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Localized ENT Amyloidosis – Literature Overview

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1. Introduction

Amyloidoses from a group of disorders characterized by extracellular tissue accumulation of amorphous hyaline material. They are categorized in two main forms: systemic and localized (Zhuang YL, 2005). Localized forms involve a single organ, whereas systemic amyloidosis involves multiple organ systems.

Localized forms often involve the head and neck. The aerodigestive tract is a common location, the nasopharynx or soft palate are rarely envaded (Panda NK, 2007) (Pitkäranta A, 2000).

The distinction between localized and systemic disease is important because localized amyloidosis can be managed conservatively with an excellent prognosis, whereas systemic amyloidosis is associated with significant morbidity and mortality (Kyle RA, 1975).

Although the pathogenesis is not completely understood, soluble protein subunits undergo a conformational change to become insoluble and aggregate in an antiparallel β-pleated sheet conformation (Panda NK, 2007). The diagnosis of amyloidosis is made based on Congo red staining on tissue biopsy which leads to apple-green birefringence on polarized microscopy (Patel A, 2002).

Amyloidomas are benign tumorlike lesions consisting of localized deposits of amyloid and are the rarest form in the group of amyloidosis-related lesions (Parmar H, 2010).

Amyloidosis should not be considered as a single clinical entity, but rather as a nonhomogeneous group of diseases characterized by the common presence of a fibrillar structure of linear, aggregated fibers with a cross β-pleated sheet conformation, and evidenced by x-ray diffraction. In primary amyloidosis, a monoclonal population of marrow cells produces either fragments of light chains that may be processed to form amyloid. Secondary amyloidosis is characterized by a defect in the metabolism of the precursor protein (Comenzo RL, 2006).

Our objective is to study the epidemioloclinical characteristics of ENT amyloidosis and the management of those localizations.

2. Study method

Data Sources a systematic literature search of MEDLINE, SCIENCEDIRECT and Web of Science Review Database from 2000 to 2010 was conducted using specific search terms: head and neck amyloidosis, localized amyloidosis, ENT Diseases, hypopharynx, larynx, oral cavity, oropharynx, nasopharynx and sinonasal cavities.

We considered thirty-three articles for our review and including a total of 43 patients.
Exclusion criteria were: systemic amyloidosis, a concomitant history of chronic inflammatory processes, monoclonal gammopathy and myeloma.

3. Epidemiology

Localized amyloidosis is characterized by the same staining properties as systemic amyloidosis but is usually fortuitously diagnosed.
The majority of patients with head and neck amyloidosis have no underlying chronic systemic disease (primary form).
Nasal or nasopharyngeal amyloidosis is a very rare condition with few case reports seen in the English scientific literature. Both sexes may be affected, with a great variation in age of disease onset (8–86 years old). Laryngeal involvement in amyloidosis is rare and accounts for less than 1% of all benign laryngeal tumors. It usually occurs in the 40–60 years age range with a male to female predominance of about 2:1 (Gallivan GJ, 2010).

3.1 Age and sex

The mean age of patients considered in our review was 52,36 years with ages ranging from 10 to 86 years; 21 were male and 22 were female.

3.2 Localization

This literature review identified 33 articles describing a total of 43 cases of localized head and neck amyloidosis, summarized in Table 1.

Localization	Number of cases	Mean age	Male	Female
Larynx	12	47,5	6	6
Nasopharynx	7	55,5	4	3
Tongue	6	58	4	2
Palate	4	66,7	1	3
Trachea	3	52,3	2	1
Hypopharynx	3	55,3	0	3
Sinonasal cavities	2	32,5	0	2
Oropharynx	1	12	0	1
Tonsil	1	71	1	0
Ear	1	47	1	0
Cervical lymph nodes	3	60	2	1

Table 1. Epidemiological characteristics of localized amyloidosis

Localized amyloidosis is an uncommon benign disorder which occurs most commonly in the head and neck region.
Among the sites, larynx is affected most frequently (61%), followed by oropharynx (23%), trachea (9%), and orbit (4%) (Glenner GG, 1980) (Scott PP, 1986). Only 3% of cases occur in the nasopharynx (Glenner GG, 1980) (Scott PP, 1986). In the larynx, the most commonly involved sites are the vestibular fold, followed by the subglotis, ventricle, vocal folds, and aryepiglottic folds.
Chin *et al*. reported a case of localized amyloidosis involving simultaneously the sinonasal cavities and the larynx (Chin SC, 2004).
Localized amyloidosis of the nasal mucosa is extremely rare with few cases reported in the English literature (Pearlman AN, 2010).
Localized amyloidosis of the palate is extremely rare and only five cases have been reported (Aono J, 2009).
Amyloidosis of Waldeyer's ring has been also described (Walker PA, 1996).
The hypopharyngeal involvement is extremely rare: extensive search of the literature retrieved only 3 primitive localized hypopharyngeal amyloidosis (Ghekiere O, 2003) (Bhavani RS, 2010) (Penner CR, 2006).
Amyloidosis in the external auditory canal is extremely rare. Indeed, only 13 cases have been reported worldwide to date (Yamazaki K, 2011).
Only two cases report of localized amyloidosis of the parotid glands were described in the literature (Stimson PG, 1988) (Nandapalan V, 1998).
No case of localized submandibular amyloidosis has been reported.
Kurokawa (Kurokawa H, 1998) reported the first case of primary localized of the sublingual gland.

4. Clinical features

There are currently 3 known forms of amyloidosis.
The first, primary systemic amyloidosis is a systemic condition with no known underlying cause. It is different from the secondary systemic amyloidosis, which occurs with other underlying medical problems, such as tuberculosis and rheumatoid arthritis. And this form also includes patients with multiple myeloma, of which 10% to 20% have associated amyloidosis (Kyle RA, 1975) (Gertz MA, 2005). Renal and cardiac diseases are seen in both primary and secondary systemic forms and are the most frequent causes of death (Kyle RA, 1975). Other symptoms may include hypesthesias, syncope, macroglossia, and carpal tunnel syndrome. The third form of amyloidosis is localized and which occurs without any evidence of systemic involvement or underlying disease.
In a review of 236 cases of amyloidosis, Kyle and Bayrd (Kyle RA, 1975) found that only 22 cases (9%) were localized. None of these patients developed systemic disease in a 10-year follow-up. Similarly, in the University of California, Los Angeles series, localized amyloidosis did not evolve to primary or secondary systemic amyloidosis or multiple myeloma (Kerner MM, 1995).
Localized amyloidosis is a clinical entity with variable presentation, depending on the organs involved.
Clinically, amyloidosis can manifest in several different forms: primary versus secondary, heredofamilial versus acquired, and generalized versus localized.
The most frequent types encountered in clinical practice are the amyloid light chain (AL, primary) and a protein called the AA protein (secondary), both of which might involve

multiple organ systems (kidney, heart, and liver). In our literature review we analyzed the articles about localized amyloidosis

Isolated deposition of fibrillar proteins in an organ may result in organ-specific syndromes (e.g. Alzheimer's disease and primary localized amyloidosis of the bladder) (Malek RS, 2002). In the head and neck region, the most common site of amyloid depositions is the larynx, and other sites include the orbit, skin, tongue, salivary glands and cervical lymph nodes (Pribitkin E, 2003).

The presentation of head and neck amyloidosis varies according to the site of involvement. A patient can present with nasal obstruction, epistaxis (due to vessel wall invasion), glue ear, dysphagia, foreign body sensation in the throat, and voice change, but most of the cases of oropharyngeal amyloid are asymptomatic (Mufarrij AA, 1990).

They will appear as yellowish mucosa, covered, and irregular polypoidal lesions (Mufarrij AA, 1990).

4.1 Sino-nasal and nasopharyngeal amyloidosis

The presenting symptoms consist of nasal obstruction, recurrent epistaxis, postnasal drip and ear problems due to eustachian tube obstruction with resultant middle ear effusion (Pang KP, 2001) (Patel A, 2002).

Facial pain can be seen in amyloidosis of the maxillary antrum (MCALPINE JC, 1964).

Occasionally, it may manifest as a clinical picture simulating an apparent malignancy with cervical nodal metastasis (Zhuang YL, 2005).

Generally, the appearance is described as a yellowish or whitish polypoid, firm mass.

Macroscopically, organs infiltrated with amyloid have a characteristic firm, rubbery firm consistency, but may also be nodular-like or irregular, mimicking neoplasm. A waxy, gray or yellowish appearance is also typical (Pribitkin E, 2003).

Geller *et al* (Geller E, 2010) reported the first case of localized nasopharyngeal amyloidosis causing bilateral nasolacrimal duct obstruction.

This localization has usually a slow evolution.

4.2 Larynx

Laryngeal amyloidosis remains a rare entity accounting for about 1% of all benign laryngeal tumors. Within the larynx, a number of sites can be involved including ventricular folds (55%), laryngeal ventricle (36%), subglottic space (36%), vocal folds (27%), aryepiglottic folds (23%) and anterior commissure (14%) (Passerotti GH, 2008).

Clinical manifestations are hoarseness, dyspnea, cough, stridor, and rarely, hemoptysis. (Finn DG, 1982)

The lesion is usually a firm, nonulcerated yellow, orange, or gray submucosal nodule, mass or pedunculated polypoid lesion.

It is rare for vocal fold fixation or cicatricial stenosis to occur unless other predisposing factors are also present (Caldarelli DD, 1979). A cystic lesion (Talbot AR, 1979) or infiltrating tumor of the vocal folds and subglottis, multinodular subglottic, tracheal, mainstem bronchial and pulmonary deposits may occur (Simpson GT 2nd, 1984).

4.3 Tracheobronchial

Respiratory symptoms may vary according to the anatomic deposition of amyloid (Gillmore JD, 1999). Patients with upper tracheal or proximal airway disease develop varying degrees of airway obstruction. They have worse prognosis than those with distal airway involvement (Utz JP, 1996).

Common presenting symptoms are dyspnea, cough, and wheezing. These symptoms are not specific and may mimic different pathologies, such as asthma, chronic bronchitis, and endobronchial tumor. Those with distal tracheal and bronchial disease may have atelectasis, recurrent pneumonias, or bronchiectasis (Thompson PJ, 1983).

4.4 Hypopharynx and upper esophagus

The presenting symptoms consist of progressive dysphagia to both solids and liquids.
Nasofibroscopy and endoscopy found salivary stasis in the pyriform sinus with regular swelling with normal mucosa amyloidosis (Ghekiere O, 2003) (Bhavani RS, 2010).

4.5 Oral cavity

Amyloid involvement of tongue is almost always secondary to systemic amyloidosis and localized involvement is extremely rare (Fahrner KS, 2004).
Oral amyloidosis usually appears as multiple soft nodules, sometimes hemorrhagic, which can cause macroglossia (Asaumi J, 2001). Alternatively the nodular masses may resemble benign tumors such as granular cell tumors, schwannomas, neurofibromas and neuromas (Muto T, 1991).
Yellow nodules or raised white lesions occurring predominately along the lateral border are also common.
Sometimes petechiae, ecchymoses and hemorrhagic blisters can also be observed (van der Waal RI, 2002).
Primary localized amyloidosis of the palate is extremely rare and only a few cases have been reported (Balatsouras DG, 2007) (Stoor P, 2004) (Alvi A, 1999).
This type of lesion tends to spread locally and occasionally may result in bleeding, epistaxis, and middle ear effusion.

4.6 Cervical lymph nodes

Few documented cases of isolated amyloidosis of cervical lymph nodes have been reported (Shi Q, 2000) (Bielsa S, 2005).

4.7 Amyloid goiter

Amyloid goiter is a rare pathology due to massive amyloid infiltration of thyroid tissue, which causes diffuse or localized enlargement of the gland. It can be totally asymptomatic or causes only non-specific symptoms (compression of adjacent structures, tracheal deviation). Thyroid dysfunction (hypothyroidism or hyperthyroidism) is rare (Cavallaro G, 2006).

4.8 Ear

Physical examination demonstrates brownish or red mass that occupied ear canal, eventually blocked the visualization of the tympanic membrane associated with conductive hearing loss in tone audiometry (Yamazaki K, 2011).
It's like an otitis externa that did not respond to local treatment.

5. Diagnosis

Localized amyloid tumors in the head and neck region are an extremely rare manifestation that is not usually associated with either multiple myeloma or systemic amyloidosis (Godbersen GS, 1992).

Amyloidosis is a disease complex resulting in the extracellular deposition of waxy insoluble fibrillar protein material called "amyloid." It was first described by Rokitansky in 1842 (Rokitansky K, 1842). Amyloid consists of relatively insoluble fibrils composed of polypeptide chains arranged in a twisted ß-pleated sheet configuration. This particular protein configuration accounts for its characteristic staining properties and permits identification by light microscopy. It imparts unique chemical properties like resistance to protease digestion and insolubility, which promote continued deposition within organs.

Therefore a biopsy from the head and neck that reveals amyloid necessitates evaluation for systemic involvement either by rectal biopsy (75% positive) or fat aspiration of the anterior abdominal wall (90% positive) (Nandapalan V, 1998). The absence of Congo red staining of biopsy specimens from either of these 2 sites establishes that the amyloidosis is not systemic (Simpson GT 2nd, 1984). Specific organ involvement may also be excluded by laboratory or radiologic studies (Nandapalan V, 1998).

While amyloid can be suspected on routine hematoxylin-eosin sections, special stains are important for definitive diagnosis. With the polarized microscope, amyloid is seen to have a green birefringence when stained with Congo red.

The etiology, treatment, and outcome of systemic amyloidosis are totally different from localized amyloidosis. The mean survival of patients with systemic amyloidosis is between 5 to 15 months, whereas patients with localized amyloidosis have excellent prognosis (Fahrner KS, 2004).

Final diagnosis relied on histopathological analysis of endonasal endoscopic biopsy samples taken under local or general anesthesia (Yakoot A, 2010).

Histopathological examination showed eosinophil areas, staining positively on Congo red (expressing birefringence under polarized light and on thioflavin) (Yakoot A, 2010).

These histologic features indicated a diagnosis of amyloidosis without signs of underlying malignancy (no abnormalities on general check-up).

On Congo red staining, green birefringence is revealed under polarized light, and electron microscopy shows 8–10-nm wide, straight, and unbranching fibrils.

6. Radiology

The computed tomography (CT) provides excellent information on the anatomic location and topography of different laryngeal benign lesions (Aspestrand F, 1989) but cannot be used to differentiate inflammatory masses from benign neoplasms.

Computed tomography scans demonstrate focal amyloidosis as a well-defined, submucosal, homogeneous, hyperdense, soft tissue mass in association with calcifications (Gean-Marton AD, 1991) (Panda NK, 2007) (Pitkäranta A, 2000).

The lesions are described as well-defined homogeneous soft-tissue masses, with no or only a slight degree of contrast enhancement (Godbersen GS, 1992).

Occasionally, areas of localized calcification within the mass are present (Hegarty JL, 1993) (Parmar H, 2010).

Although the amyloidosis does not generally induce osteolysis, one case of nasopharyngeal amyloidosis with sinus extension and bone destruction suggestive of malignancy has been reported (Zhuang YL, 2005).

MRI is considered as the technique of choice for diagnosis, showing characteristic aspects: intermediate T1-weighted signal and T2-weighted hyposignal, as in striated muscle, without modification in fat-suppression sequences (Zhuang YL, 2005). This signal pattern is due to the amyloid protein deposits, which are fibrillar as in striated muscle fiber (Gilad R, 2007).

The laryngeal lesion reveals intensity equal to that of surrounding muscles on T1 and remains isointense or slightly hyperintense on T2. In comparison, chondrosarcoma of the larynx is hypo- to isointense on T1 and hyperintense on T2, which parallels the appearance of mature hyaline cartilage. Therefore, T2 signal-intensity characteristics might help differentiate amyloidoma from chondrosarcoma.
Following administration of a contrast agent, the lesion exhibits slight or no enhancement. A "fluffy" appearance of bones surrounding the lesion has been shown, which is thought to result from an osteoblastic reaction provoked by submucosal deposition of amyloid fibrils (Chin SC, 2004).
MRI studies show a submucosal mass with low signal intensity in both T1- and T2-weighted images, and mild enhancement in postcontrast T1-weighted images. However, Motosugi *et al*. (Motosugi U, 2007) reported a case of localized nasopharyngeal amyloidosis which showed high signal intensity on T2-weighted images and marked early enhancement in the periphery of the mass on dynamic contrast-enhanced MRI. This phenomenon was attributed to plasmacyte infiltration which occasionally occurs with amyloidosis, leading to hypervascularity.
The exact mechanism underlying the signal hypointensity of amyloid in T2-weighted images is unknown. There are 3 possible mechanisms described (Gean-Marton AD, 1991): (1) signal loss results from enhanced T2 decay because of the presence of static or slowly fluctuating internal magnetic fields within adjacent amyloid proteins held in fixed positions within the folded protein; (2) rapid chemical exchange and spin-spin interactions may occur between the amyloid protein and adjacent water molecules; and (3) because the amyloid microenvironment is composed of a heterogeneous micromagnetic mixture of collagen, calcification, and vessels as well as the amyloid fibrils, the T2 hypointensity may result from differences in diamagnetic susceptibility (Gean-Marton AD, 1991).

7. Differential diagnosis

7.1 Systemic amyloidosis

Localized amyloidosis has been described in nearly every organ system. Development of systemic disease following a diagnosis of localized amyloidosis was rare, occurring in 2% of cases. One-quarter of patients with primary systemic amyloidosis presents with clinical involvement of only one organ, but investigation leads to the recognition of widespread involvement (Merlini G, 2003).

7.2 Larynx

In the larynx, the amyloid deposits are submucosal and homogeneous and are not associated with the cartilage changes.
They are firm lesions that tend to occur in the supraglottic larynx, although all laryngeal sites may be affected.
The differential diagnosis includes other submucosal diseases such as laryngeal sarcoidosis, lymphoma, and pseudotumor.

7.3 Tracheobronchial

Wegener's granulomatosis, laryngotracheobronchial papillomatosis, idiopathic laryngotracheal stenosis, sarcoidosis, or tracheobronchitis with ulcerative colitis may radiologically mimic tracheobronchial amyloidosis.

7.4 Tongue

There is a wide differential diagnosis that should be considered in patients presenting with macroglossia or nodular tongue lesions.

For generalized macroglossia, amyloid, tuberculosis, lymphangioma, hypothyroidism, acromegaly, lingual infarction caused by giant-cell arteritis, idiopathic muscular hypertrophy, and Beckwith-Wiedemann syndrome should be considered.

For nodular lesions, fibroma, lipoma, granular cell tumor, sarcoma, lingual thyroid. and salivary gland tumors would be among the differential.

7.5 Palate

The differential diagnosis includes candidiasis, vascular lesions and Kaposi's sarcoma.

7.6 Amyloid goiter

Microscopic infiltration of the thyroid gland by amyloid is an uncommon but well recognized phenomenon and significant enlargement of the thyroid due to deposition of amyloid is rarely seen. This condition has to be distinguished from other types of goiters and at times from malignancy.

Amyloid goiter may be diagnosed by cytopuncture (Kapadia HC, 2001), although there is a problem of differential diagnosis with medullary carcinoma, where amyloid deposit is also found in 50% to 80% of cases.

However, cytopuncture excludes other thyroid cancers, notably anaplastic cancer and lymphoma.

Anatomopathology enables positive diagnosis in case of amyloid deposit detected in the form of an amorphous substance showing yellow-green double refraction under polarized light on Congo red staining.

7.7 Sino-nasal and nasopharyngeal amyloidosis

The differential diagnosis for lesion within the maxillary antrum includes malignant conditions such as squamous cell carcinoma, lymphoma, metastatic disease, plasmacytoma, melanoma, neuroblastoma, paraganglionoma and haemangiopericytoma, benign lesions including hamartoma, and local fungal infections such as mucormycosis and aspergilloma (Birchall D, 1997).

Although amyloidoma of the nose is rare, it should be considered as part of the differential diagnosis of a nasal mass presenting with nasal obstruction and epistaxis, even in pediatric patients. The approach should include a scan to exclude a possible vascular tumor, the as the appearance may be fleshy.

Because of its infrequent occurrence, nasopharyngeal amyloidosis is rarely considered in the differential diagnosis of nasopharyngeal tumor (Panda NK, 2007). Initial endoscopic examination in the present case was rather suggestive of adipose tumor.

8. Treatment

In systemic disease, high-dose chemotherapy followed by stem cell reconstitution seems to provide the most effective treatment (Gertz MA, 2005).

The localized amyloidosis has excellent prognosis and never evolves towards systemic forms (Pitkäranta A, 2000). Hence, it may be treated with surgical excision (Stoor P, 2004) , especially to reduce local soft palate mass (Zhuang YL, 2005) (Pitkäranta A, 2000).

Certain reports consider surgery for nasopharyngeal forms to be merely palliative (Stoor P, 2004). In case of recurrence, surgical revision may be recommenced, depending on symptomatology.
For localized amyloidosis, symptomatic removal is required. But, close follow-up is necessary because the recurrence rate reported is as high as 50%.

8.1 Sinonasal cavities and nasopharynx

For localized amyloidosis of the nasopharynx, surgical resection is usually chosen as the treatment modality for symptomatic relief. However, the extent of surgical excision has changed from radical excision to a more conservative approach.
The rationale behind this alteration is that a conservative surgical approach may be associated with a slower rate of recurrence (Lesserman JA FD, 1995). Unfortunately, failure to prevent recurrence by means of surgical excision has been found in the majority of case reports. Simpson *et al.* (Simpson GT 2nd, 1984) have suggested a reduction in the recurrence rate following laser resection.
Emerging treatment approaches employing agents that disrupt fibril formation, destabilize amyloid deposits, or interfere with interaction between amyloidogenic proteins and accessory molecules have shown promising results in animal models, and clinical applications are underway (Lachmann HJ, 2003).
The surgical treatment must be done in case of airway obstruction, bleeding, and other severe symptoms.
Radiotherapy may be a treatment option in nasal and nasopharyngeal amyloidosis.
Symptoms may be improved with steroid treatment (Pearlman AN, 2010).

8.2 Larynx

It is important to treat laryngeal amyloidosis because untreated cases can progress to vocal fold fixation, severe dysphonia, and airway obstruction. Alternatively, "expectant" management with no intervention may be indicated in certain carefully selected cases, owing to the slow progression of the disease over many years (Avitia S, 2007).
In isolated laryngeal amyloidosis, treatment is primarily endoscopic surgical removal of masses that interfere with laryngeal or airway function. The likelihood of recurrent or residual disease is significant (Talbot AR, 1979).
Because of the propensity of amyloid to infiltrate blood vessels, cold resection of amyloid lesions may be complicated by bleeding. Although CO2 laser excision may be advantageous, if used in very extensive lesions, there may be scarring (Talbot AR, 1979) (Simpson GT 2nd, 1984).
In very large lesions, external approaches, employing laryngofissure for treatment of diffuse subglottic and tracheal amyloidosis have been used. Repeated excision and curettage may be necessary to achieve stabilization of lesions and subsequent decannulation of those patients who are tracheotomy dependent.
Local or systemic steroids are ineffective in controlling or reversing lesions of amyloidosis (Finn DG, 1982).
Bartels *et al* (Bartels H, 2004) found five patients over a period of 13 years who had localized laryngeal amyloidosis and free light chains were found in one patient. Amyloid interfering with laryngeal or airway function was excised during microlaryngoscopy. Cold endoscopic excision for glottic deposits and CO2 laser for supraglottic lesions provided best results. Four patients developed recurrent disease.

Localization	Number of cases	Surgery	Phonosurgery or laser treatment	IMRT	External beam radiation (45 Gy)	Tracheostomy	Tympanostomy tube	Steroid	Supervision
Larynx	12	0	9	2	1				
Nasopharynx	7	4					1		2
Tongue	6	3	1						2
Palate	4	1							3
Trachea	3						1		2
Hypopharynx	3	1							
Sinonasal cavities	2	1						1	
Oropharynx	1								1
Tonsil	1								
Ear	1								
Cervical lymph nodes	3								

IMRT: Intensity Modulated Radiotherapy

Table 2. Distribution by the therapeutic procedure

Dedo and Izdebski (Dedo HH, 2004) reported 10 consecutive laryngeal amyloid patients in whom amyloid was found on the undersurface of both vocal folds in two and submucosally in unilateral or bilateral vestibular folds in eight cases. Direct microlaryngoscopy with CO2

laser excision was done on one side at a time to try to prevent anterior commissure scarring. Followup after the first operation was 6 months to 16 years, with an average of 6.5 years. Four vestibular fold patients required re-excision on the same side after the first operation.
Because full resection is difficult, Neuner GA (Neuner GA, 2010) recommends a combination of surgery and radiation therapy to cure localized amyloidosis of the larynx.
Other treatments options that have been described include corticosteroids, radiotherapy, and agents like colchicine and melphalan. However, these modalities have yielded variable results (Avitia S, 2007).

8.3 Trachea

The management is dependent on symptoms. Treatment options are bronchoscopic approaches, surgery, radiation, and observation. Therapy may not be required for asymptomatic patients. Cases with proximal airway obstruction are difficult to manage.
Bronchoscopic methods are mechanical debulking, endobronchial laser ablation, and stent or balloon dilatation in selected patients with stenosis. These methods are preferable and safer than surgery. However, such methods may be ineffective and repeated procedures may be required (Gillmore JD, 1999) (O'Regan A, 2000). Bleeding is a common complication of these procedures.
Sometimes external beam radiation or surgical resection is necessary (Neben-Wittich MA, 2007).

8.4 Oral cavity

The definitive treatment of localized amyloidosis was cited to be surgery. Surgery alone may be 100% curative (Fahrner KS, 2004).

8.5 Cervical lymph node

There is no specific treatment for this uncommon entity (Bielsa S, 2005). The cervical lymph nodes dissection may be done (Shi Q, 2000).

8.6 Amyloid goiter

Fine-needle aspiration biopsy can be performed to exclude malignant lesions. In order to diagnose amyloid goiter, definitively thyroidectomy is often necessary. Surgical intervention is indicated either for aesthetic purposes or to relieve the pressure symptoms (Villa F, 2008).

9. Conclusion

Amyloidosis is a rare disease with multifactorial pathogenesis.
Localized amyloidosis affecting the head and neck region is an uncommon and benign process, which has almost no clinical consequences. Once the diagnosis has been made, an extensive workup for systemic amyloidosis should be undertaken. This should include abdominal fat biopsy or rectal biopsy.

10. References

Alvi A, Goldstein MN. (1999). Amyloidosis of the palate. *Otolaryngol Head Neck Sur,* 120, 2, (1999), pp. 287.

Aono J, Yamagata K & Yoshida H. (2009). Local amyloidosis in the hard palate: a case report. *Oral Maxillofac Surg,* 13, 2, (2009), pp. 119-22.

Asaumi J, Yanagi Y & Hisatomi M. (2001). CT and MR imaging of localized amyloidosis. *Eur J Radiol*., 39, 2, (2001), pp. 83-7.

Aspestrand F, Kolbenstvedt A & Boysen M. (1989). CT findings in benign expansions of the larynx. *J Comput Assist Tomogr,* 13, 2, (1989), pp. 222-5.

Avitia S, Hamilton JS & Osborne RF. (2007). Surgical rehabilitation for primary laryngeal amyloidosis. *Ear Nose Throat J,* 86, 4, (2007), pp. 206- 208.

Balatsouras DG, Eliopoulos P & Assimakopoulos D. (2007). Primary local amyloidosis of the palate. *Otolaryngol Head Neck Surg,* 137, 2, (2007), pp 348-9.

Bartels H, Dikkers FG & van der Wal JE. (2004). Laryngeal amyloidosis: localized versus systemic disease and update on diagnosis and therapy. *Ann Otol Rhinol Laryngol,* 113, 9, (2004), pp. 741-8.

Bhavani RS, Lakhtakia S & Sekaran A. (2010). Amyloidosis presenting as postcricoid esophageal stricture. *Gastrointest Endosc,* 71, 1, (2010), pp. 180-1, discussion 181.

Bielsa S, Jover A & Porcel JM. (2005). Isolated lymph node amyloidosis. *Eur J Intern Med,* 16, 8, (2005), pp. 619.

Birchall D, Fields JM & Poon CL. (1997). Case report: focal amyloidosis of the maxillary antrum: plain film, CT and MR appearances. *Clin Radiol*. 52, 5, (1997), pp.392-4.

Caldarelli DD, Friedberg SA & Harris AA. (1979). Medical and surgical aspects of the granulomatous diseases of the larynx. *Otolaryngol Clin North Am,* 12, 4, (1979) pp. 767-81.

Cavallaro G, Polistena A & Fornari F. (2006). A case of primitive amyloid goiter. *G Chir,* 27, 8-9, (2006), pp. 315-7.

Chin SC, Fatterpeckar G & Kao CH. (2004). Amyloidosis concurrently involving the sinonasal cavities and larynx. *AJNR Am J Neuroradiol,* 25, 4, (2004), pp. 636-8.

Comenzo RL. (2006). Amyloidosis. *Curr Treat Options Oncol,* 7, 3, (2006), 225-36.

Dedo HH, Izdebski K. (2004). Laryngeal amyloidosis in 10 patients. *Laryngoscope,* 114, 10, (2004), pp. 1742-6.

Fahrner KS, Black CC & Gosselin BJ. (2004). Localized amyloidosis of the tongue: a review. *Am J Otolaryngol,* 25, 3, (2004), pp. 186-9.

Finn DG, Farmer JC Jr. (1982). Management of amyloidosis of the larynx and trachea. *Arch Otolaryngol,* 108, 1, (1982), pp. 54-6.

Gallivan GJ, Gallivan HK. (2010). Laryngeal amyloidosis causing hoarseness and airway obstruction. *J Voice,* 24, 2, (2010), pp. 235-9.

Gean-Marton AD, Kirsch CF & Vezina LG. (1991). Focal amyloidosis of the head and neck: evaluation with CT and MR imaging. *Radiology,* 81, 2, (1991), pp. 521-5.

Geller E, Freitag SK & Laver NV. (2010). Localized Nasopharyngeal Amyloidosis Causing Bilateral Nasolacrimal Duct Obstruction, *Ophthal Plast Reconstr Surg,* [Epub ahead of print].

Gertz MA, Lacy MQ & Dispenzieri A. (2005). Amyloidosis: diagnosis and management. *Clin Lymphoma Myeloma,* 6, 3, (2005), pp. 208-19.

Ghekiere O, Desuter G & Weynand B. (2003). Hypopharyngeal amyloidoma. *AJR Am J Roentgenol* 2003, 181, 6, (2003), pp. 1720-1.

Gilad R, Milillo P & Som PM. (2007). Severe diffuse systemic amyloidosis with involvement of the pharynx, larynx, and trachea: CT and MR findings. *AJNR Am J Neuroradiol,* 28, 8, (2007) pp. 1557-8.

Gillmore JD, Hawkins PN. (1999). Amyloidosis and the respiratory tract. *Thorax,* 54, 5, (1999) pp. 444-51.

Glenner GG. (1980). Amyloid deposits and amyloidosis: the beta-fibrilloses (second of two parts). *N Engl J Med,* 302, 24, (1980), pp. 1333-43.

Godbersen GS, Leh JF & Hansmann ML. (1992). Organ-limited laryngeal amyloid deposits: clinical, morphological, and immunohistochemical results of five cases. *Ann Otol Rhinol Laryngol,* 101, 9, (1992), pp. 770-5.

Hegarty JL, Rao VM. (1993). Amyloidoma of the nasopharynx: CT and MR findings. *AJNR Am J Neuroradiol* , 14, 1, (1993), pp. 215-8.

Kapadia HC, Desai RI & Desai IM. (2001). Amyloid goiter--a case report. *Indian J Pathol Microbiol,* 44, 2, (2001), pp. 147-8.

Kerner MM, Wang MB & Angier G. (1995). Amyloidosis of the head and neck. A clinicopathologic study of the UCLA experience, 1955-1991. *Arch Otolaryngol Head Neck Surg,* 121, 7, (1995), pp. 778-82.

Kurokawa H, Takuma C & Tokudome S. (1998). Primary localization amyloidosis of the sublingual gland. *Fukuoka Igaku Zasshi,* 89, 7, (1998), pp. 216-20.

Kyle RA, Bayrd ED. (1975). Amyloidosis: review of 236 cases. *Medicine (Baltimore),* 54, 4, (1975), pp. 271-99.

Lachmann HJ, Hawkins PN. (2003). Novel pharmacological strategies in amyloidosis. *Nephron Clin Pract,* 94, 4, (2003), pp. c85-8.

Lesserman JA FD. (1995). Endoscopic treatment of amyloidosis of the nasopharynx. *Am J Rhinol,* 9, (1995), pp. 43–7.

Lewis JE, Olsen KD & Kurtin PJ. (1992). Laryngeal amyloidosis: a clinicopathologic and immunohistochemical review. *Otolaryngol Head Neck Surg,* 106, 4, (1992), pp. 372-7.

Malek RS, Wahner-Roedler DL & Gertz MA. (2002). Primary localized amyloidosis of the bladder: experience with dimethyl sulfoxide therapy. *J Urol,* 168, 3, (2002), pp. 1018-20.

Mcalpine JC, Fuller AP. (1964). Localized Laryngeal Amyloidosis, A Report Of A Case With A Review Of The Literature. *J Laryngol Otol,* 78, (1964), pp. 296-314.

Merlini G, Bellotti V. (2003). Molecular mechanisms of amyloidosis. *N Engl J Med,* 349, 6, (2003), pp. 583-96.

Motosugi U, Ichikawa T & Araki T. (2007). Localized nasopharyngeal amyloidosis with remarkable early enhancement on dynamic contrast-enhanced MR imaging. *Eur Radiol,* 17, 3, (2007), pp. 852-3.

Mufarrij AA, Busaba NY & Zaytoun GM. (1990). Primary localized amyloidosis of the nose and paranasal sinuses. A case report with immunohistochemical observations and a review of the literature. *Am J Surg Pathol,* 14, 4, (1990), pp. 379-83.

Muto T, Sato K & Lutcavage GJ. (1991). Multiple nodules of the lip, cheeks, and tongue. *J Oral Maxillofac Surg.* 49, 9, (1991), pp. 1003-6.

Nandapalan V, Jones TM & Morar P. (1998). Localized amyloidosis of the parotid gland: a case report and review of the localized amyloidosis of the head and neck. *Head Neck,* 20, 1, (1998), pp. 73-8.

Neben-Wittich MA, Foote RL & Kalra S. (2007). External beam radiation therapy for tracheobronchial amyloidosis. *Chest,* 132, 1, (2007), pp.262-7.

Neuner GA, Badros AA & Meyer TK. (2010). Complete resolution of laryngeal amyloidosis with radiation treatment. *Head Neck,* (2010), [Epub ahead of print].

O'Regan A, Fenlon HM & Beamis JF Jr. (2000). Tracheobronchial amyloidosis. The Boston University experience from 1984 to 1999. *Medicine (Baltimore),* 79, 2, (2000) pp. 69-79.

Panda NK, Saravanan K & Purushotaman GP. (2007). Localized amyloidosis masquerading as nasopharyngeal tumor: a review. *Am J Otolaryngol,* 28, 3, (2007), pp. 208-11.

Pang KP, Chee LW & Busmanis I. (2001). Amyloidoma of the nose in a pediatric patient: a case report. *Am J Otolaryngol,* 22, 2, (2001), pp. 138-41.

Parmar H, Rath T & Castillo M. (2010). Imaging of focal amyloid depositions in the head, neck, and spine: amyloidoma. *AJNR Am J Neuroradiol,* 31, 7, (2010), pp. 1165-70.

Passerotti GH, Caniello M & Hachiya A. (2008). Multiple-sited amyloidosis in the upper aerodigestive tract: case report and literature review. *Braz J Otorhinolaryngol,* 74, 3, (2008), pp.462-6.

Patel A, Pambuccian S & Maisel R. (2002). Nasopharyngeal amyloidosis. *Am J Otolaryngol.* 23, 5, (2002), pp. 308-11.

Pearlman AN, Jeffe JS & Zynger DL. (2010). Localized amyloidosis of the nasal and paranasal mucosa: a rare pathology. *Am J Otolaryngol,* 31, 2, (2010) pp. 130-1.

Penner CR, Muller S. 2006. Head and neck amyloidosis: a clinicopathologic study of 15 cases. *Oral Oncol,* 42,4, (2006), pp.421-9.

Pitkäranta A, Malmberg H. (2000). Localized amyloid tumor of the nasopharynx. *Otolaryngol Head Neck Surg,* 122, 2, (2000) pp. 309-10.

Pribitkin E, Friedman O & O'Hara B. (2003). Amyloidosis of the upper aerodigestive tract. *Laryngoscope,* 113, 12, (2003), pp. 2095-101.

Rokitansky K. (1842). Handbuch der Pathologischen Anatomie. *Vienna, Austria: Braunmuller and Seide,* 311, (1842) pp. 384–424.

Scott PP, Scott WW Jr & Siegelman SS. (1986). Amyloidosis: an overview. *Semin Roentgenol,* 21, 2, (1986), pp. 103-12.

Shi Q, Fan K & Chen H. (2000). Localized amyloidosis of cervical lymph nodes. *Chin Med J,* 113, 2, (2000), pp.184-5.

Simpson GT 2nd, Strong MS & Skinner M. (1984). Localized amyloidosis of the head and neck and upper aerodigestive and lower respiratory tracts. *Ann Otol Rhinol Laryngol.* 93, (4 Pt 1), (1984), pp. 374-9.

Stimson PG, Tortoledo ME & Luna MA. (1988). Localized primary amyloid tumor of the parotid gland. *Oral Surg Oral Med Oral Pathol,* 66, 4, (1988), pp. 466-9.

Stoor P, Suuronen R & Lindqvist C. (2004). Local primary (AL) amyloidosis in the palate. A case report. *Int J Oral Maxillofac Surg,* 33, 4, (2004), pp. 402-3.

Talbot AR . (1979). Laryngeal amyloidosis. *Otolaryngol Clin North Am,* 12, 4, (1979), pp. 767-81.

Thompson PJ, Citron KM. (1983). Amyloid and the lower respiratory tract. *Thorax,* 38, 2, (1983), pp. 84-7.

Utz JP, Swensen SJ & Gertz MA. (1996). Pulmonary amyloidosis. The Mayo Clinic experience from 1980 to 1993. *Ann Intern Med,* 15, 124, 4, (1996), pp.407-13.

Van Der Waal RI, Van De Scheur MR & Huijgens PC. (2002). Amyloidosis of the tongue as a paraneoplastic marker of plasma cell dyscrasia. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod,* 94, 4, (2002), pp. 444-7.

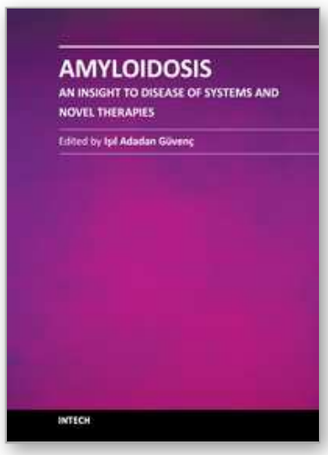
Villa F, Dionigi G & Tanda ML. (2008). Amyloid goiter. *Int J Surg,* 6, Suppl 1, (2008), pp. S16-8.

Walker PA, Courey MS & Ossoff RH. (1996). Staged endoscopic treatment of laryngeal amyloidosis. *Otolaryngol Head Neck Surg,* 114, 6, (1996), pp. 801-5.

Yakoot A, Giusiano S & Sanjuan M. (2010). An unusual cause of serous otitis media. *Eur Ann Otorhinolaryngol Head Neck Dis,* 127, 4, (2010), pp. 156-8.

Yamazaki K, Sato H & Ishijima K. (2011). A case of hemodialysis-associated amyloidosis localized to the external auditory canal. *Auris Nasus Larynx*, pp. 295-9. Epub 2010 Oct 28.

Zhuang YL, Tsai TL & Lin CZ. (2005). Localized amyloid deposition in the nasopharynx and neck, mimicking nasopharyngeal carcinoma with neck metastasis. *J Chin Med Assoc.*68, 3, (2005), pp. 142-5.



Amyloidosis - An Insight to Disease of Systems and Novel Therapies

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Amyloidosis is a benign, slowly progressive condition characterized by the presence of extracellular fibrillar proteins in various organs and tissues. It has systemic or localized forms. Both systemic and localized amyloidosis have been a point of interest for many researchers and there have been a growing number of case reports in the literature for the last decade. The aim of this book is to help the reader become familiar with the presentation, diagnosis and treatment modalities of systemic and localized amyloidosis of specific organs or systems and also cover the latest advancements in therapy.

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