Chapter 3

Trigeminal Schwannomas

Breno Nery, Marcelo Nery Silva,
Rodrigo Antônio Fernandes Costa,
Leandro César Tângari Pereira, Eduardo Quaggio,
Fred Bernardes Filho and George Peter Stevens

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Abstract

Trigeminal schwannomas (TS) are rare entities occurring in various trigeminal nerve locations and present a peak incidence between the fourth and fifth decades of life, being more common in women. Patients usually present with symptoms of trigeminal nerve dysfunction. Depending on the tumor's topography, various approaches might be used to obtain its gross total resection. Trigeminal schwannoma's classification, nuances of the approaches, pathology, postoperative care, and outcomes are revised as follows. In conclusion, anatomical knowledge and the disease's comprehension are essential when dealing with such lesions, and despite their rarity, we must be obstinately committed to the surgical technique and devoted to the patient's functional postoperative outcome.

Keywords: schwannoma, Schwann cells, trigeminal nerve, cranial nerves

1. Introduction

Schwannomas are benign tumors originating from Schwann cells, which form the myelin sheath around cranial and peripheral nerves. When occurring in various trigeminal nerve locations, these tumors account for 0.1–0.4% of all intracranial tumors and 1–8% of intracranial neurinomas [1, 2]. There is no doubt that vestibular schwannomas represent the vast majority of brain schwannomas, but other unusual topographies can be seen, in descending order, in the following cranial nerves: glossopharyngeal, vagus, facial, accessory, hypoglossal, oculomotor, trochlear, and abducent [3–5]. The trigeminal schwannomas (TS) present a peak incidence between the fourth and fifth decades of life, being more common in women. Since their

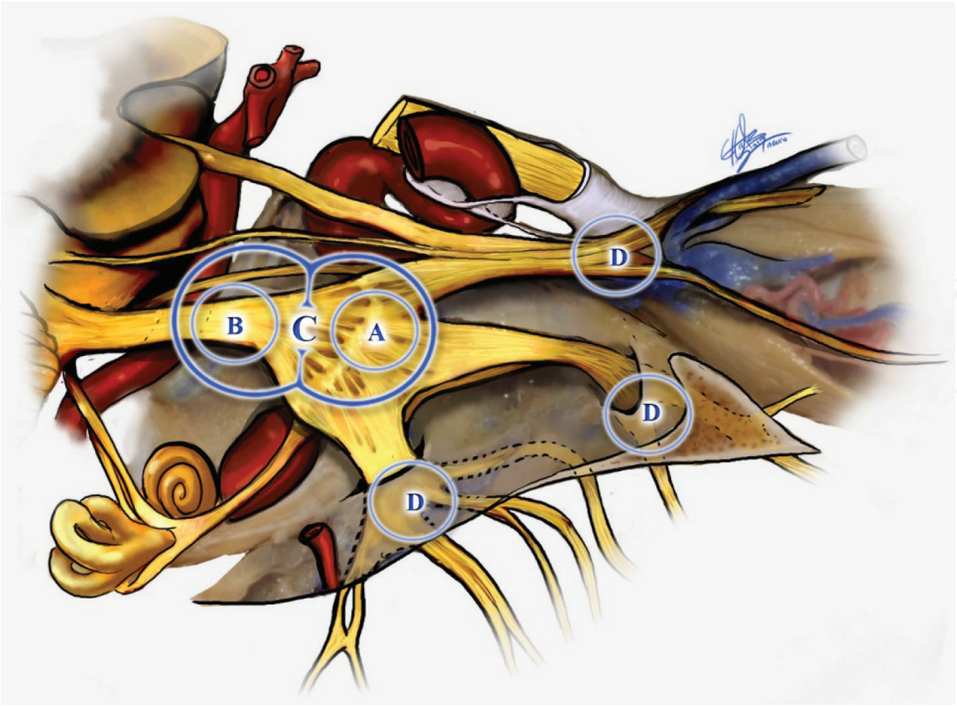

Figure 1. Jefferson's classification revised by day and Fukushima. Type A: tumors of the middle fossa in the interdural space; type B: tumors of the posterior fossa in the subdural space; type C: dumbbell-shaped tumors (afflicting both middle and posterior fossae); and type D: TS which may arise from any extracranial division of the trigeminal nerve.

location directly affects the surgical approach, several classification systems were proposed for trigeminal schwannomas. The first classification scheme was proposed by Jefferson [6] and modified by Day and Fukushima who classified the TS according to their anatomical location and apparent origin of the trigeminal nerve in their classical paper [7]. According to those authors, TS can grow in one, two, or all three of the following compartments: subdural compartment (pontocerebellar angle), interdural compartment (lateral wall of the cavernous sinus and Meckel's cavity), and extradural or extracranial compartment (orbit, pterygopalatine fossa, and infratemporal fossa). Advances in imaging modalities and the ability to accurately diagnose these lesions in nuclear magnetic resonance imaging (MRI) allow us to easily demonstrate these extension patterns for the posterior, middle, and infratemporal fossae. In this classification system, tumors can therefore be divided into four groups: type A: tumors of the middle fossa in the interdural space; type B: tumors of the posterior fossa in the subdural space; type C: dumbbell-shaped tumors (afflicting both middle and posterior fossae); and type D: TS which may arise from any extracranial division of the trigeminal nerve (**Figure 1**).

2. Clinical presentation

Most patients present with trigeminal dysfunction in the opening of its clinical picture, more common being the decreased sensitivity in the ophthalmic divisions (V1), maxillary (V2), and mandibular (V3). The progressive decrease in sensitivity is compatible with the slow growth form of this type of tumor; one of its major concerns is the involvement of V1 segment, with consequent decreased corneal sensitivity and keratitis. Pain may also be a part of the clinical picture, mainly in the ganglionic subtype, having been found in more than 40% of patients

in the initial series of Day and Fukushima [7]. Wanibuchi et al. published a series with 105 patients operated on with trigeminal neurinomas, and the most frequent preoperative clinical picture was facial hypoesthesia, present in more than 65% of the patients. Facial pain was found in approximately 23% of the cases, followed by diplopia related to paresis of the abducent nerve, headache, and ataxia/vertigo as symptoms in 17, 14, and 10%, respectively [8]. Pain with longer duration, without a specific trigger, and associated with low response to carbamazepine therapy or other anticonvulsant medications may occur, characterizing atypical facial pain and always raising the hypothesis of a secondary cause for trigeminal neuralgia.

Compression of intracranial nerves that travel through the cavernous sinus can determine clinical diplopia (due to the compression of III, IV, and VI cranial nerves), *tic douloureux* (V compression), exophthalmos (due to the invasion of the orbit), and decrease of auditory acuity or facial mimic by compression of the VII/VIII complex in the posterior fossa. Patients with small and oligosymptomatic tumors can be clinical and imaging followed with intervals between 6 and 12 months. If there is worsening of symptoms, the revaluation must be anticipated, due to a small possibility of malignant tumor of the trigeminal nerve. Patients who are symptomatic or do not respond to drug therapy should be promptly operated, and good neurosurgical technique offers low morbidity and mortality for these tumors today.

2.1. Preoperative preparation

Detailed clinical examination and imaging study with MRI with and without contrast constitute the primary evaluation to define tumor extension and neurovascular relationship of the tumor. Computed tomography (CT) helps in assessing bone involvement, and digital angiography should be performed in cases of suspected engulfment of the internal carotid arteries (ICA) during growth through the middle fossa or of the vertebral artery (VA) in case of growth of the tumor lesion into the posterior fossa. The audiogram may be necessary in the preoperative evaluation in cases with the presence of hearing loss, or suspected vestibulocochlear nerve invasion, for preoperative documentation of hearing status.

2.2. Intraoperative monitoring

Monitoring through somatosensory-evoked potential (SEP) and motor-evoked potential (MEP) is mandatory in this surgery. In case of tumor extension to the posterior fossa and compression of the brainstem/cranial nerves, brainstem auditory-evoked potential is also part of the armamentarium needed during surgery, reducing the possibility of injury to the intracranial nerves.

3. Approaches

The choice of the ideal approach for the surgical treatment of trigeminal schwannomas depends essentially on the location of the tumors of this region. Tumors originating in Gasserian's ganglion or whose major component is found in the cavernous sinus (Type A) may benefit from temporal craniotomy associated or not with zygomatic osteotomy for anterolateral interdural access (Dolenc's approach) [9] or frontotemporal craniotomy for temporopolar extradural resection [7]. Tumors originating from the root of the fifth cranial nerve (Type B) can be approached via simple suboccipital craniotomy and retrosigmoid approach [10]. Combined accesses may

be used in cases of tumors with extension to the middle fossa and posterior fossa (Type C), and in some cases, the combined accesses may be necessary (temporal craniotomy and combined presigmoid approach) [10]. In lesions that extend from the middle fossa to the posterior fossa (DSTS), we can also utilize a two-step surgery in short surgical times for complete resection of the lesion. We believe that in these cases one should choose the initial approach for the most symptomatic lesion and after 2 or 3 months to resect the residual lesion through alternative approaches. Endoscopic-assisted approaches may also help in such complex situations with tumors extending for both middle and posterior cranial fossae, such as the endoscope-assisted retrosigmoid intradural suprameatal approach (EA-RISA), which may help to achieve gross total resection in such situations [11]. Type D tumors may be resected with the help of various approaches, depending on which extracranial division of the V nerve is attacked [7, 12]. **Table 1** resumes the mainly utilized approaches depending on the tumor's topography. DSTS or type C trigeminal schwannomas represent a unique pathology that requires special attention and are properly discussed in the following sections.

3.1. Frontotemporal approach with zygomatic osteotomy (middle fossa approach)

The extended access to the middle fossa is ideal for schwannomas originating in the middle fossa or with associated invasion of the orbit or infratemporal fossa. We believe that this approach has advantages over the subtemporal approach because it allows better access and visualization of the lateral wall of the cavernous sinus, with less temporal lobe retraction and less possibility of traction of the Labbé venous group and a lower incidence of venous infarction or thrombosis. Larger tumors with extension to the posterior fossa can be approached via this access, by anterior petrosectomy in the Kawase triangle. The patient should be positioned in dorsal decubitus position, with the head rotated between 30 and 45° to the contralateral side of the lesion, with a slight elevation of the ipsilateral shoulder. The head should be fixed in the Mayfield head holder, and the use of lumbar drainage is optional.

An inverted question mark incision should be performed, starting at the lower margin of the zygoma, anterior to the tragus, and should be directed posteriorly and superiorly above the external acoustic meatus, and then again directed anteriorly behind the hairline, toward the midline. The temporal muscle should be dissected through careful incision of the superficial and deep fascial layers anteriorly, in order to preserve the frontal branches of the facial nerve. The zygoma must be dissected in its subperiosteal plane and resected obliquely. Its anterior and posterior oblique resection facilitates its refitting at the end of the surgical procedure. We prefer to maintain the

Tumor's topography	Approaches
Type A	Frontotemporal approach associated or not with zygomatic osteotomy (anterolateral, subtemporal)
Type B	Retrosigmoid approach
Type C	One-step surgery (middle fossa approach or EA-RISA) or combined approaches (anterior and posterior petrosectomy)
Type D	Frontotemporal approach + orbital osteotomy (V1); frontotemporal approach + temporopolar via (V2); frontotemporal approach with zygomatic osteotomy + infratemporal approach (V3)

Table 1. Possible approaches for TS based on their topographies.

superficial fascia of the temporal muscle adhered to the zygoma, and then, we rebound the muscle and zygoma in the inferior direction. Four burr holes must be made, two of them being as close as possible to the floor of the middle fossa, one in the keyhole and another in the posterior aspect of the superior temporal line. The holes are then connected, completing the craniotomy (**Figure 2**).

After craniotomy, which can be combined with orbital osteotomy in case of anterior extension of the tumor, access may be made by extradural or intradural route. When the tumor involves only a small portion of the anterior or inferior cavernous sinus, extradural approach is the most appropriate route. When the tumor extends to the uppermost and posterior portion of the cavernous sinus, the intradural access may help its resection.

3.2. Extradural approach

After craniotomy, the dura mater should be gradually elevated opposite to the floor of the middle fossa, from anterior to posterior (Hakuba), inferior to superior (Kawase), or superior to inferior directions (Dolenc). Medially to the posterior root of the zygomatic arch is the middle meningeal artery, which should be coagulated and sectioned near the dura mater, avoiding its retraction into the spinal foramen with the presence of bleeding that may be difficult to control. The middle fossa should be drilled with exposure of the superior orbital fissure and the round and oval foramina, where the trigeminal branches of V2 and V3 leave, respectively. After adequate exposure of the lateral wall of the cavernous sinus, sutures can be performed at the base of the temporal dura throughout its length, facilitating its superior retraction and allowing exposure of the lateral wall of the cavernous sinus without the inconvenient fall of the temporal lobe over the surgeon's visual field, as well as greatly reducing the manipulation of the temporal lobe. Tumor bulging is visualized on the lateral wall of the cavernous sinus, and an incision in the superficial surface, running through

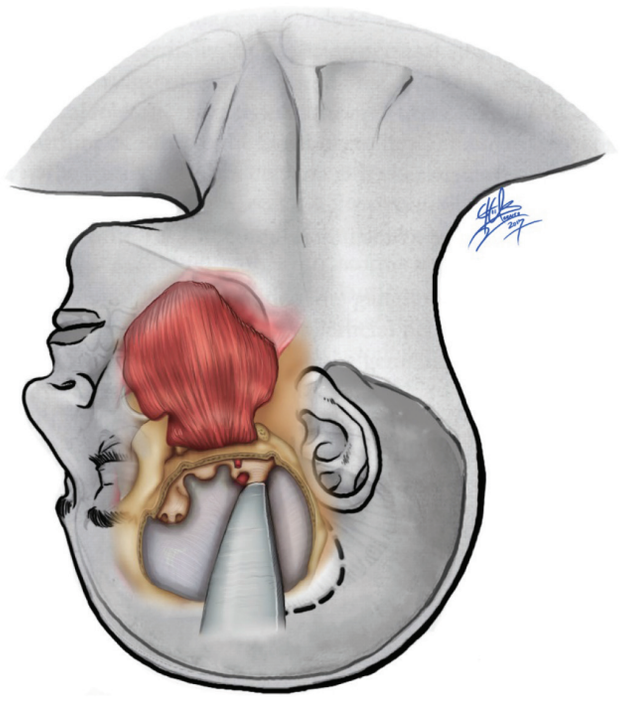

Figure 2. Frontolateral approach without orbital osteotomy associated with zygomatic osteotomy to facilitate the visualization of the middle fossa. The center of the exposure is the branches of the trigeminal nerve.

the superior orbital fissure, V2 and V3, should be performed (peeling of the cavernous sinus), exposing the tumor. The tumor should then be decompressed using an ultrasonic aspirator.

After sufficient dissection, the tumor's capsule should be dissected from the fascicles of the trigeminal nerve with microsurgical technique. While the surgeon dissects, the assistant should help with fine suction, in order to facilitate adequate visualization of the neural structures.

A Doppler ultrasound helps to find the pathway of the intracavernous internal carotid artery, enhancing the surgeon's safety when manipulation of the medial portion of the trigeminal nerve is needed. Venous bleeding of the cavernous sinus is easily controllable by fibrin glue or surgical and local compression. In the presence of tumor portion that enters the posterior fossa, we generally find the Meckel's cavity enlarged, and the drilling of the petrous apex enhances the access to the posterior fossa after opening the posterior fossa dura and ligation of the superior petrosal sinus. In some cases, the dilated Meckel's cave permits access to the posterior fossa without the drilling of the petrous apex (**Figure 3**).

3.3. Intradural approach

Soon after the previously described craniotomy, we should make the opening of the frontotemporal dura mater, continuing with the dissection of the Sylvian fissure over its entire extension to allow mobilization of the temporal lobe. If necessary, we must perform the coagulation and sectioning

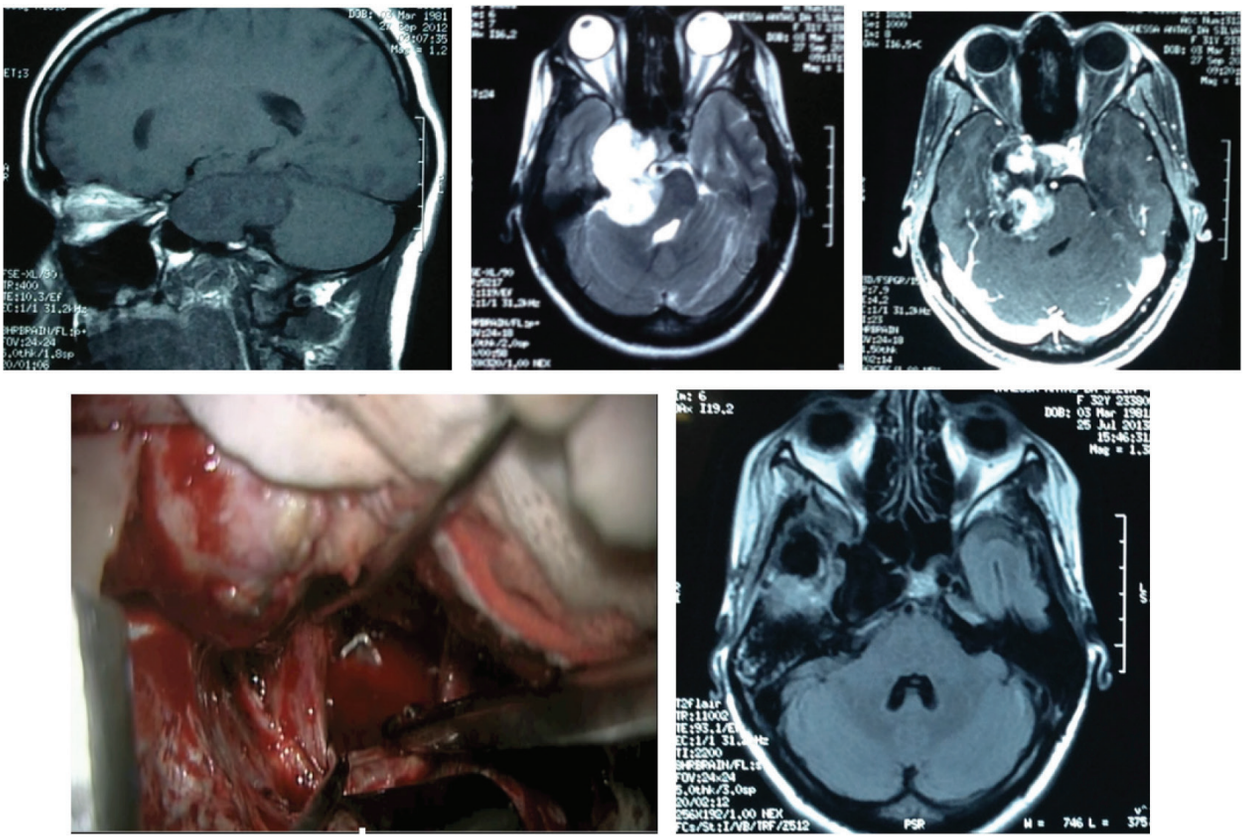

Figure 3. From the left to the right: T1, T2, and T1 post-gadolinium images. Since no dural enhancement or bone hyperostosis or destruction was observed, the main hypothesis was trigeminal schwannoma. Bone remodeling and trigeminal pore dilation can also be seen. After extradural peeling of the middle fossa and dural opening, we can see V3 retracted and tumor exeresis.

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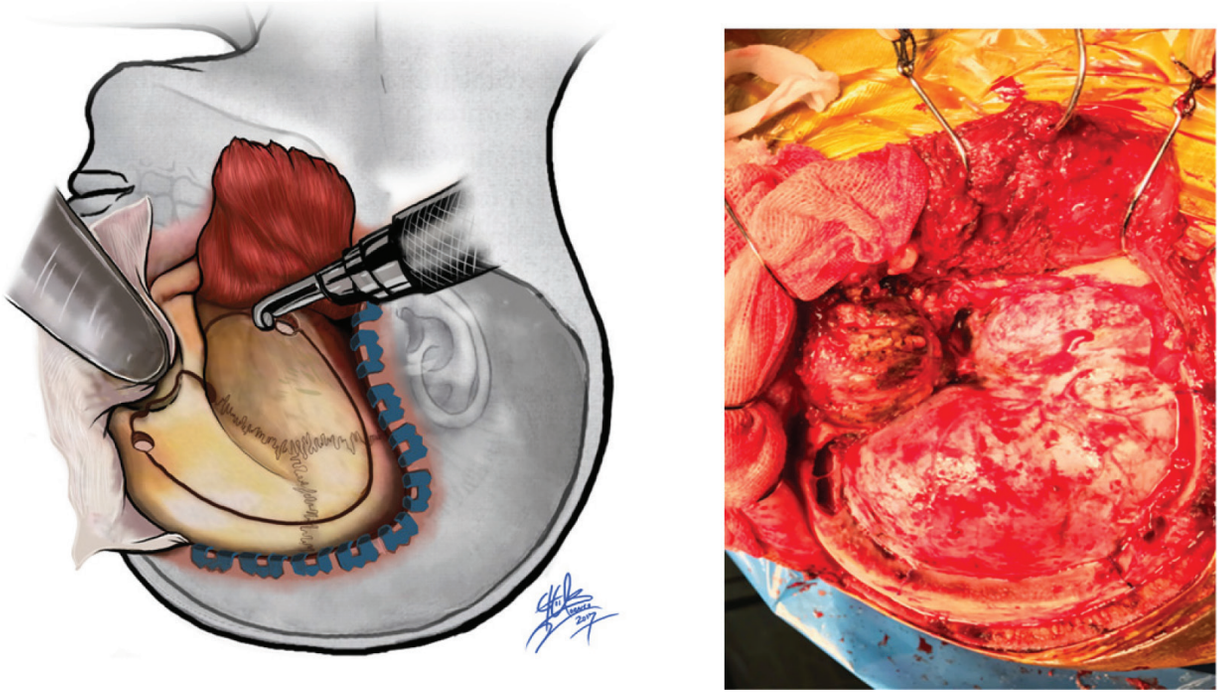

Figure 4. From the left to the right, Cranio orbital zygomatic approach (drawing) and intraoperative view (on the right), after osteotomy of the roof of the orbit and zygomatic arch.

of the temporal lobe connecting veins to the sphenoparietal sinus, especially in cases where the drainage pattern of the superficial Sylvian veins is not exclusively anterograde. Preoperative angiographic study assists in determining the drainage pattern of Sylvian veins. With the temporal lobe released, its mobilization will be enough to expose the lateral wall of the cavernous sinus through the temporopolar, subtemporal, or trans-Sylvian routes. The opening of the lateral wall of the cavernous sinus is followed where a greater tumor bulging can be seen. Tumor excision is then followed after opening of the lateral wall of the cavernous sinus (**Figures 4–6**).

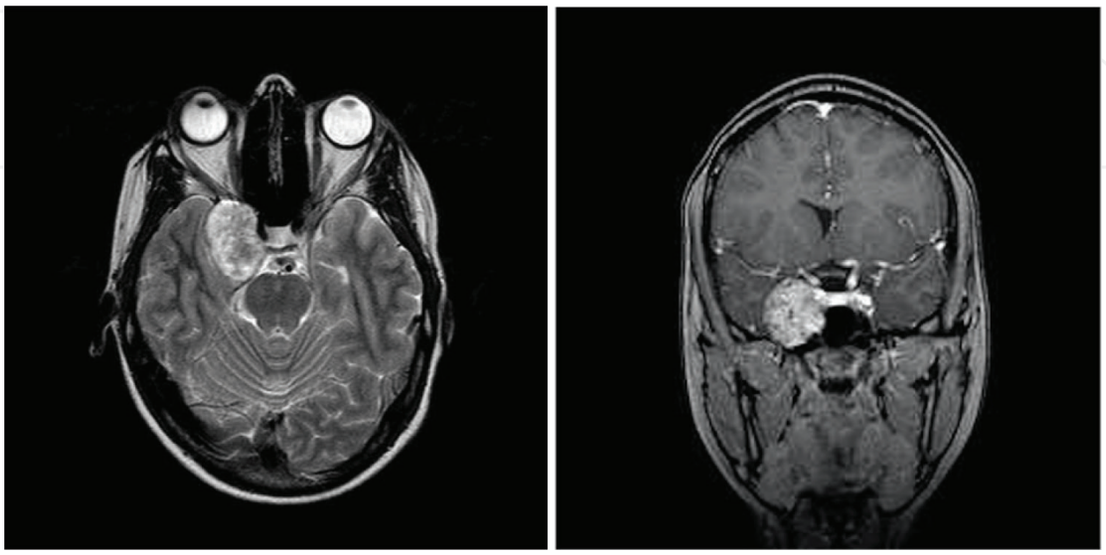

Figure 5. From the left to the right, preoperative T2-weighted image (axial) and T1 coronal with gadolinium of a type a right trigeminal schwannoma operated in our institution.

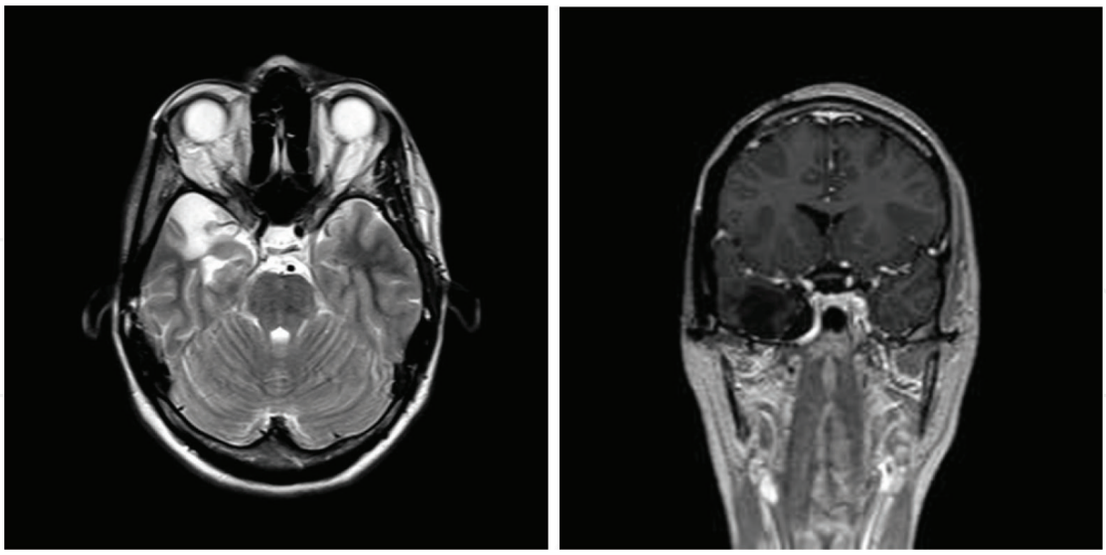

Figure 6. From the left to the right, postoperative T2-weighted image (axial) and T1 coronal with gadolinium after 5 years showing gross total resection without recidive of the tumor.

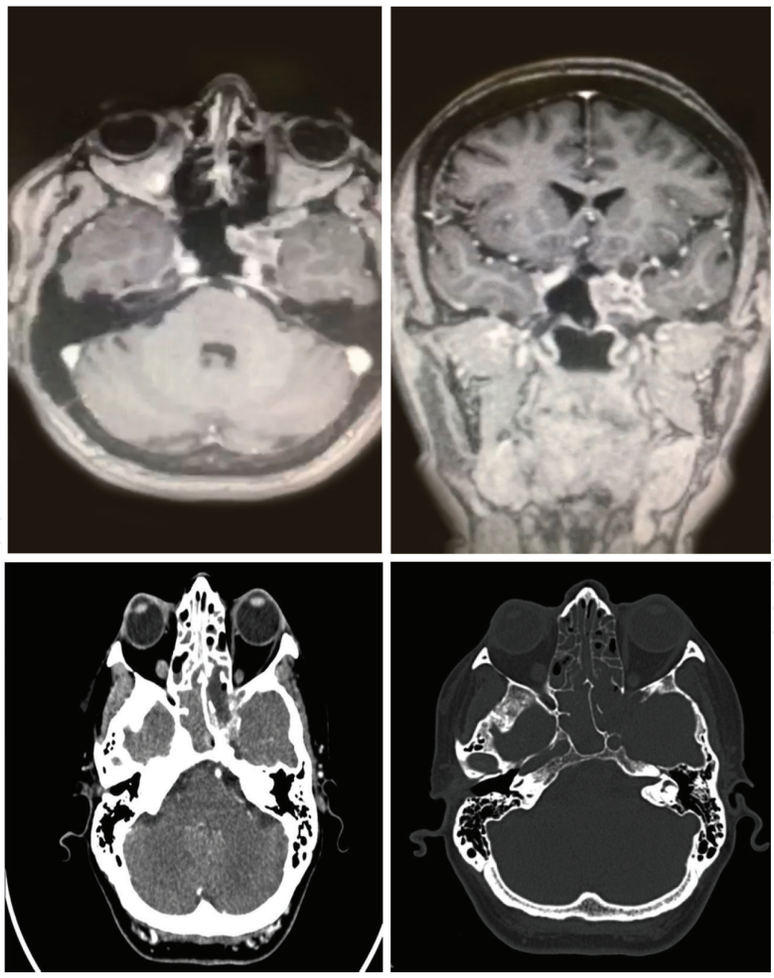

Figure 7. From the left to the right: T1 gadolinium enhanced TS. The patient underwent endoscopic transpterygoid endonasal approach for an anteromedially located trigeminal schwannoma. In the middle, postoperative CT scan, and bone scan on the right show nuances of the approach and gross total resection of the tumor.

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3.4. Lateral combined approach

The extent of the tumor to the posterior fossa may require combined approaches when the tumor volume in this region is more significant. Presigmoidal retrolabyrinthine approach associated with partial labyrinthectomy may be used, aiming for the preservation of hearing when it is preserved before surgery.

3.5. Endoscopic endonasal approach

Anteromedial-located trigeminal schwannomas can be better resected by endoscopic endonasal approaches. This is a very rare condition among these tumors, but such an approach avoids direct lesion of the trigeminal nerve since the fifth nerve will be located lateral to the approach (**Figure 7**).

4. Pathology

Schwannomas are benign Schwann cell tumors that, when in the central nerve system, affect sensitive roots. The most frequent location is in the vestibular root of the VIII and rarely is present in the trigeminal location. Microscopically, the tumor is formed by elongated cells, arranged in bundles that intersect. The nuclei tend to be arranged parallel to each other, an aspect called the arrangement in palisades, very typical of the schwannoma. The spaces with few nuclei between palisades are constituted only by the cytoplasm of Schwann cells and are called bodies of Verocay. There is no necrosis or mitosis (**Figure 8**).

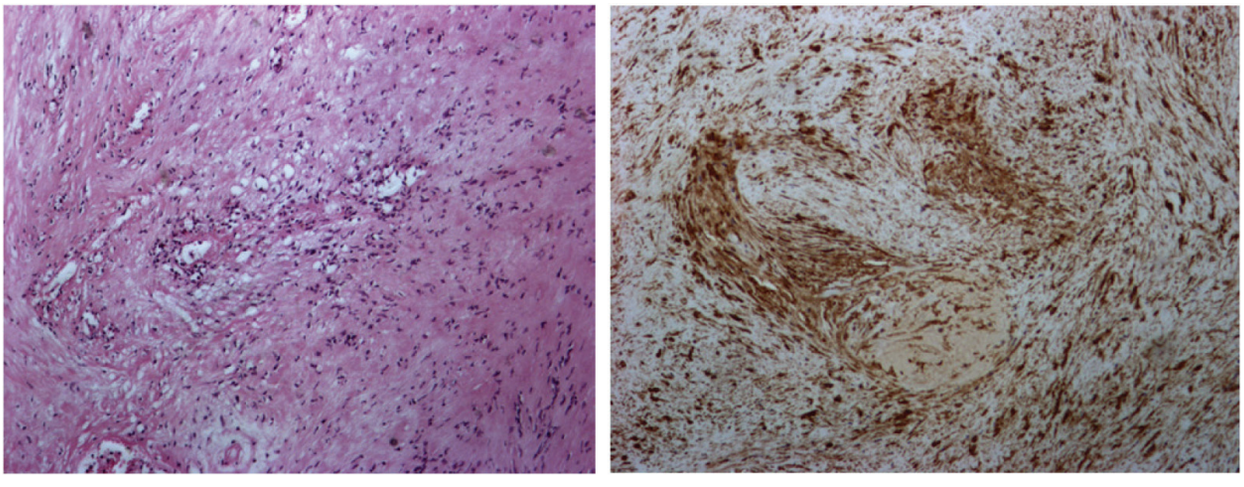

Figure 8. On the left side, an increase of 400× shows benign fusocellular neoplasia next to blood vessels with thickened walls. On the right side, immunohistochemistry with a magnification of 400×, presenting strong expression of neoplastic cells to protein S100.

5. Postoperative care

Early trigeminal dysfunction can be observed after surgery but usually presents progressive improvement. Trigeminal pain has significant reduction or even resolution after surgery. A complication that might occur and should be exhaustively addressed is the hypoesthesia

of the ophthalmic branch of the trigeminal nerve, which may develop keratitis as the most severe complication due to decreased corneal reflux. The traction of the dura mater during the peeling of the middle fossa may lead to retraction of the major superficial petrosal nerve and even its avulsion with lesion of the geniculate ganglion, culminating in facial nerve injury. Other complications are dependent on the manipulation of the cranial nerves in contact with the tumor, in addition to cerebrospinal fluid fistula, infection, and venous infarction due to excessive retraction of the temporal lobe.

6. Results

In the largest series of patient cases operated by trigeminal schwannomas, total or near-total resection could be reached in up to 82% of cases [2, 8]. The most common symptom in the postoperative period was facial hypoesthesia, occurring in 65–86% of cases [2, 8]. Residual facial pain was seen in 23% of the patients in the series of Wanibuchi et al. and diplopia was reported in this same series in 20% of cases, with persistent deficit in only 5% of patients and worsening of the deficit in only 1 patient of 105 operated patients [8]. Tumor resection, in lesions that extend to multiple regions, is feasible with a high rate of total resection [12].

7. Conclusion

Advances in imaging modalities and the progressive improvement of microsurgical instruments and surgical techniques have greatly improved surgical results in the treatment of this pathology. We must be obstinately committed to the surgical technique and devoted to the patient's functional postoperative outcome for their reintegration into normal life and improvement of their quality of life.

Conflict of interest

None.

Author details

Breno Nery[1]*, Marcelo Nery Silva[2], Rodrigo Antônio Fernandes Costa[1], Leandro César Tângari Pereira[1], Eduardo Quaggio[1], Fred Bernardes Filho[3] and George Peter Stevens[4]

*Address all correspondence to: brenonery84@gmail.com

1 Department of Neurosurgery, São Francisco Hospital, Ribeirão Preto, São Paulo, Brazil

2 Department of Neurosurgery, Heliopolis Hospital, São Paulo, São Paulo, Brazil

3 Department of Internal Medicine, São Francisco Hospital, Ribeirão Preto, São Paulo, Brazil

4 Department of Neurosurgery, SSM Health Saint Louis University Hospital, St. Louis, Missouri, USA

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