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Article

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Giant trigeminal schwannomas-case report and a short literature review

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Abstract: Trigeminal schwannomas (TS) are rare tumors that are located in the posterior and middle cranial fossae in about a quarter of the cases. The presentation pattern is different according to the involved compartment and the goal of surgery is complete removal. We present the case of a 35-year old woman who presented a left sided TS extending from the posterior to the middle fossa and cavernous sinus. We review the relevant literature regarding diagnosis and surgical treatment of these tumors.
Key words: trigeminal nerve, schwannoma, multiple cranial fossae

Introduction

Trigeminal nerve schwannomas (TS) are rare entities, accounting for 1 to 8% of all intracranial schwannomas and less than 0.4% of all intracranial tumors [4]. Women seem to be affected more often than men and the majority of cases present in the third or fourth decade of life [2]. The tumors can arise from the trigeminal dorsal root entry zone, the Gasserian ganglion or any of the three branches of the Vth cranial nerve and this variability in origin can lead to complex tumors, which extend in one or in multiple cranial fossae [3]. The symptomatology is not related to tumor size as most patients usually present with mild facial hypoesthesia, various visual disturbances, and sometimes facial pain [14]. Various authors presented their series in the literature, but surgical experience in giant trigeminal schwannomas is still limited. We present a case of a giant trigeminal schwannoma and perform a short review of the current literature.

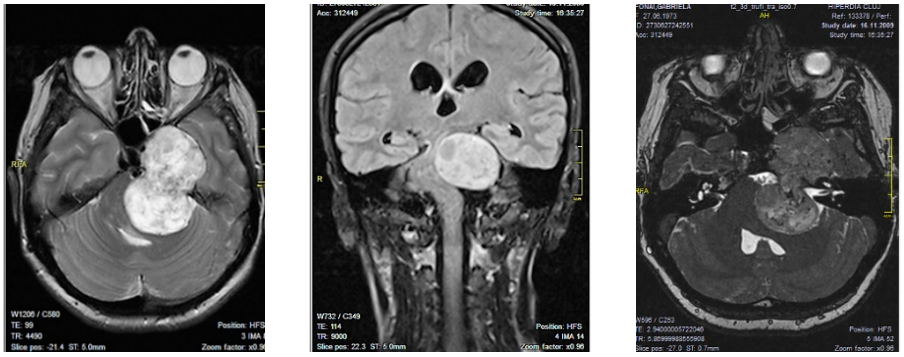
Case report

A 35-year old woman presented in our department complaining of a 6 months old left-side facial paresthesia, difficulty in chewing, loss of balance, facial pain that was lancinating, alternating from moderate to severe, irradiating to the lower face, aggravated by chewing. The clinical exam revealed a mild left hemifacial spasm, left hemifacial paresthesia and a mild facial asymmetry. Cerebellar signs included ataxic gait and dysdiadochokinesia.

A gadolinium-enhanced MRI was performed and revealed a defined, solid tumor extending from the left posterior fossa to the middle fossa and cavernous sinus (Figure 1). The tumor dislocated the brainstem medially and exerted cerebellar compression, and compressed without incorporating the carotid artery. The tumor was hyperintense both on T1 and T2-weighted images and enhanced significantly after contrast injection. Based on

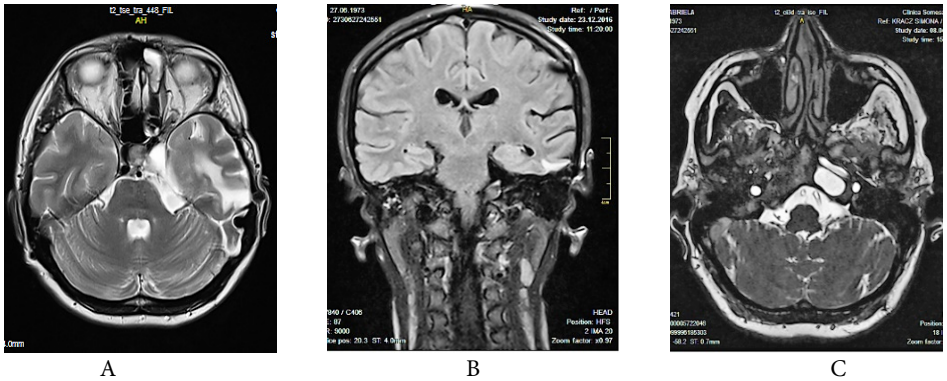
the clinical and radiological findings, a provisional diagnosis of trigeminal schwannoma was made and surgery for tumor removal was recommended.

The patient was positioned on the left side and a subtemporal craniotomy, with a retrosigmoid extension, was performed. The tumor was solid, showed a good delineation from vascular and nervous elements, and was mildly vascularized. A complete removal was performed. After surgery, the patient has a transient worsening of the facial paresthesia, which remitted in a couple of days. Pathology analysis showed spindle-shaped bipolar cells that formed fascicles and palisading appearance. Immunohistochemistry of the tumor was positive for S100, CD34, vimentin and confirmed the diagnosis of TS. The postoperative MRI showed complete tumor removal and no tumor regrowth was noted on the one and five years MRI (Figure 2). At the one year control, the patient did not show any neurological deficits and declared she returned to a completely normal life.



A B C

Figure 1 - Preoperative MRI showing a hyperintense tumor in T2-weighted imaging extending from the posterior to the middle fossa and cavernous sinus (A), which compresses the brainstem (B), and has a cystic structure (C)



A B C

Figure 2 - Postoperative MRI at 5 years showing axial (A) and coronal (B) complete tumor removal, with no signs of brainstem compression and a liquid filled area where the tumour was located (C)

Discussions

In 1953, Jefferson tried to characterize trigeminal schwannomas and designed a classification which divided these tumors into three categories, according to their origin [7]. Type A included tumors originating anterior of the Gasserian ganglion and located in the middle cranial fossa, type B included tumors in the posterior fossa, originating from the trigeminal root and type C defined tumors located both in the posterior and middle cranial fossae [7]. Yoshida and Kawase also offered a classification of TS into six categories and divided those that affect multiple cranial fossae [13]. About 25 % of tumors appear in multiple compartments and because of the rarity of TS, these tumors are infrequent and no standard treatment exists [8]. What authors agree on is the fact that complete surgical excision is usually curative and as such, whenever feasible, this should be the goal of surgery [10].

Whenever TS arise from the root of the trigeminal nerve, they involve the posterior fossa [5]. Because of the fact that they can also appear on any of the nerve branches, these tumors can also grow epidurally into the infratemporal fossa or orbit [5]. Pathological findings are usually those of any schwannoma, but it must be considered that in about 1% of cases these tumors are associated with neurofibromatosis and therefore other consecutive tumors or, very rarely, a type of melanotic tumor can occur in the trigeminal nerve [13].

The trigeminal nerve has a mix component, of both sensory and motor fibers. Therefore symptoms of decreased facial sensibility or diminished force of mastication can raise the suspicion of a TS [1]. Facial hypoesthesia is the initial symptom in 60% of cases, but it must be kept in mind that because of their large size, symptoms related to any other cranial nerve, cerebellar or brainstem symptoms are also possible [9].

Radiological appearance plays a pivotal role in diagnosing and planning the surgical treatment of TS. MRI is the method of choice. Tumors usually appear as hypo or isointense on T1 and as hyperintense on T2-weighted imaging, with high enhancement after contrast administration [1]. A CT scan offers a complementary view of these tumors, as it can demonstrate the existence of bone erosion [14]. Usually, contrast enhancement on CT is homogeneous or it can have a circum enhancement in cystic lesions [14].

Surgical strategy in TS is generally tailored on the tumor and no definitive recommendations can be made. Because of the difference in the classification systems, lack of volumetric studies and report of trigeminal function, there is an inevitable difficulty in comparing different series, the same as in the case of other complex skull base tumors [6, 9]. Some authors divide the possible approaches into conventional approaches and skull-base approaches [14]. The conventional approaches include the subtemporal intradural, frontotemporal infratemporal, while the skull base approaches include anterior transpetrosal, zygomatic infratemporal or zygomatic transpetrosal [12, 13]. Some authors describe the use of adjuvant technical means, e.g. endoscopy, especially in those tumors that extend into multiple cranial compartments [8, 9, 11]. As previously mentioned, the goal of surgery should be a complete removal, without sacrifice of neurological function. It must be kept in mind that sometimes, transient neurological deficits

can appear after surgery, but these disappear in a few days [12]. In our experience, the use of a conventional approach is sufficient in most cases, in order to achieve complete removal. Tumors are usually solid and soft and detachment from ICA or nervous structures is performed easily. The preservation of function of the trigeminal nerve is possible in those cases in which the tumor is well delimitated, although this is rarely the case in giant tumors.

Conclusions

Our article presents the case of a giant TS and discusses relevant aspects of the neurosurgical literature regarding this aspect. TS are rare tumors and they're treatment is still not standardized. Patients usually present with symptoms related to impaired trigeminal function, but cerebellar and brainstem signs are also possible in the case of TS which invade multiple cranial fossae. Complete surgical removal is usually curative and should be the purpose whenever possible without affecting neurologic function. Surgery through a modified subtemporal approach is a feasible option in those tumors that do not extend in the infratemporal sinus.

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