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Head and Neck Soft Tissue Sarcoma

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1. Introduction

Soft tissue sarcomas are a group of heterogeneous tumors that have their origin primarily in the embryonic mesoderm; more than 50 histological subtypes and diverse clinical behaviors have been identified. Soft tissue sarcomas can range from relatively slow growth, causing little destructive growth, to being locally aggressive, regionally destructive and having a great potential for systemic metastases [Greene et al, 2002; Pelliteri et al, 2003]. The approximate incidence for this kind of neoplasia is 3-4.5/100,000 [Zahm et al, 1997], representing approximately 1% of all malignant adult neoplasias. Soft tissue sarcomas are rare in the head and neck and have an approximate frequency of 5-15% of all adult sarcomas and less than 1% of all head and neck neoplasias [Patel et al, 2001; Pandey et al, 2003; Colville et al, 2005]. The age at presentation is variable with a mean of 50 to 55 years (minimum is 3 months and the maximum is 89 years old) and the male/female ratio is approximately 2:1, which varies depending on the review series. The symptoms depend on location, but the most frequently reported symptoms are the following: headache, nasal obstruction, dysphagia, hoarseness and dyspnea. However, the majority of patients are asymptomatic [Farhood et al]. The most frequently reported involved sites include the following: the face, neck, scalp, nasopharynx, maxillary antrum, cranial base and parotid gland. However, frequencies at each site differ depending on the published series [Colville et al, 2005; Bentz et al, 2004; de Bree et al, 2006]. The histological varieties are diverse, but the most frequent are malignant fibrous histiocytoma (MFH) and fibrosarcoma [de Bree et al, 2006]. In Mexico, a total of 27 cases were reported by the National Institute of Cancerology (INCan) from 1982-1993, and the most frequent histological types were rhabdomyosarcoma and malignant peripheral nerve sheath tumors [Barrera et al, 1997]. The General Hospital of Mexico reported a total of 29 head and neck sarcomas cases 1993 to 1997, and the most frequent histological types were neurogenic sarcomas and leiomyosarcomas [Lazos et al, 1999]. The natural history of head and neck sarcomas is similar to that of sarcomas in other parts of the body; however, because of their location, they present a greater surgical difficulty, and residual disease is often left behind thus reducing the patient's life expectancy [Barrera et al, 1997]. Despite this variety of histologic subtypes, soft tissue sarcomas have some clinical and pathologic features in common. The current American Joint Committee on Cancer (AJCC) and International Union Against Cancer (UICC) staging

criteria for soft tissue sarcomas are universal for almost all histologic subtypes and rely on the histologic grade, tumor size and depth and the presence of distant or nodal metastases. Therefore, the particular subtype seems to be of less importance [de Bree et al, 2006]. The main prognostic factor for soft tissue sarcomas is histological grade and tumor size. Staging is performed according to the AJCC, which has four clinical stages ranging from I to IV [Mendenhall et al, 2005]. Regarding the evolution of the disease, head and neck sarcomas frequently present metastases, most commonly lung metastases, and the initial management therefore includes a chest X-ray or a computed tomography scan. The absence of metastatic lesions excludes the possibility of a systemic disease [Mendenhall et al, 2005]. In children, head and neck sarcomas respond appropriately when treated with chemotherapy and radiation therapy. In adults, the main treatment modality is surgical, although multidisciplinary treatment is also important because these kinds of tumors frequently invade or are in close proximity to vital structures. As a result, surgical resection could be incomplete, making it necessary to locally control the disease by means of adjuvant therapies [Le QT et al, 1997].

The purpose of this chapter is a review of frequency, clinical features, histopathology, molecular biology, metastasis and treatment of soft tissue sarcomas of the head and neck.

2. Angiosarcoma of head and neck

Angiosarcoma is a malignant neoplasm that frequently occurs in the skin and subcutis or in a visceral location and very rarely affects the oral cavity [Loudon JA et al, 2000, Oliver AJ et al, 1991]. Clinically, angiosarcoma appears as a poorly demarcated nodular tumor that is red-blue to purplish in color [Toth BB, et al, 1981]. Angiosarcomas of the head and neck most commonly involve the scalp, and only 4–5% of them form in the pharynx, oral cavity or maxillary sinus [Loudon JA et al, 2000, Lanigan DT, et al 1989]. They may represent either primary or metastatic lesions. In a recent review of the literature, only 23 cases were reported to involve the head and neck, with the exception of the scalp, at an age of presentation ranging from 1 day to 68 years [Abdullah BH et al, 2000, Loudon JA et al, 2000]. Angiosarcomas predominantly affect elderly men and may be present in any region of the body, but they usually occur in the skin or superficial soft tissues (head and neck) in the post-radiotherapy area. The prognosis is poor because of frequent local recurrence and metastatic spread to the lymph nodes, bones (vertebrae) and lungs [Forton Glen EJ et al, 2005]. Primary angiosarcoma of the non-irradiated parotid gland is extremely rare. There are only a few articles in literature that discuss patients with angiosarcoma, but most of these angiosarcomas affected the irradiated region or originated in the skin and secondarily affected the parotid gland. All angiosarcomas tend to be aggressive, and they are often multicentered. These tumors have a high local recurrence rate and metastasize because of their intrinsic biological properties and because they are often misdiagnosed, which leads to a poor prognosis and high mortality rate. This malignant vascular tumor is clinically aggressive, difficult to treat, and has a reported five-year survival rate of less than 20%. Advanced stage at presentation and lack of extensive excision are associated with higher recurrence, distant metastasis rates, and worsened survival [Mahdhaoui A et al, 2004]. About 30% of oral metastases are the first sign of an undiscovered malignancy at a distant site. Angiosarcomas represent 1% of all soft tissue sarcomas. Their clinical outcomes are poor due to a rapid growth and high risk of metastatic extension. The median overall

survival of all clinical types of angiosarcomas ranges between 15 and 30 months [Penel N et al, 2003, Favia G et al, 2002]. Angiosarcoma can occur in pre-existing benign or intermediate vascular lesions; there are several case reports of angiosarcomas arising spontaneously from hemangiomas or vascular malformations. The mechanism of malignant transformation in benign vascular tumors is unclear because there are few case reports [Hunt SJ et al, 2004]. A history of trauma may be an etiological factor in angiosarcomas, but most authors think that a traumatic event only alerts the patient to a lesion that already existed. Other factors that have been implicated include hormonal influences from anabolic steroids, synthetic estrogens or pregnancy, exposure to environmental toxins, such as thorium dioxide, vinyl chloride, thorotrast (used for angiography in the past), or insecticides. Immunohistochemical analysis is an important adjunctive diagnostic approach for angiosarcoma [Koch M et al, 2008]. The tumors are usually positive for factor VIII-related antigen, vimentin, CD31, CD34 and UEA-1. Among these factors, CD31 is positive in almost 80– 90% of angiosarcomas, with relatively good specificity and excellent sensitivity [Morgan MB et al, 2004]. The microscopic appearance of angiosarcoma varies from epithelioid to spindled areas, with the former being more common. Various prognostic factors have been reported, including older age, tumor size larger than 5 cm, high grade, positive margin and lymphedema field location [Skubitz KM et al, 2005]. A prospective clinical study with a larger sample size is needed to determine the prognostic factors in angiosarcoma patients. Early diagnosis is important for early treatment. A medical history should be obtained, and a thorough physical examination should be performed. Magnetic resonance imaging (MRI) and contrast-enhanced computed tomography (CT) are nonspecific for diagnosis, but they may be used to define the extent of the primary tumor and evaluate distant metastasis. The diagnosis of angiosarcoma can only be established by microscopic examination. The macroscopic and microscopic appearance of this tumor can lead to a misdiagnosis of pyogenic granuloma. The differential clinical diagnoses should include pyogenic granuloma, giant cell granuloma, Kaposi sarcoma, hemangioma, and malignant melanoma [Mullick SS et al, 1997]. Likewise, the differential microscopic diagnoses should include hemangioma, hemangiopericytoma, papillary endothelial hyperplasia, angiolymphoid hyperplasia with eosinophilia, Kaposi sarcoma, malignant melanoma, metastatic renal cell carcinoma, and pyogenic granuloma [Mullick SS et al, 1997]. Treatment of angiosarcomas is greatly complicated by the diffuse infiltration typical of these tumors. Various interventions have been attempted to stem the disease process. In a recent detailed treatment analysis, surgery in combination with radiation therapy affords the most favorable opportunity for angiosarcoma control [Mark RJ et al, 1996]. The prognosis for patients with angiosarcoma is generally considered to be rather poor, although tumor size (and hence stage), site, and the histopathologic grade may influence survival. Investigations indicate that one-half of patients die within 15 months of diagnosis, with only approximately 12% surviving 5 years or longer. However, those patients presenting with lesions less than 10 cm in diameter respond better to therapy [Mentzel T et al, 1998].

Recurrence after local treatment manifests primarily in local failure, yet distant metastasis is not insignificant in the post-treatment failure group [Mark RJ et al, 1996]. These factors, both for local and distant spread, are reflective of the highly aggressive nature of this illness and serve to explain the poor survival statistics.
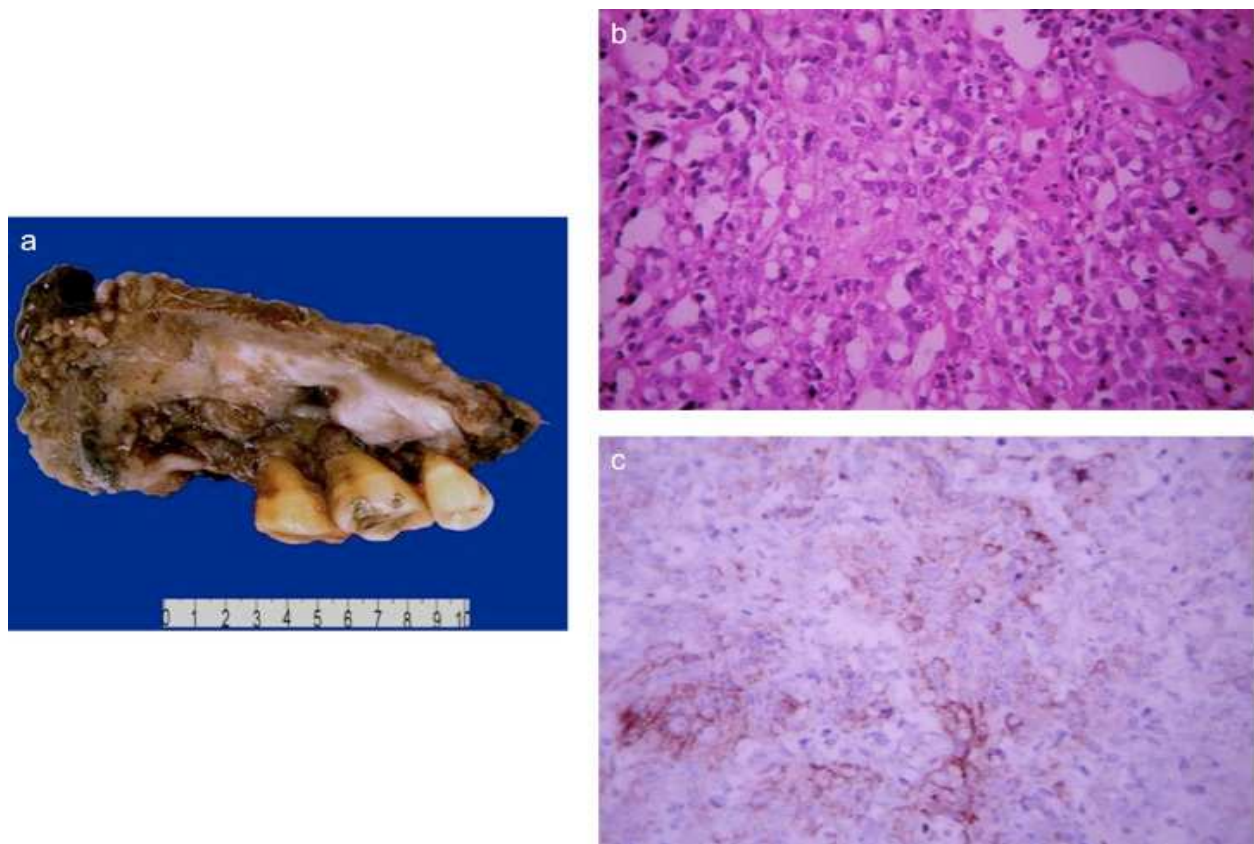
Fig. 1. a) Angiosarcoma of the maxilla in an elderly woman, b) Epithelioid angiosarcoma of oral mucosal maxillar region, 40X, c) Inmunostain for factor VIII shows intense staining of most angiosarcoma cells, 40X.

3. Fibrosarcoma of head and neck

Fibrosarcoma is defined as a malignant spindle cell tumor that shows a herringbone or interlacing fasicular pattern without the expression of other connective tissue cell markers [Sapp JP et al, 2004]. Fibrosarcoma can arise in soft tissues or within bones. Intraossesous fibrosarcomas may develop enosteally or possibly periosteally, affecting the bone by spreading from adjacent soft tissue. Fibrosarcomas can occur in any location, but the bone extremities are the main affected sites; occurrence in the maxilla is rare, with an incidence ranging from 0-6.1% of all primary fibrosarcomas of the bone. The mandible is the most common site for fibrosarcomas [Soares et al 2006, Pereira CM et al, 2005]. The clinical behavior of fibrosarcoma is characterized by a high local recurrence rate and a low incidence of locoregional lymph node and/or distant hematogenous metastases. However, hematogenus metastases may involve the lungs, mediastinum, abdominal cavity and bone [Conley J et al, 1967]. Local recurrence poses a serious and complex problem, particularly with occurrence of mediastinum infiltration, local destruction, airway compression, esophageal compression and extension. Radiation therapy is generally considered only in cases for which resection is impossible; chemotherapy is only used for palliative treatment. Prognosis is directly related to adequate, complete resection, which obviously requires early detection before the extensive involvement of soft tissue [Lukinmaa P et al, 1988].

The histological appearance of fibrosarcoma does not allow a distinction between a tumor of the bone from one arising in soft tissue [Chen Y et al, 2007]. Histologically, the degree of differentiation is variable, from being comparable to a benign fibroma to a highly anaplastic tumor, thus presenting a diagnostic dilemma to histopathologists. Fibrosarcoma can be graded as either a low or high grade of malignancy. Low-grade fibrosarcoma shows spindle cells arranged in fascicles with low to moderate cellularity and a herringbone appearance. This type of fibrosarcoma has a mild degree of nuclear pleomorphism and rare mitosis, with a collagenous stroma. High-grade lesions show an intense nuclear pleomorphism, greater cellularity and atypical mitosis. The nuclei can be spindle shaped, oval or round. The histological appearance of high-grade fibrosarcoma may be similar to other tumors, such as malignant fibrous histiocytoma, liposarcoma or synovial sarcoma. The positive immunostaining for vimentin, together with negative staining for muscular immunomarkers, helps to diagnose fibrosarcoma [Wadhwan V et al, 2010].

3.1 Sclerosing epithelioid fibrosarcoma of head and neck

Sclerosing epithelioid fibrosarcoma was originally described in 1995 by Meis-Kindblom et al [Meis-Kindblom JM et al, 1995], as an uncommon low-grade variant of fibrosarcoma that is found mainly in the deep soft tissue of the extremities. It is characterized by distinctive epithelioid cytomorphology associated with extensive stromal hyalinization. Tumors with definitive intraosseous origin are uncommon, despite the fact that sclerosing epithelioid fibrosarcoma has a predilection for bone invasion [Antonescu CR et al, 2001]. Histologically, it is characterized by a multinodular proliferation of sheets, nests, or cords of uniform round to oval epithelioid cells with distinctive cell borders. The cytoplasm of the tumor cells is pale eosinophilic to clear, and the nuclei are rounded, although in some areas where the tumor cells are tightly packed together, nuclear angulation may be evident. These cells are embedded in a densely hyalinized stroma. Areas of myxoid degeneration, cystic change, foci of metaplastic bone, hyaline cartilage, and calcification are additional features that may be present [Eyden BP et al, 1998]. Immunohistochemically, the tumor cells of sclerosing epithelioid fibrosarcoma do not display definitive evidence of differentiation along a specific lineage. Most examples are immunoreactive with vimentin, but variable staining for EMA, S100, AE1/AE3, Bcl-2, CD34 and CD99 has been reported [Antonescu et al, 2001, Donner LR et al, 2000].

3.2 Ameloblastic fibrosarcoma

Ameloblastic fibrosarcoma, first described by Heath in 1887, is an extremely rare malignant odontogenic tumor. It is composed of a benign odontogenic epithelium and a malignant ectomesenchymal component. It is regarded as the malignant counterpart of the ameloblastic fibroma. Ameloblastic fibrosarcoma normally presents as a painful swelling and intraosseous mass (2-6 cm), with occasional ulceration in the posterior regions of the mandible and/or maxilla. The posterior mandibular area is the most commonly affected site; the disease is more likely to occur in males than females (1.6:1). Ameloblastic fibrosarcoma occurs in a wide age range, from 3 to 83 years (mean age, 27.3 years). Only one case of peripheral presentation has been reported. The histopathology of ameloblastic fibrosarcoma is characterized by a consistent appearance in which a malignant ectomesenchymal component is mixed with a benign epithelial odontogenic component; the malignant ectomesechymal component consistently takes up more than 70% of the tumor

area compared with 30% by the odontogenic epithelium. Ameloblastic fibrosarcoma resembles a malignant connective tissue. The World Health Organization distinguishes odontogenic sarcomas devoid of dental hard tissue ameloblastic fibrosarcoma from those displaying focal evidence of dentinoid (ameloblastic fibrodentinosarcoma) or dentinoid plus enameloid (ameloblastic fibro-odontosarcoma) but acknowledges that the presence or absence of dental hard tissue in an odontogenic sarcoma is of no prognostic significance. This sacorma has an unknown etiology, with some cases representing malignant transformation of a preexistent ameloblastic fibrosarcoma. Although approximately two-thirds of ameloblastic fibrosarcomas seemed to have arisen de novo, several authors have demonstrated that ameloblastic fibroma to be the precursor of ameloblastic fibrosarcoma, i.e., malignant transformation. This neoplasm has a higly localized behavior with low potential for distant metastasis. The treatment of choice is the radical extensive surgery, usually necessitating partial or total mandibulectomy. The prognosis of ameloblastic fibrosarcoma seems better than that for other fibrosarcomas of the orofacial region [Carlos R et al, 2005, Reichart PA et al, 2004]

4. Malignant fibrous histiocytoma of head and neck

Malignant fibrous histiocytoma (MFH) was first described by Ozzelo et al, in 1963 [Pezzi CM et al, 1992] and by O'Brien and Stout in 1964 [Sabesan T et al, 2006]. It was widely accepted as a clinicopathological entity after the description of cases by Kempson and Kyriakos in 1972 [Barnes L et al, 1988]. The etiology is unknown, and the histiogenesis remains controversial. Several hypotheses have been suggested, including an origin from true histiocytes, fibroblasts, both fibroblasts and histiocytes, or from primitive mesenchymal cells. [Ogawa A et al, 2005].

Malignant fibrous histiocytoma is now recognized as one of the most common soft tissue sarcomas in adults. In addition to occurring in soft tissue, it can also occur as a primary intraosseous tumor in bones. It affects, in order of frequency, the lower extremity, the upper extremity, the retroperitonoum and abdominal cavity, and lastly, the head and neck, where it accounts for 1–3% of all cases [Gibbs JF et al, 2001]. Therefore, it is relatively uncommon in the head and neck region. Surgery is the most reliable treatment for MFH, but the five year survival rate for MFH in the head and neck is low compared with MFH in the extremities and trunk. In the head and neck, MFH has been observed in the nasal sinus, salivary gland, oral cavity, mandible, larynx, auricula and eyelid, and Barnes et al [Barnes L et al, 1988], reported that MFH onset occurs most commonly in the accessory nasal sinuses, followed by the salivary glands. In a study of 11 sarcomas of the parotid gland, histological classification showed that three of the lesions were MFH and two cases each of neurosarcoma, rhabdomyosarcoma, fibrosarcoma, and osteosarcoma. A single case of MFH in the buccal region was also reported [Ogawa A et al, 2005]. In the literature, in cases of MFH in the head and neck, the onset age ranged from 16 years old in a patient with MFH in the mandible to 85 years old in a patient with MFH in the eyelid, and the mean age of 12 patients with head and neck MFH was reported to be 55 years old [Khong JJ et al, 2005, Barnes L et al, 1988, Narvaez JA et al, 1996]. The occurrence of MFH in membranous bones is unusual. Involvement of the mandible accounts for only 3% of all MFH of the bone. MFH of the head and neck that extend into bony structures are associated with a much more aggressive clinical course than those that are restricted to soft tissues [Rinaldo A et al, 2004, Iguchi Y et al, 2002] The occurrence of MFH in the head and neck region was primarily in middle-aged

adults (mean age, 45 years), with men affected more frequently than women. The sinonasal tract is the most common site of origin, followed by the soft tissue of the face and neck, the oral cavity, and the craniofacial region [Park SW et al, 2009]. The signs and symptoms of MFH of the maxilla include swelling of the cheek, facial pain, nasal obstruction and rhinorrhea. The rarer symptoms include infraorbital nerve paresthesia, visual disturbance, and ocular proptosis. In cases of MFH of the mandible, Kanazawa et al [Kanazawa H et al, 2003]. reported in their review that those lesions are usually first noticed due to swelling, paresthesia, and loosening of teeth. A history of antecedent trauma in about 20% of the cases suggests that some of these tumors may represent an initial proliferative response to the trauma [Senel FC et al, 2006]. Although radical tumor resection with adequate tumor-free margins is essential, from the anatomic point of view, it is often difficult to perform in the head and neck region [Kearney MM et al, 1980]. Park et al [Park SW et al, 2009]. reported in a review that CT and MRI features of MFH of the head and neck have also been nonspecific. On CT scans, MFH is usually seen as a large lobulated soft-tissue mass, which is isoattenuated to muscle. Sato et al [Sato T et al, 2001] . reported that lesions of the maxilla sometimes present radiographically as fairly well-demarcated bone margins, with uniform density or no necrotic areas, and with a clear separation from surrounding soft tissues in CT images, which lead to a misdiagnosis of low-grade malignant tumors or benign tumors. On the MRI, MFH is seen as a heterogeneous hyperintense pattern on T2-weighted images, with an isointensity that is almost the same as that of the muscles on T1-weighted images [Park SW et al, 2009]. Intraosseous MFH has a tendency to indicate a poor prognosis. However, in the maxillary area, it is difficult to differentiate intraosseous MFHs from extraosseous MFHs and to definitively determine the origin when these MFHs were large in size and had aggressive bone involvement [Yamaguchi S et al, 2004]. Also, survival rates of MFH of the maxilla are difficult to evaluate due to the small number of documented cases Chan YW et al, 2004]. With regard to sinonasal MFH, the five year disease-free survival rate and the five year overall survival rate are only 21.5% and 25.1%, respectively [Wang CP et al, 2009]. Anavi et al [Anavi Y et al, 1989]. reported in their review of mandibular MFH that the overall survival estimate at five years was 46% regardless of the type of treatment. Clinical stage, histological grade of malignancy, and local recurrences were the most important prognostic factors for MFH in the bone. Huvos et al [Huvos AG et al, 1985]. suggested that metastatic spread in patients with MFH primarily in the bone was not to the regional lymph nodes, but rather a hematogenous dissemination predominantly to the lungs. The reported frequency of nodal metastases for head and neck MFH varies between 0% and 15%. Prognosis differs according to the morphologic subtypes of MFH [Park SW et al, 2009], Derbel F et al [Derberl F et al, 2010], reported a case of MFH in the neck with metastases to liver. Metastases from MFH in liver are rare, representing 1% of reported metastasis of MFH [Derbel F et al, 2010]. Metastasis occurs in 42% of cases. Lung (82%) and lymph nodes (32%) metastases are most frequent [Derbel et al, 2010]. Factors that influence the rate of metastasis included depth, size and the inflammatory component of the tumour [Derbel F et al, 2010].

The myxoid type has a higher rate of local recurrence, with recurrence in approximately 50–60% of patients and with an overall risk of subsequent metastases at 20–35% [Marotta D et, al, 2009, Mentzel T et al, 2002]. Histologically, the tumor contains both fibroblast-like and histiocyte-like cells in varying proportions, with spindle and round cells exhibiting a storiform arrangement. These tumors have been divided into four morphologic subtypes that depend on the predominant cellular components: storiform – pleomorphic (50–60%), myxoid (25%), giant cell (5–10%), and inflammatory (about 5%). The myxoid variant has

been reported to have better prognosis when compared with the storiform-pleomorphic type [Park SW et al, 2009]. In the treatment of MFH of the region, adjuvant chemotherapy is considered for high-grade tumors because these tumors may present subclinical or microscopic metastases at the time of diagnosis [Pereira CM et al, 2005]. The effectiveness of surgery in combination with radiotherapy and/or chemotherapy has not been well established. Three-drug regimens with high-dose MTX, CDDP and DOX or four-drug regimens with high-dose IFO, high-dose MTX, CDDP, and DOX used in patients with osteosarcoma have been evaluated.
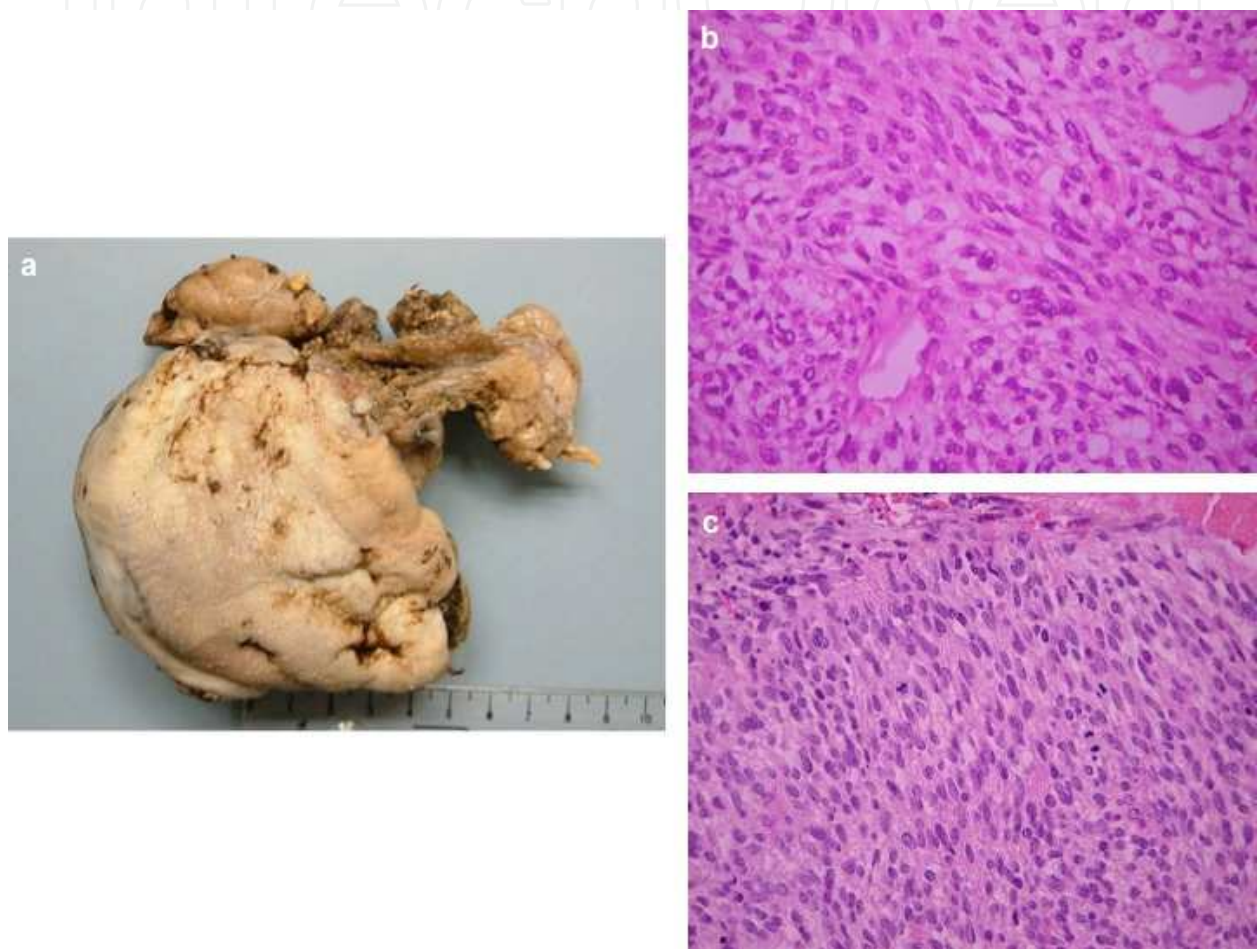

Fig. 2. a) Malignant fibrous histiocytoma of the dorsum tongue , b) Gross appearance of intramuscular malignant fibrous histiocytoma involving tongue, c) Malignant fibrous histiocytoma with fascicular pattern degree nuclear atypia and mitotic activity.

The regimen of neoadjuvant chemotherapy with three or four drugs has been tested in a randomized study of MFH in the bone [Hugate RR et al, 2008]. Few studies estimate the survival benefits of chemotherapy with IFO, CDDP and DOX for osteosarcoma [Zalupski MM et al, 2004]. Further research is needed to determine if adjuvant chemotherapy with the three-drug combination of IFO, CDDP and DOX would be effective for MFH of the jaws. Radiotherapy alone may be reserved for inoperable patients and patients with high surgical risk or those with regional or systemic metastases. Incomplete excision may lead to a high rate of recurrence. High rates local recurrence of MFH in the bone are due to the fact that this tumor infiltrates skeletal muscle fibers and fascial planes [Enjoji M et al, 1980]. Complete

excision has been achieved under frozen section control, and all margins were free of tumor. The prognosis depends on the size and site of the tumor and its malignant potential in terms of metastasis. The lungs are the most common site of metastasis. Post-radiation sarcomas have been reported to have poor prognosis [LinkTM et al, 1998]. One patient, however, has been reported to have long-term disease- free survival despite earlier exposure to radiation. Overall, the five year survival for mandibular malignant fibrous histioctyoma has been reported to be 46%, irrespective of the treatment type [Anavi et al, 1989].

5. Leiomyosarcoma of head and neck

Leiomyosarcoma is a malignant tumor derived from smooth muscle that accounts for 5–6% of all soft tissue sarcomas. Leiomyosarcoma usually occurs in the soft tissues of the extremities and trunk; only 3% of leiomyosarcomas are in the head and neck. In the head and neck region, most leiomyosarcomas occur in the nasal cavity and paranasal sinuses, mouth, and larynx [Marioni G et al, 2000]. The signs and symptoms of leiomyosarcoma involving the head and neck region depend on the site and the size of the tumor. Hoarseness, stridor, dyspnea and dysphagia are the most common complaints of the laryngeal and the parapharyngeal tumors. While the initial symptom in a patient with laryngeal leiomyosarcoma in one report was dysphonia, the main symptom in a second patient with parapharyngeal leiomyosarcoma was dysphagia due to the mechanic compression of the cervical esophagus. The scarcity of the smooth muscle in the head and neck region may be the probable reason for the rarity of leiomyosarcoma. Because blood vessels are the only structures in the larynx and parapharyngeal region with smooth muscles, leiomyosarcoma may develop from the smooth muscle in the tunica media of vessel walls. Aberrant mesenchymal differentiation and metastasis are the other possible modes of origin [Freije JE et al, 1992, Chen JM et al, 1991]. Leiomyosarcoma that originate in the head and neck region are very rare. Leiomyosarcoma of the hypopharynx has been reported only four times in the English medical literature. The rarity of a leiomyosarcoma in the hypopharynx can be attributed to the limited smooth muscle tissue in the hypopharynx. A possible candidate for the origin of leiomyosarcoma in the hypopharynx is the smooth muscle surrounding blood vessels. Among the four cases presented in these previous reports, two were located in the posterior wall of the hypopharynx, one in the pyriform sinus, and one in the postcricoid. The age at onset was 39–65 years (mean age, 55.4 years).

The lack of any distinguishing clinical features and the rarity of these lesions often result in their being mistaken for the more common lesions affecting the oral cavity, and correct diagnosis is made only following definitive histological examination [Freedman AM et al, 1989, Cocks H et al, 1999]. The sites of the tumors in this series are similar to the regional variation within the oral cavity noted in the literature, with the maxilla being the most commonly involved site followed by the mandible, tongue, cheek and floor of mouth, in descending frequency. The cause of this apparent variation is unknown and different from benign leiomyomas, which occur more frequently in the lips, tongue, cheeks and palate. Almost 65% of reported tumors are in the maxilla or mandible, and they often involve the jaw bones [Dry SM et al, 2000, Kratochvil FJ et al 1982, Montgomery E et al, 2002]. In cases of head and neck leiomyosarcoma, the success of initial surgical management is an important prognostic factor because complete surgical excision is associated with low local recurrence and longer survival. Resection with microscopically tumor-free margins is of

paramount importance for long-term survival. Adjuvant radiation treatment or chemotherapy appears to be ineffective in achieving local control when there is residual tumor postoperatively [Dry SM et al, 2000]. However, gemcitabine has been one of the few agents that shows activity in these tumors, with an observed response rate of 20.5%. Hensley et al [Hensley ML et al, 2002], combined gemcitabine with docetaxel and reported a response rate of 53% in 34 patients, some of whom had failed doxorubicin therapy. The cause of leiomyosarcoma remains uncertain, although cases may be associated with trauma, oestrogenic stimulation, and ion involvement. Other publications suggest that the prognosis of oral leiomyosarcoma that affects the tongue is good if clear excision can be achieved. In the previous eight cases reported in which follow-up ranged from one to five years, there has been no evidence of recurrence [Bass B et al, 1986]. This suggests that primary leiomyosarcoma has a better prognosis if it arises within the tongue than elsewhere in the body. The recurrence rate for tumors that arise from and are limited to the dermis is reported to be between 14% and 42% [Farman AG et al, 1977]. Histopathologically displayed a prominent spindle cell component, with the cells arranged in intersecting fascicles, containing characteristic `cigar-shaped' nuclei. These features are typical for leiomyosarcoma. Nevertheless, similar findings may occur in a wide number of diferent neoplasms, such as fibrobrosarcoma, myofibrosarcoma, synovial sarcoma, solitary fibrous tumor, malignant peripheral nerve sheath tumors (MPNSTs), spindle cell rhabdomyosarcoma, spindle cell liposarcoma, spindle cell carcinoma and other spindle cell neoplasms [Izumi et al, 1995]. The arrangement of neoplastic fascicles in perpendicular, intersecting bundles, the peculiar `cigar-shaped' nuclear morphology, the occurrence of paranuclear vacuolization and intracytoplasmic PAS-positive granularity are indicative, but not exclusive, of leiomyosarcomatous diferentiation. In addition, the diagnosis of spindle cell carcinoma should always be ruled out when a spindle cell neoplasm occurs in a visceral location. In most instances, immunohistochemistry may provide useful clues to the diagnosis; muscle-specific markers for smooth muscle actin and vimentin are most frequently detectable, but myofibroblastic tumor cells, rhabdomyosarcoma and, inconstantly, spindle cell carcinomas may display a similar immunoprofile. Cytokeratin and EMA negativity, as in the case reported herein, rules out the diagnosis of spindle cell carcinoma as well as that of synovial sarcoma, the latter being vimentin and CD99 positive. Desmin immunoreactivity is not detectable in myofibroblasts, neither normal or neoplastic, but is encountered in other myogenic neoplasms, such as rhabdomyosarcoma [Muzio L et al, 2000]. Although numerous prognostic factors, including size, site, grade and TNM stage, have been identified for leiomyosarcoma arising in other sites, there are no reliable prognostic factors in the case of primary oral leiomyosarcoma. The TNM classification of soft-tissue tumors is not directly applicable to oral leiomyosarcoma, especially in terms of the T stage, and the histological grade of the tumor has rarely been reported in oral leiomyosarcoma. It is important to identify factors of prognostic significance in this patient group. The estimated five year survival for the whole group is 55%. Tumors that demonstrated bony involvement (maxilla/mandible) and metastasis were associated with poorer prognosis. Increasing age and male gender showed a trend toward worse prognosis, although this was not statistically significant. Interestingly, neither the increased size of the tumor nor recurrence was associated with poor survival, unlike tumors occurring at other sites [Miyajima K et al 2002, Weiss SW et al, 2001, Dry SM et al, 2000, Nikitakis NG et al, 2002].

6. Liposarcoma of head and neck

Liposarcoma is the second most common soft tissue sarcoma in adults. Its occurrence in the head and neck region is reported to be very rare. The majority of liposarcomas occur in middle-aged adults; however, very uncommon cases of liposarcoma can be generated in infancy and early childhood [Hicks J et al, 2001]. The occurrence of liposarcoma in the head and neck region is rare, comprising 5.6–9% of all cases [Enzinger FM et al, 2001]. Liposarcoma of the oral cavity is even less frequent, making up about 10% of all cases in the head and neck. In a 1995 literature review, Golledge et al [Golledge J et al, 1995]. found that liposarcoma of the head and neck was poorly addressed and that its incidence is rare, representing approximately 4-5% of liposarcomas (50 reported cases). Most cases originated in the neck (28%), followed by the head (scalp and face, 26%), larynx (20%), pharynx (18%), and mouth (8%) [Golledge J et al, 1995].

Since the publication of that review, liposarcomas of the head and neck have gained more attention; however, most of the subsequent reports have addressed the primary liposarcomas of the head and neck. Fifteen cases of liposarcoma metastases to the head and neck region have been previously reported. The associated primary tumors from these reports occurred most fequently in the thigh and retroperitoneum and equally between men and women, showing no deviance from the normal trends found in liposarcoma. The average age of the patients was 53 years old. Most of the head and neck metastases were to the orbit, thyroid, and dura mater in 31%, 25%, and 19% of the cases, respectively. Other metastatic foci included the gingival mucosa, submandibular region, and scalp [McElderry et al, 2008]. Although head and neck liposarcomas are rare, specialists should be aware of the natural history, prognosis and treatment. As with liposarcomas elsewhere in the body, most cases present in adults, and there is a male predominance. Factors considered to be important in the etiology of liposarcomas include genetics, trauma and irradiation. Usually, the tumor appears firm, relatively fixed to adjacent tissues, encapsulated, and to be growing steadily but not rapidly. Patients do not usually have regional lymph node or distant metastasis at presentation [Feles RA et al, 1993, Freedman AM et al, 1989]. Radiological investigations are necessary in most cases to determine precise size, localization, limits, extensions of the tumor and its relations with neurovascular structures. It is also necessary for detecting distant metastases [Enzinger FM et al, 2001].. Liposarcoma originates from primitive mesenchymal cells rather than mature fat cells. Thus, liposarcoma seldom originates from normal fat tissue or lipoma. Furthermore, in contrast to lipoma, which ordinarily arises in the subcutaneous, submucous, or subserous tissue, liposarcoma rarely arises in such tissues, but rather in the perimuscular or perifascial structures of spindle cell liposarcoma. The next major type is the myxoid type, which is composed of three main tissue components: proliferating lipoblasts at varying stages of differentiation, a delicate plexiform capillary pattern, and a myxoid matrix containing abundant nonsulfated glycosaminoglycans. A subset of myxoid liposarcomas shows histological progression to hypercellular or round cell morphology, which is associated with poor prognosis [Christopher DM et al, 2002]. The most infrequent type is pleomorphic liposarcoma, which contains huge lipoblastic multinucleated cells by which it distinguishes itself from other pleomorphic sarcomas, such as malignant fibrohistiocytoma.The dedifferentiated type was added to these criteria recently and is composed of a well-differentiated area and a poorly differentiated area in parts of the same neoplasm, recurrent tumor or metastatic lesion. The prognosis of this tumor depends on the tumor subtype; well-differentiated and myxoid

liposarcomas are considered to be low-grade malignancies, whereas the pleomorphic and round cell types are regarded to be high-grade. Golledge et al [Golledge J et al, 1995]. reported 5-year survival rates of 100% for well-differentiated, 73% for myxoid, 42% for pleomorphic, and 0% for round cell liposarcomas in 76 cases in the head and neck area. The best choice for treatment is complete surgical excision, where a wide or radical excision with sufficient margins is possible. McCulloch et al [McCulloch et al, 1992]. reported an 80% rate of local or distant recurrence in patients with incomplete surgical excision compared with a rate of only 17% when complete excision was accomplished.

Liposarcoma appears to have a clear capsule and is easy to separate, but it often infiltrates the surrounding structures microscopically, so occasionally, incomplete resection occurs. In contrast to its characteristic high recurrence, lymph node metastasis is quite rare, so neck lymph node dissection is considered unnecessary [Weing BM et al, 1995]. Cases of distant metastasis, which result from hematogenous metastasis, are occasionally reported, and the most common metastatic sites are the lung, intra-abdominal area, bone, skull and liver [Wong CK et al, 1997]. In the head and neck area, the majority of metastasized cases appear to be round cell or pleomorphic liposarcomas. Radiotherapy, particularly postoperative radiation, is considered useful and might delay or prevent local recurrence.

7. Rhabdomyosarcoma of head and neck

Rhabdomyosarcomas (RMS) account for 40% of all sarcomas found in the head and neck region, and they are a morphologically and clinically heterogeneous family of malignant soft tissue tumors of a myogenic lineage [Abali H et al, 2003]. Alveolar rhabdomyosarcoma (ARMS) and embryonal rhabdomyosarcoma (ERMS) represent the two main histologic patterns and must be differentiated from other small round cell tumors. RMS is the most common soft tissue sarcoma in the pediatric population, comprising approximately 5% of all childhood cancers and nearly 50% of soft tissue sarcomas arising in children 0 to 14 years of age [Ferlito A et al, 1999]. By contrast, RMS is remarkably uncommon in older adults, representing merely 2-5% of all malignant soft tissue tumors, with the majority being the pleomorphic subtype [Sivanandan R et al, 2004]. Head and neck tumors are divided into three major groups based on anatomic location and propensity for invasion of the central nervous system: orbital, parameningeal, and nonparameningeal. Parameningeal tumors carry the worst prognosis. Orbital RMS represents 75% of those tumors in the head and neck and is associated with the best prognosis. Oral rhabdomyosarcomas are classified within the non-orbital, nonparameningeal group of tumors, which present a better prognosis and tend not to invade the central nervous system. The five-year survival rate is approximately 85% for this RMS subtype. RMS is more aggressive in adults compared with children. Poor prognosis in adults is thought to be due to a combination of advanced tumor stage, unfavorable histology, a decreased tolerance to treatment and other unknown biological factors [Franca CM et al, 2006, Bras J et al 1987, Pavithran K et al, 1997]. Alveolar rhabdomyosarcoma is known to be rare in adults aged over 45 years and to be commonly located in the extremities; the clinical features of the case presented were exceptional. As a result of the lack of characteristic histopathologic features, the final diagnosis of alveolar rhabdomyosarcoma is difficult to establish. The histochemical findings suggested differentiation from other small anaplastic round cell tumors, such as undifferentiated carcinoma, neuroblastoma, neuroepithelioma, Ewing's sarcoma, and malignant lymphoma. The immunohistochemical character of the tumor cells increases the diagnostic accuracy;

positivity for vimentin, MSA, and desmins confirm the diagnosis. Rhabdomyosarcoma is a malignant neoplasm consisting of undifferentiated mesodermal tissue that expresses myogenic differentiation. Histopathologic diagnosis is based on conventional light microscopy and confirmed by immunhistochemistry. Antibodies against desmin, muscle-specific actin, and myoglobulin are most widely used for diagnostic purposes. The three major morphologic categories of rhabdomyosarcomas are embryonal, alveolar, and pleomophic. The embryonal subtype is the most common, accounting for 70–75% of all rhabdomyosarcomas, followed by the alveolar (20–25%) and pleomorphic differentiations (5%). While the embryonal and alveolar subtypes are most commonly seen in children – therefore termed juvenile - the pleomorphic subtype occurs almost exclusively in adults. Rhabdomyosarcomas are generally seen in children, usually consisting of the embryonal type, which represents the most frequent form of soft tissue sarcomas at this age. The pleomorphic subtype is one of the most malignant sarcomas. In a publication in 1998, Akyol [Akyol MU et al, 1998] reviewed the 13 documented cases of laryngeal pleomorphic rhabdomyosarcoma. Radiotherapy as the primary treatment has been reported with varying success, while adjunctive radiation therapy is given whenever a tumor is not completely surgically removed. Randomized trials on adult sarcoma in general found that
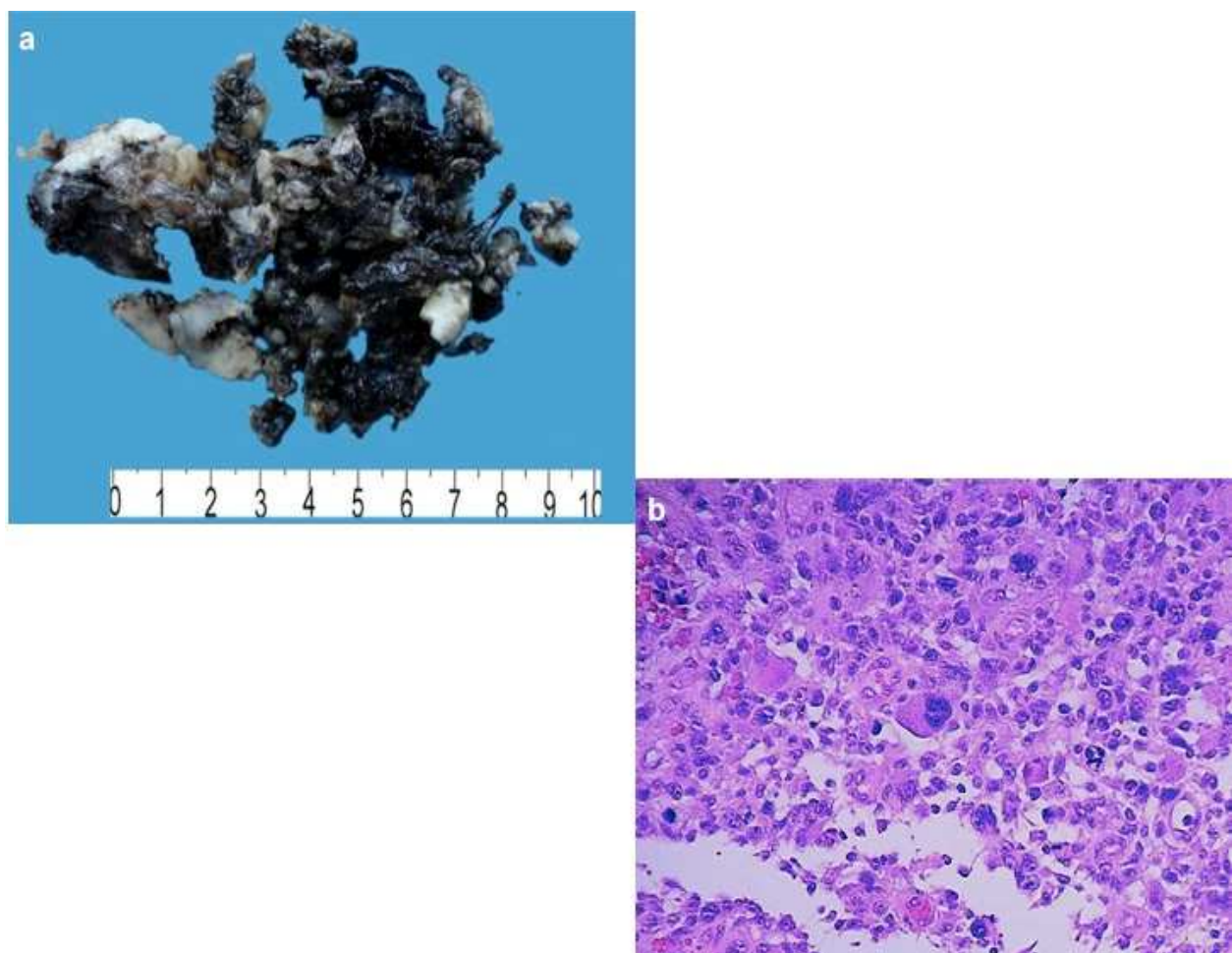

Fig. 3. a) Rhabdomyosarcoma of larynx showing a multinodular white and brown mass, b) Pleomorphic Rabdomiosarcoma with large cells containing deeply eosinophilic rhabdomyosarcoma

the addition of radiation resulted in significant improvement in local control over surgery alone. However, rhabdomyosarcomas in adults are not as radiosensitive as those in children [Haerr RW et al, 1987, Little DJ et al, 2002].

8. Synovial sarcoma of head and neck

Synovial sarcoma is a malignant soft tissue neoplasm that occurs most frequently in the extremities of young adults, near large joints [Enzinger FM et al, 2001]. The most common site is around the knee. As opposed to most other soft tissue sarcomas, these lesions are occasionally painful [Devita TJ et al, 2001]. Synovial sarcoma is more common in males than in females, although there is no evidence of a race difference. This sarcoma represents 5.6-10% of all soft tissue sarcomas. The tumor usually occurs in close association with tendon sheaths, bursae, and joint capsules, primarily in the paraarticular regions of the extremities. The origin of synovial sarcoma remains unknown, but the neoplasm is thought to arise from primitive undifferentiated pluripotential mesenchymal cells unrelated to synovial tissue [Grayson et al, 1998]. Although 85% of synovial sarcomas arise in the extremities, sites in the head and neck, trunk, abdomen, pelvis, mediastinum and lung are rarely involved. Approximately 3.7% of patients of all ages with synovial sarcoma have it in the head and neck [Ferrari A et al,2004]. Jernstrom [Jernstrom P et al, 1994] was the first to report the occurrence of synovial sarcoma in the head and neck region in 1954, and since then, more than 100 cases have been reported. The parapharyngeal region is the most frequently affected site, and few cases have been documented to arise within the orofacial region [Almeida – Lawall M et al, 2009]. The most important and accurate prognostic factors in synovial sarcoma are the extent of tumor resection and the presence of metastatic disease, the most common sites of which are the lung and regional lymph node [Rangheard AS et al, 2001]. The other prognostic factors associated with synovial sarcoma are conflicting. Previously reported favorable prognostic factors include age below 20-25 years, tumor size smaller than 5 cm, distal extremity location, and biphasic histological type. However, some studies have shown that age, location of the tumor, and histology type are not significantly associated with prognosis. In many instances, synovial sarcomas are so poorly differentiated that they do not show any specific features sufficient to suggest their true origin, and hence, they may be confused with other poorly differentiated sarcomas [Limon J et al, Turc – Carel et al, 1986].

The following factors have potential prognostic value: age at diagnosis, sex, tumor site, size, histology, mitotic count, necrosis, histological grade, stage, surgical margin status, and fusion type. The presence of poorly differentiated areas was the strongest prognostic factor associated with local recurrence, metastases, and tumor-related death [de Silva MV et al, Guillou J et al, 2004]. Three histologic subtypes originating from the presence of 2 cell types exist along a continuous spectrum: biphasic, monophasic (predominantly fibrous or rare epithelial), and poorly differentiated. Monophasic epithelial synovial cell sarcoma and gland-predominant biphasic synovial cell sarcoma are similar to adenocarcinoma, leading to potential misidentification as adenocarcinoma [Weinreb et al, 2008]. The poorly differentiated type poses a diagnostic challenge and has structures that resemble those of high-grade small round cell tumors: high cellularity, frequent mitosis, and necrosis. The common diagnosis of synovial cell sarcoma is the biphasic pattern or monophasic fibrous type but can also be easily confused with spindle cell carcinoma, myofibromatosis, leiomyosarcoma, primitive neuroectodermal tumors, malignant peripheral nerve sheath

tumors, and malignant fibrous histiocytoma. A panel of antibodies in an immunohistochemistry assay must be assessed together with other observations. Several investigations showed that the epithelial component of biphasic synovial sarcomas stains for cytokeratin in almost 90% of cases and that EMA is also frequently expressed. The spindle cells in some, but not all, tumors express cytokeratin or EMA focally and less intensely than epithelial cells. In monophasic synovial sarcoma, the expression of these epithelial markers is less evident, necessitating the examination of many sections from different sites. Reactivity for vimentin is observed in epithelial elements in about 15-30% of biphasic tumors; in spindle cells, it is observed in about 80-90% of both biphasic synovial sarcomas and monophasic synovial sarcomas and a wide variety of epithelial neoplasms express vimentin, making its reactivity less significant in the diagnosis of synovial sarcoma. The S-100 protein may be detectable in 30% of these tumors, causing confusion with malignant peripheral nerve-sheath tumors. In addition, bcl-2 protein was reported in 75-100% of Sinovial sarcomas, typically in a strong and diffuse fashion, especially in spindle cells [Fletcher et al, 2002, Kempson et al, 2001]. CD99 can be detected in 60% to 70% of cases. Nevertheless, these findings are of little diagnostic value because bcl-2 and CD99 are also expressed in a variety of distinct neoplasms. Chromosomal studies showed a balanced reciprocal translocation t (X;18)(p11.2;q11.2) in more than 90% of all synovial sarcoma subtypes in all anatomic sites, including the oral cavity. The translocation results in the fusion of SYT or SSXT from chromosome 18 to either SSX1, SSX2, or SSX4 genes from the X-chromosome [Almeida – Lawall M et al., 2009, Kempson RL et al, 2001].

9. Malignant Peripheral Nerve Sheath Tumors of head and neck

Malignant Peripheral Nerve Sheath Tumors (MPNSTs) are rare neoplasms with an estimated incidence of 0.1 per 100,000 per year in the general population. They account for approximately 5-10% of all soft tissue sarcomas and have a strong association with neurofibromatosis type 1 (NF-1), also known as von Recklinghausen's neurofibromatosis [Hajdu SI, 1993]. MPNSTs have been defined as any malignant tumor that arises from or differentiates toward cells of the peripheral nerve sheath, with the exception of tumors that originate from the epineurium or the peripheral nerve vasculature [Wong WW et al, 1998]. Various misleading synonyms, including the terms neurofibrosarcoma, neurogenic sarcoma, malignant neurilemmoma and malignant schwannoma, have previously all been applied to this neoplasm and are merely a reflection of the controversial clinicopathologic classification of this rare tumor [Wanebo JE, 1993]. MPNSTs of the head and neck, in particular, represent exceptional neoplasms; MPNSTs are one of the most aggressive malignant tumors and have the highest local recurrence rate of any sarcoma, and they have a marked propensity for dissemination and metastatic spread [Stark AM. 2001]. Despite multimodal therapy, including radical surgical resection and adjuvant radiochemotherapy, the prognosis of MPNSTs is said to remain dismal, particularly in the head and neck. However, prognostic factors and treatment modalities have not been consistently identified [Anghileri M et al, 2006]. Up to 30-50% of all MPNSTs are found in association with NF-1, with a reported incidence of MPNSTs in this subgroup ranging from 2-29% [al - Otieschan AA et al, 1998] MPNSTs can affect all age groups but usually present in adult life between 20 and 50 years of age, with no established predilection for sex or race. However, the mean age of patients with NF-1-associated MPNSTs is approximately a decade younger [Nagayama I et al, 1993]. The most common sites of involvement are the extremities, trunk, chest, and

retroperitoneum. While benign peripheral nerve sheath tumors, such as benign schwannoma and neurofibroma, have a propensity for the head and neck, fewer than 10% of MPNSTs affect this anatomic region. The majority of MPNSTs arise either de novo or from pre-existing neurofibromas, with an estimated incidence of malignant transformation ranging 3-30%. Only very rare examples of MPNSTs arise in the schwannoma, ganglioneuroma or phaeochromocytoma, and they arise from all cranial nerves except the optic and olfactory nerves, which have no nerve sheath [Shingh B et al, 2001]. CT and MRI scans delineate the extent of the disease and the involvement of vital structures and allow staging. MPNSTs infiltrate local tissues extensively and spread preferentially, as with other sarcomas, via the bloodstream to the liver, lungs and bone rather than the lymphatic system.

 Regional lymph node involvement occurs in less than 1% of deep-seated disease [Ducatman BS et al, 2006] and is even rarer in the superficial form, but no studies have described the statistics. Punjabi described an MPNST in the left parotid that metastased to the contralateral parotid and auditory canal with intra-cranial extension, but it is unclear whether the right parotid was the site of the metastasis or was secondarily invaded. Histological diagnosis has become more stringent because of similarities with other spindle cell malignancies, e.g., leiomyosarcomas, malignant fibrous histiocytomas and neurotropic malignant melanoma. In the past, it was necessary to demonstrate the origin from a nerve; however, cutaneous nerves are generally too small to be grossly identified, and hence, electron microscopy and immunochemistry are used to show ultrastructural evidence of Schwann cell differentiation with the absence of premelanosomes and absence of epidermal melanocyte proliferation. There are no specific immunohistological markers, and therefore, to make a diagnosis, a panel of antibodies and immunoreactivity with at least two of the following antibodies is required: S100, Leu7, myelin basic protein, glial fibrillary acid protein and PGP 9.5. [Wick MR et al, 1990]. Surgery is the main treatment for MPNSTs, which require radical resection and additional frozen sectioning of the proximal nerve to ensure clear margins, although reviews do not suggest a margin of excision. Because lymph node involvement is very unusual, elective neck dissection is not recommended. Adjuvant high-dose radiotherapy is used, particularly after incomplete resection or where radical excision is impossible, and chemotherapy remains controversial. The superficial form of the disease after primary surgery has a local recurrence rate of 78%, metastatic spread rate of 22% and a 4-year survival rate of 66%. Unfortunately, Dabski's series was small, and only half the lesions were located in the head and neck region [Dabski C et al, 1990]. The five-year survival of all patients with MPNSTs of the head and neck ranges from 15 to 34%, with 50% of the cases developing local recurrences and 33% metastases, particularly to the lung [Punjabi et al, 1996, Hujala K et al, 1993]. MPNSTs have been described in English in the literature, and most of these cases presented with a complex karyotype with triploid or tetraploid clones. The most frequent structural aberrations found in MPNST involve chromosomes 7, 9, 13, and 17, whereas the most frequent numerical chromosomal abnormalities include trisomy 7 and 2 and monosomy X and 17. Breakpoints at 12q24, as a part of a complex karyotype, were previously described in six cases of MPNSTs and in two cases of Xq22. Breakpoints at 2q35 or 4q31 have not been described for MPNSTs. Moreover, of the previously described cases of MPNSTs, only eight displayed a diploid karyotype, and six displayed a simple karyotype with unbalanced chromosomal aberrations 47,XY, 7/45,X,

Y; the literature also revealed another gene, PTPN11, which encodes the non-receptor protein tyrosine phosphatase SHP-2. This gene was mapped to the chromosome 5 region on 12q24.1, the same region involved in the second translocation of our patient. Noonan syndrome is an autosomal dominant disorder characterized by dysmorphic body features, heart disease, mental retardation, and bleeding diatheses. Interestingly, a child with Noonan syndrome and malignant schwannoma of the left forearm has been reported. These findings initially raised our suspicion, and we have further tested the possibility that the patient carried concealed characteristics of Noonan syndrome [Rao UN et al, 1996, Tartaglia M et al, 2001, Noonan JA et al, 1968, Kaplan et al, 1968].

9.1 Malignant Triton Tumor (MTT) of head and neck

MTT was first described by Masson in 1932. It is a rare subtype of MPNSTs in which the malignant schwannoma has rhabdomyoblastic differentiation, a so-called mosaic tumor with both a muscular and a neurogene component. One third of these tumors arise in the head and neck region, and at least one third of these are associated with neurofibromatosis type 1. In sporadic cases, the mean age at debut is 38 years, while cases associated with neurofibromatosis are, on average, 12 years younger [Kim ST et al, 2001, Barnes L. 2004]. The MTT is named after the Triton salamander, which is capable of regenerating limbs consisting of both muscle and nerve tissue after the cut end of the sciatic nerve is implanted into the soft tissue of its back. The MTT pathogenesis is not known. One hypothesis is that malignant schwan cells differentiate into rhabdomyoblasts. Another hypothesis is that both cell lines arise from less differentiated neural crest cells with both ectodermal and mesodermal potential. Although MTT can occur as a sporadic tumor, approximately half to two thirds of cases occur in association with neurofibromatosis type 1 (NF1), usually affecting individuals in their third or fourth decade of life. MTT is generally considered a high-grade malignant neoplasm with a poor outcome [Yakulis et al, 1996]. Previous studies [Daimaru Y et al, 1984, Victoria L et al, 1999], reviewed the treatment and outcome of 27 MTTs arising in the head and neck and commented that there may be a subset of MTTs that occurr in this region as low-grade malignancies with favorable long-term prognosis. The primary involvement of the oral cavity is extremely rare. MTTs occur predominantly in the trunk, head and neck, and lower extremities [Enzinger FM, 2001]. Among all reported cases, approximately one third appear to arise in the head and neck region. In a recent detailed review of the literature, Victoria et al [Victoria L et al, 1999] summarized the anatomic distribution of head and neck cases. The data showed that 12 of the 27 cases described were found to arise in the structures of the neck or upper thorax.

Taken together with a recent report describing MTT of the maxilla, it can be concluded that intraoral presentation of MTT is rare. The hallmark of this tumor is the presence of rhabdomyoblasts scattered throughout the stroma indistinguishably from ordinary MPNST. The number of rhabdomyoblasts varies greatly from tumor to tumor and even from area to area in the same tumor. Chromosome analysis of MTTs showed a complex hyperdiploid karyotype with multiple unbalanced translocations, large markers, and ring formations. Although some of the markers were highly variable, other markers were reasonably stable and were seen in the majority of the abnormal metaphases. Haddadin et al [Haddadin et al, 2003], compared the chromosomal breakpoints in MTT, MPNST, and rhabdomyosarcoma to identify common regions of involvement. These included 7p22, 7q36, 11p15, 12p13, 13p11.2, 17q11.2, and 19q13.1.
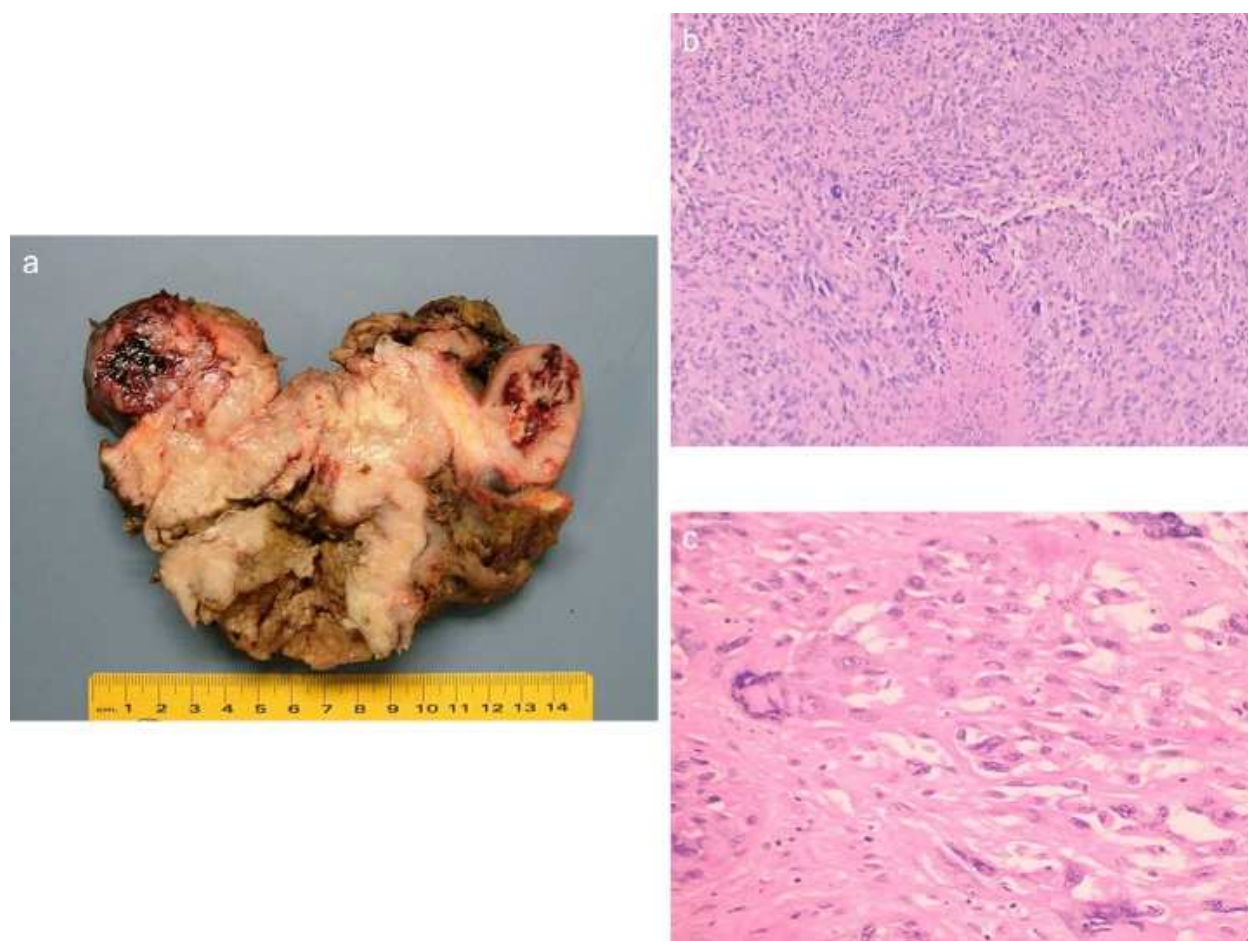
Fig. 4. a) Malignant Triton Tumor high-grade located in the neck with extensive areas of necrosis and hemorrhage, b) Malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation 40X, c) Rounded or enlongated rhabdomyoblasts throughout the tumor, 40X.

10. Conclusions

Head and neck soft tissue sarcomas are rare and represent 1-5% of all corporal neoplasias. The cause of the majority of sarcomas is yet unknown; however, it is thought that a number of environmental and genetic factors are closely linked to the development of these types of neoplasias. The survival range depends on the histological grade and the clinical stage. At five years, survival is approximately 60–70%, and with local control, it becomes 60–80%. The survival range depends on the histological grade and the clinical stage. Approximately 10–30% of patients presented with distant metastases within the first two years. In general, young patients with low-grade, small and superficial sarcomas have a better prognosis than high-grade sarcomas. Patients with sarcomas greater than 5 cm in clinical stage III or IV with positive surgical margins, can die from progression and metastases. To determine how a neoplasm is likely to behave, with or without treatment, it is necessary to know certain facts about the disease. The management of soft tissue sarcomas of the head and neck is particularly challenging and depends upon some prognostic factors. The assessment of prognostic factors, which correlate baseline clinical

and experimental covariables to outcomes, is one of the major objectives of clinical research.

11. References

Abali H, Aksoy S, Sungur A, Yalcin S (2003) Laryngeal involvement of rhabdomyosarcoma in an adult. World J Surg Oncol; 1:17.

Abdullah BH, Yahya HI, Talabani NA, Alash NI, Mirza KB. (2000) Gingival and cutaneous angiosarcoma. J Oral Pathol Med; 29 410 - 2.

Abeloff (2008) Abeloff: Clinical Oncology, 4th ed. Copyright Churchill Livingstone.

Akyol MU, Sozeri B, Kucukali T, Ogretmenoglu O. (1998) Laryngeal pleomorphic rhabdomyosarcoma. Eur Arch Otorhinolaryngol; 255:307–10.

al-Otieschan AA, Saleem M, Manohar MB, Larson S, Atallah A: (1998) Malignant schwannoma of the parapharyngeal space. J Laryngol Otol 112: 883-87.

Almeida – Lawall M, Mosqueda – Taylor A, Bologna – Molina R, Dominguez – Malagon H, Cano – Valdez AM, Luna – Ortiz K, Werneck da Cumba I (2009). Synovial sarcoma of the tongue: case report and review of the literature J Oral Maxillofac Surg; 67:914-920.

Anavi Y, Herman GE, Graybill S. (1989) Malignant fibrous histiocytoma of the mandible. Oral Surg Oral Med Oral Pathol; 68:436–43.

Anghileri M, Miceli R, Fiore M, Mariani L, Ferrari A, Mussi C, Lozza L, Collini P, Olmi P, Casali PG, Pilotti S, Gronchi A (2006). Malignant peripheral nerve sheath tumours: prognostic factors and survival in a series of patients treated at a single institution. Cancer 07: 1065 – 74.

Antonescu CR, Rosenblum MK, Pereira P, et al. (2001) Sclerosing epithelioid fibrosarcoma: a study of 16 cases and confirmation of a clinicopathologically distinct tumor. Am J Surg Pathol; 25:699–709.

Barnes L, Kanbour A. (1988) Malignant fibrous histiocytoma of the head and neck A report of 12 cases. Arch Otolaryngol Head Neck Surg; 114:1149–56.

Barnes L. (2001) Surgical pathology of the head and neck, 2nd ed., vol. 2, Marcel Dekker, Springer Link.

Barrera FJL, López GCM, Gómez GE, Meneses GA (1997). Sarcomas de cabeza y cuello en adultos. Rev Inst Nal Cancerol Méx; 43: 184-88.

Bass B, Archard H, Sussman R, Stern M, Saunders V. (1986) Case 62: expansile radiolucent lesion of the mandible. J Oral Maxillofacial Surg: 44: 799–803.

Bentz BG, Singh B, Woodruff J, Brennan M, Shah JP, Kraus D (2004). Head and neck soft tissue sarcomas: a multivariate analysis of outcomes. Ann Surg Oncol; 11: 619-28.

Bras J., Batsakis J.G., Luna M.A., (1987) Rhabdomyosarcoma of the oral soft tissues. Oral Surg Oral Med Oral Pathol; 64 585–96.

Carlos R, Altini M, Takeda Y (2005). Odontogenic Sarcomas, pathology and genetics of tumours of the head and neck. In: Barnes L, Eveson JW, Recihart P, Sidransky D, editors. WHO Classification of tumours. Pathology and genetics. Tumours of head and neck. Lyon: IARC Press.

Chan YW, Guo YC, Tsai TL, Tsay SH, Lin CZ. (2004) Malignant fibrous histiocytoma of the maxillary sinus presenting as toothache. J Chin Med Assoc; 67:104–7.

Chen JM, Novick WH, Logan CA. (1991) Leiomyosarcoma of the larynx. J Otolaryngol;20:345–8.

Chen Y, Wang JM, Li JT. (2007). Ameloblastic fibroma: A review of published studies with special reference to its nature and biological behavior. Oral Oncology; 43:960-9.

Cocks H, Quraishi M, Morgan D, Bradley P. (1999)   Leiomyosarcoma of the larynx. Otolaryngol Head Neck Surg; 121:643–6.

Colville RJ, Charlton F, Kelly CG, Nicoll JJ, McLean NR (2005). Multidisciplinary management of head and neck sarcomas. Head Neck; 27: 814-24.

Conley J, Stout A, Healey W.(1967) Clinicopathologic analysis of eighty four patients with an original diagnosis of fibrosarcoma of the head and neck. Am J Surg 114: 564–569.

Dabski C, Reiman Jr HM, Muller SA (1990). Neurofibrosarcoma of skin and subcutaneous tissues. Mayo Clin Proc; 65: 164 - 72.

Daimaru Y, Hashimoto H, Enjoji M. (1984) Malignant "triton" tumors: a clinicopathologic and immunohistochemical study of nine cases. Hum Pathol;15:768-78.

de Bree R, van der Valk P, Kuik DJ, van Dies PJ, Doornaert P, Buter J, Eerenstein SE, Langendijk JA, van der Waal I, Leemans CR (2006). Prognostic factors in adult soft tissue sarcomas of the head and neck: A single-centre experience. Oral Oncology; 42: 703-9.

de Silva M.V., McMahon A.D., Reid R., (2004) Prognostic factors associated with local recurrence, metastases, and tumorrelated death in patients with synovial sarcoma, Am. J. Clin. Oncol; 27:113 -121.

Derbel F, Hajji H, Mitmet A, Hadj -Hamida MB, Mazhoud J, Youssef S, Ben – Ali A, Khochtali H, Chaouch A, Mokni M, Jemni H, Hadj-Hamida BR (2010). Liver metastasis of malignant fibrous histiocytoma: A case report. Arab J Gastroenterol; 11:113-15.

Devita TJ Jr, Hellman S, Rosenberg SA. (2001) Cancer principles and practice of oncology. 7th ed. New York: Lippincott Williams and Wilkins.

Donner LR, Clawson K, Dobin SM. (2000) Sclerosing epithelioid fibrosarcoma: a cytogenetic, immunohistochemical, and ultrastructural study of an unusual histological variant. Cancer Genet Cytogenet; 119:127–31.

Donner LR, Clawson K, Dobin SM. (2000) Sclerosing epithelioid fibrosarcoma: a cytogenetic, immunohistochemical, and ultrastructural study of an unusual histological variant. Cancer Genet Cytogenet; 119:127–31.

Dry SM, Jorgensen JL, Fletcher CDM. (2000) Leiomyosarcomas of the oral cavity: an unusual topographic subset easily mistaken for nonmesenchymal tumours. Histopathology: 36: 210–220.

Dry SM, Jorgensen JL, Fletcher CDM. (2000) Leiomyosarcomas of the oral cavity: an unusual topographic subset easily mistaken for nonmesenchymal tumours. Histopathology; 36: 210–220.

Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM. (2006) Malignant peripheral nerve sheath tumours: a clinicopathologic study of 120 cases. Cancer; 1986:57.

Eeles RA, Ficher C, A'Hern RP, Robinson M, Rhys-Evans P, Henk JM, et al. Head and neck sarcomas: prognostic factors and implications for treatment. Br J Cancer 1993; 68:201–7.

Enjoji M, HashimotoH, TsuneyoshiM, Iwasaki H. (1980) Malignant fibrous histiocytoma. A clinico pathologic study of 130 cases. Acta Pathol Jpn; 30:727–41.

Enzinger FM, Weiss SW. Liposarcoma. In: Weiss SW, Goldblum JR, editors. (2001) Soft tissue tumors. 4th ed. St. Louis: Mosby.

Enzinger FM, Weiss SW. Soft tissue tumors. Foruth edition. St Louis: Mosby, 2001.

Farman AG, Kay S. (1977) Oral leiomyosarcoma: report of a case and review of the literature pertaining to smooth muscle tumours of the oral cavity. Oral Surg; 43: 402–409.

Favia G, Lomuzio L, Serpico R, Mariorano E. (2002) Angiosarcoma of the head and neck with intraoral presentation. A clinicopathologic study of four cases. Oral Oncol; 38:757 - 62.

Ferlito A, Rinaldo A, Marioni G. (1999) Laryngeal malignant neoplasms in children and adolescents. Int J Pediatr Otorhinolaryngol; 49:1–14.

Ferrari A, Gronchi A., Casanova M., Meazza C., Gandola L., P. Collini, et al (2004). Synovial sarcoma: a retrospective analysis of 271 patients of all ages treated at a single institution, Cancer; 101:627 - 634.

Fletcher CDM, Unni KK, Mertens F: (2002)World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Soft Tissues and Bone. Lyon, IARC Press.

Forton - Glen EJ, Van Parys G, Hertveldt K. (2005) Primary angiosarcoma of the non-irradiated parotid gland: a most uncommon, highly malignant tumor. Eur Arch Oto Rhino Laryngology; 262:173 - 7.

Franca CM, Caran EM, Alves MTS (2006) Rhabdomyosarcoma of the oral tissues two new cases and literature review. Medicina Oral, Patologia Oral y Cirugia Bucal; 11:E136 – 40.

Freedman AM, Reiman HM, Woods JE. (1989) Soft-tissue sarcomas of the head and neck. Am J Surg;158:367–72.

Freedman AM, Reiman HM, Woods JE. Soft tissue sarcomas of the head and neck. Am J Surg 1989;158:367–72.

Freije JE, Gluckman JL, Biddinger PW,Wiot G. (1992) Muscle tumors in the parapharyngeal space. Head Neck;14:49–54.

Gibbs JF, Huang PP, Lee RJ, McGrath B, Brooks J, McKinley B et al. (2001). Malignant fibrous histiocytoma: an institutional review. Cancer Invest; 19:23–7.

Golledge J, Fisher C, Rhys-Evans PH. Head and neck liposarcoma. Cancer 1995; 76:1051-8.

Grayson W, Nayler SJ, Jena GP.(1998) Synovial sarcoma of the parotid gland. A case report with clinicopathological analysis and review of the literature. S Afr J Surg; 36:32-5.

Greene FL, Page DL, Flemming FD (2002). American Joint Committee on cancer: cancer staging manual. 6th ed. New York (NY); Springer: 221–6.

Guillou, J. Benhattar, F. Bonichon, G. Gallagher, P. Terrier, E. Stauffer, et al (2004). Histologic grade, but not SYT-SSX fusion type, is an important prognostic factor in patients with synovial sarcoma: a multicenter, retrospective analysis, J. Clin. Oncol; 22:4040 – 50.

Haddadin MH, Hawkins AL, Morsberger LA, Depew D, Epstein JI, Griffin CA (2003). Cyotgenetic study of malignant triton tumor: case report. Cancer Genet Cytogenet; 144:100-5.

Haerr RW, Turalba CI, el-Mahdi AM, Brown KL. (1987) Alveolar rhabdomyosarcoma of the larynx: case report and literature review. Laryngoscope; 97:339–44.

Hajdu SI (1993). Peripheral nerve sheath tumours. Histogenesis, classification, and prognosis. Cancer 72: 3549 – 52.

Hensley ML, Maki R, Venkatraman E, Geeler G, Lovegren M, Aghajanian C, et al. (2002) Gemcitabine and docetaxel in patients with unresectable leiomyosarcoma: results of a phase II trial. J Clin Oncol;20(12):2824–31.

Hicks J, Dilley A, Patel D, Barrish J, Zhu SH, Brandt M (2001). Lipoblastoma and lipoblastomatosis in infancy and childhood: histopathologic, ultrastructural, and cytogenetic features. Ultrastruct Pathol; 25:321–33.

Hoffmann DF, Everts EC, Smith JD, Kyriakopoulos DD, Kessler S (1988). Malignant nerve sheath tumours of the head and neck. Otolaryngol Head Neck Surg 99: 309-314.

Hujala K, Martikainen P, Minn H, Grenman R (1993). Malignant nerve sheath tumours of the head and neck: four case studies and review of the literature. Eur Arch Otorhinolaryngol; 250:379 - 82.

Hunt SJ, Santa Cruz DJ. (2004) Vascular tumors of the skin: a selective review. Semin Diagn Pathol; 21: 166–218.

Huvos AG, Heilweil M, Bretsky SS. (1985) The pathology of malignant fibrous histiocytoma of bone. A study of 130 patients. Am J Surg Pathol; 9:853–71.

Iguchi Y, Takahashi H, Yao K, Nakayama M, Nagai H, Okamoto M. (2002) Malignant fibrous histiocytoma of the nasal cavity and paranasal sinuses: review of the last 30 years. Acta Oto Laryngol; 122:75–8.

Izumi K, Maeda T, Cheng J, Saku T. (1995) Primary leiomyosarcoma of the maxilla with regional lymph node metastasis. Report of a case and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod; 80:310 - 9.

Jernstrom P: (1954) Synovial sarcoma of the pharynx: Report of a case. Am J Clin Pathol; 24:957.

Kanazawa H, Watanabe T, Kasamatsu A. (2003) Primary malignant fibrous histiocytoma of the mandible: review of literature and report of a case. J Oral Maxillofac Surg; 61:1224–7.

Kaplan MS, Opitz JM, Gosset FR (1968). Noonan's syndrome. A case with elevated serum alkaline phosphatase levels and malignant schwannoma of the left forearm. Am J Dis Child; 116:359–66.

Kaplan MS, Opitz JM, Gosset FR (1968). Noonan's syndrome. A case with elevated serum alkaline phosphatase levels and malignant schwannoma of the left forearm. Am J Dis Child; 116:359–66.

Kearney MM, Soule EH, Ivins JC. (1980) Malignant fibrous histiocytoma: a retrospective study of 167 cases. Cancer; 45:167–78.

Kempson RL, Fletcher CDM, Evans HL, Hendrickson MR, Sibley RK: (2001) Atlas of Tumor Pathology. Tumors of the Soft Tissues. Bethesda, Armed Forces Institute of Pathology.

Khong JJ, Chen CS, James CL, Huilgol SC, O'Donnell BA, Sullivan TJ, et al. (2005) Malignant fibrous histiocytoma of the eyelid: differential diagnosis and management. Ophthal Plast Reconstr Surg; 21: 103–8.

Kim ST, Kim CW, Han GC, Park C, Jang IH, Cha HE, et al (2001). Malignant Triton Tumor of the nasal cavity. Head Neck;23:1075–8.

Koch M, Nielsen GP, Yoon SS. (2008) Malignant tumors of blood vessels: angiosarcomas, hemangioendotheliomas, and hemangioperictyomas. J Surg Oncol; 97: 321–329.

Kratochvil FJ, MacGregor SD, Budnick SD, Hewan-Lowe K, Allsup HW. (1982) Leiomyosarcoma of the maxilla. Report of a case and review of the literature. Oral Surg Oral Med Oral Pathol; 54:647-55.

Lanigan DT, Hey JH, Lee L. (1989) Angiosarcoma of the maxilla and maxillary sinus: report of a case and review of the literature. J Oral Maxillofac Surg; 47:747–53.

Lazos OM, Ávila TA, Hernández GM. Sarcomas de cabeza y cuello (1999). Estudio clinicopatológico de 29 casos. Rev Med Hosp Gen Mex; 62: 176-82.

Le QT, Fu KK, Kroll S, Fitts L, Massullo V, Ferrell L, Kaplan MJ, Phillips TL (1997). Prognostic Factors in adult soft tissue sarcomas of the head and neck. Int J Radiat Oncol Biol Phys; 37: 975-84.

Limon J, Dalcin P, Sandberg AA. (1986) Cytogenetic findings in a primary leiomyosarcoma of the prostate. Cancer Genet Cytogenet;22:159–67.

Link TM, Haeussler MD, Poppek S, Woertler K, Blasius S, Lindner N ,et al. (1998) Malignant fibrous histiocytoma of bone: conventional X-ray and MR imaging features. Skeletal Radiol; 27:552–8.

Little DJ, Ballo MT, Zagars GK, Pisters PWT, Patel SR, El-Naggar AK, Garden AS, Benjamin RS. (2002) Adult rhabdomyosarcoma. Outcome following multimodality treatment. Cancer; 2:377–88.

Loudon JA, Billy ML, DeYoung BR, Allen CM. (2000) Angiosarcoma of the mandible: a case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod; 89:471–6.

Magrini E, Pragliola A, Fantasia D, Calabrese G, Gaiba A, Farnedi A, et al (2003). Acquisition of i(8q) as an early event in Malignant Triton Tumors. Cancer Genet Cytogenet;154:150–5. Cancer Genetics and Cytogenetics 144 100 105.

Mahdhaoui A, Bouraoui H, Chenivor M, Trimech B, Mesghani M. (2004) Right atrium angiosarcoma disclosed by alveolar hemorrhage. Rev Med Sci; 12:115 - 6.

Marioni G, Bertino G, Mariuzzi L, Bergamin-Bracale AM, Lombardo M, Beltrami CA. (2000) Laryngeal leiomyosarcoma. J Laryngol Otol ;114:398–401.

Mark RJ, Poen JC, Tran LM, Fu YS, Juillard GF. (1996) Angiosarcoma. A report of 67 patients and a review of the literature. Cancer; 77: 2400 - 6.

Mark RJ, Poen JC, Tran LM, Fu YS, Juillard GF. (1996) Angiosarcoma. A report of 67 patients and a review of the literature. Cancer; 77: 2400 - 6.

Marotta D, Angeloni M, Salgarello M, Ricciardella ML, Chalidis B, Maccauro G. (2009) Surgical treatment of a giant tibial high-grade mixofibrosarcoma with preservation of limb function: a case report. Int Semin Surg Oncol; 17: 16.

McCulloch TM, Makielski KH, McNutt MA (1992). Head and neck liposarcoma. A histopathologic reevaluation of reported cases. Arch Otolaryngol Head Neck Surg;118:1045–9.

McElderry J, McKenney JK, Stack BC. (2008), High – grade liposarcoma metastasic to the gingival mucosa: case report and literature review. Am J Otol; 29: 130-34.

Meis-Kindblom JM, Kindblom LG, Enzinger FM. (1995) Sclerosing epithelioid fibrosarcoma. A variant of fibrosarcoma simulating carcinoma. Am J Surg Pathol; 19:979–93.

Mendenhall WM, Mendenhall CM, Werning JW, Riggs CE, Mendenhall NP (2005). Adult head and neck soft tissue sarcomas. Head Neck; 27: 916-22.

Mentzel T, Kutzner H, Wollina U. (1998) Cutaneous angiosarcoma of the face: clinicopathologic and immunohistochemical study of a case resembling rosacea clinically. J Am Acad Dermatol; 38:837 - 40.

Mentzel T, van den Berg E, Molenaar WM. (2002) Myxofibrosarcoma, pathology and genetics of tumors of soft tissue and bone. In: Fletcher C, Unni KK, Mertens F, editors. WHO Classification of tumours. Pathology and genetics. Tumours of soft tissue and bone. Lyon: IARC Press.

Miyajima K, Oda Y, Oshiro Y, Tamiya S, Kinukawa N,Masuda K, Tsuneyoshi M. (2002) Clinicopathological prognostic factors in soft tissue leiomyosarcoma: a multivariate analysis. Histopathology; 40: 353–359.

Montgomery E, Goldblum JR, Fisher C. (2002) Leiomyosarcoma of the head and neck: a clinicopathological study. Histopathology; 40: 518–525.

Morgan MB, Swann M, Somach S, Eng W, Smoller B. (2004) Cutaneous angiosarcoma: a case series with prognostic correlation. J Am Acad Dermatol; 50: 864–867.

Mullick SS, Mody DR, Schwarz MR. (1997) Angiosarcoma at unusual sites, a report of two cases with aspiration cytology and diagnostic pitfalls. Acta Cytol; 41:839 - 44.

Muzio L, Favia G, Mignogna MD, Piattelli A, Maiorano E. (2000) Primary intraoral leiomyosarcoma of the tongue: an immunohistochemical study and review of the literature. Oral Oncology; 36 519-524.

Nagayama I, Nishimura T, Furukawa M (1993) Malignant schwannoma arising in a paranasal sinus. J Laryngol Otol 107: 146 – 148.

Narvaez JA, Muntane A, Narvaez J, Martin F, Monfort JL, Pons LC. (1996) Malignant fibrous histiocytoma of the mandible. Skeletal Radiol; 25:96–9.

Nikitakis NG, Lopes MA, Bailey JS, Blanchaert RH, Ord RA, Saul JJ. (2002) Oral leiomyosarcoma: review of the literature and report of two cases with assessment of the prognostic and diagnostic significance of immunohistochemical and molecular markers. Oral Oncol: 38: 201–208.

Noonan JA (1968) Hypertelorism with Turner phenotype. A new syndrome with associated congenital heart disease. Am J Dis Child; 116:373–80.

Ogawa A, Kawashima K. (2005) Rigid reconstruction of the cheek by free skull bone grafting: a case report. J Jpn Soc Plast Reconstr Surg; 25:454–8.

Oliver AJ, Gibbons SD, Radden BG, Busmanis I, Cook RM. (1991) Primary angiosarcoma of the oral cavity. Br J Oral Maxillofac Surg; 29:38–41.

Pandey M, Chandramohan K, Thomas G, Mathew A, Sebastian P, Somanathan T, Abrham EK, Rajan B, Krishnan Nair M (2003). Soft tissue sarcoma of the head and neck in adults. Int J Oral Maxillofac Surg; 32: 43-8.

Park SW, Kim HJ, Lee JH, Ko YH. (2009) Malignant fibrous histiocytoma of the head and neck: CT and MR imaging findings. Am J Neuroradiol 2009; 30:71–6.

Park SW, Kim HJ, Lee JH, Ko YH. (2009) Malignant fibrous histiocytoma of the head and neck: CT and MR imaging findings. Am J Neuroradiol 2009; 30:71–6.

Patel SG, Shaha AR, Shah JP (2001). Soft tissue sarcomas of the head and neck: an update. Am J Otolaryngol; 22: 2–18.

Pavithran K, Doval DC, Mukherjee G, Kannan V, Kumaraswamy SV, Bapsy PP (1997). Rhabdomyosarcoma of the oral cavity, Acta Oncol; 36: 819–821.

Pellitteri PK, Ferlito A, Bradley PJ, Shaha AR, Rinaldo A (2003). Management of sarcomas of the head and neck in adults. Oral Oncol; 39: 2–12.

Penel N, Depadt G, Vilain M-O, Vanseymortier L, et al. (2003) Fre´quence des ge´nopathies et des ante´ce´dents carcinologiques chez 493 adultes atteints de sarcomes visce´raux ou des tissues mous. Bull Cancer;90(10):887–95.

Pereira CM Jr, Jorge J Jr, Hipolito OD, Kowalski LP, Lopes MA.(2005) Primary intraosseous fibrosarcoma of jaw. Int J Oral Maxillofac Surg; 34:579-81.

Pereira CM, Jorge J, Di Hipolito O, Kowalski LP, Lopes MA. (2005) Primary intraosseous fibrosarcoma of jaw. Int J Oral Maxillofac Surg; 34:579–81.

Pezzi CM, Rawlings Jr MS, Esgro JJ, Pollock RE, Romsdahl MM. (1992) Prognostic factors in 277 patients with malignant fibrous histiocytoma. Cancer; 69:2098–103.

Punjabi AP, Haug RH, Chung-Park Moon JA, Likavek M. J (1996). Oral Maxillofac Surg; 54:765 - 9.

Rangheard AS, Vanel D, Viala J, Schwaab G, Casiraghi O, Sigal R. (2001)Synovial sarcomas of the head and neck: CT and MR imaging findings of eight patients. AJNR Am J Neuroradiol; 22:851-7.

Rao UN, Surti U, Hoffner L, Yaw K (1996) Cytogenetic and histologic correlation of peripheral nerve sheath tumors of soft tissue. Cancer Genet Cytogenet; 88:17–25.

Reichart PA, Philipsen HP, Hans P (2004). Odontogenic Tumours. Quintessence Publishing Co Ltd, UK.

Rinaldo A, Shaha AR, Pellitteri PK, Bradley PJ, Ferlito A. (2004) Management of malignant sublingual salivary gland tumors. Oral Oncol; 40:2–5.

Sabesan T, Xuexi W, Yongfa Q, Pingzhang T, Ilankovan V. (2006) Malignant fibrous histiocytoma: outcome of tumors in the head and neck compared with those in the trunk and extremities. Br J Oral Maxillofac Surg; 44:209–12.

Sapp JP, Eversole LR, Wysocki GP, editors.(2004) Contemporary oral and maxillofacial pathology. St. Louis: Mosby.

Senel FC, Bektas D, Caylan R, Onder E, Gunhano O. (2006) Malignant fibrous histiocytoma of the mandible. Dentomaxillofac Radiol; 35:125–8.

Singh B, GogineniSK, Sacks PG, Shaha AR, Shah JP, Stoffel A, Rao PH (2001). Molecular cytogenetic characterization of head and neck squamous cell carcinoma and refinement of 3q amplification. Cancer Res; 61:4506–13.

Sivanandan R, Kong CS, Kaplan MJ, Fee Jr WE, Thu-Le Q, Goffinet DR. (2004) Laryngeal embryonal rhabdomyosarcoma. Arch tolaryngol Head and Neck surg; 130:1217–22.

Skubitz KM, Haddad PA. (2005) Paclitaxel and pegylated-liposomal doxorubicin are both active in angiosarcoma. Cancer; 104: 361–366.

Soares AB, Lins LHS, Mazedo AP, Neto JSP, Vargas PA.(2006) Fibrosarcoma originating in the mandible. Med Oral Patol Oral Cir Bucal; 11: E 243-6.

Stark AM, Buhl R, Hugo HH, Mehdorn HM (2001). Malignant peripheral nerve sheath tumours e report of 8 cases and review of the literature. Acta Neurochir (Wien) 143: 357 – 364.

Tartaglia M, Mehler EL, Goldberg R, Zampino G, Brunner HG, Kremer H, van der Burgt I, Crosby AH, Ion A, Jeffery S, Kalidas K, Patton MA, Kucherlapati RS, Gelb BD (2001). Mutations in PTPN11, encoding the protein tyrosine phosphatase SHP-2, cause Noonan syndrome. Nat Genet; 29:465–8.

Toth BB, Fleming TJ, Lomba JA, Martin JW. (1981) Angiosarcoma metastatic to the maxillary tuberosity gingiva. Oral Surg Oral Med Oral Pathol; 52:71–4.

Turc-Carel C, Pietrzak E, Kakati S, Kinniburgh AJ, Sandberg AA.(1987)The human int-1 gene is located at chromosome region 12q12-12q13 and is not rearranged in myxoid liposarcoma with t(12;16)(q13;p11). Oncogene Res;1:397–405.

Victoria L, McCulloch TM, Callaghan EJ, Bauman NM. (1999). Malignant Triton Tumor of the head and neck: a case report and review of the literature. Head Neck;21:663–70.

Wadhwan V, Chaudhary MS, Gawande M. (2010) Fibrosarcoma of the oral cavity. Indian J Dent Res; 21:295-8.

Wang CP, Chang YL, Ting LL, Yang TL, Ko JY, Lou PJ. (2009) Malignant fibrous histiocytoma of the sinonasal tract. Head Neck; 31:85–93.

Weiss SW, (2001) 4th ed: Enzinger and Weiss's soft tissue tumours 4th edn. St Louis: CV Mosby.

Weiss SW, Goldblum JR. (2001) Malignant soft tissue, in Weiss SW, Goldblum JR (eds): Enzinger and Weiss's Soft Tissue Tumors. St Louis, Mosby.

Wenig BM, Heffner DK. Liposarcomas of the larynx and hypopharynx: a clincopathologic study of eight new cases and a review of the literature. Laryngoscope 1995;105:747–56.

Wick MR (1990). Malignant peripheral nerve sheath tumours of the skin. Mayo Clin Proc; 65:279 - 82.

Wong CK, Edwards AT, Rees BI. Liposarcoma: a review of current diagnosis and management. Br J Hosp Med 1997;58: 589–91.

Wong WW, Hirose T, Scheithauer BW, Schild SE, Gunderson L (1998). Malignant peripheral nerve sheath tumour: analysis of treatment outcome. Int J Radiat Oncol Biol Phys 42: 351 – 360.

Yakulis R, Manack L, Murphy AI Jr. (1996) Postradiation malignant triton tumor: a case report and review of the literature. Arch Pathol Lab Med;120:541-8.

Zahm SH, Fraumeni JFJr (1997). The epidemiology of soft tissue sarcoma. Semin Oncol; 24: 504-14.

Soft Tissue Tumors

Edited by Prof. Fethi Derbel

Soft tissue tumors include a heterogeneous group of diagnostic entities, most of them benign in nature and behavior. Malignant entities, soft tissue sarcomas, are rare tumors that account for1% of all malignancies. These are predominantly tumors of adults, but 15% arise in children and adolescents. The wide biological diversity of soft tissue tumors, combined with their high incidence and potential morbidity and mortality represent challenges to contemporary researches, both at the level of basic and clinical science. Determining whether a soft tissue mass is benign or malignant is vital for appropriate management. This book is the result of collaboration between several authors, experts in their fields; they succeeded in translating the complexity of soft tissue tumors and the diversity in the diagnosis and management of these tumors.

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