CASE REPORTS

Surgical Treatment of Cerebellopontine Angle Trigeminal Schwannoma Via a Retrosigmoid Intradural Approach: A Case Report

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OBJECTIVES: Schwannoma of the trigeminal nerve is a rare entity: those located in the posterior fossa account for 20% of all cases. The aims of this report are to describe a case of cerebellopontine angle schwannoma and to define the clinically common presentation, the medical imaging needed, and the surgical procedure used to treat this lesion.

MATERIALS AND METHODS: We report the clinical features, neuroradiologic imaging, and management of a case of trigeminal schwannoma located in the cerebellopontine angle, arising from the sensitive root of the trigeminal nerve, and managed at the Lyon Department of Otoneurosurgery at the Centre Hospitalier Lyon-Sud in Pierre Bénite, France.

RESULTS: This trigeminal schwannoma developed in the cerebellopontine angle without extension into the Meckel's cave. Our patient presented with facial pain and facial palsy, and medical imaging revealed a cerebellopontine mass. Surgery, using a retrosigmoid intradural route, was performed, and a diagnosis of trigeminal schwannoma from the roots of the sensitive part of the nerve was made. The lesion was resected, and the patient left the hospital 7 days after surgery. This article discusses the differential diagnosis of cerebellopontine angle tumors. It also shows the different surgical approaches to removing a trigeminal schwannoma, according to its location and its volume.

CONCLUSION: The retrosigmoid intradural approach is an effective and safe procedure, with low morbidity, for removing cerebellopontine angle trigeminal schwannomas—even in cases of large tumors—when the Meckel's cave is not invaded. It is important to define the extent of the lesion site to determine the most appropriate surgical technique.

Schwannomas of the trigeminal nerve are benign, rare tumors, accounting for 0.07% to 0.36% of all intracranial tumors and 0.8% to 8% of intracranial schwannomas.[1,2] For the vestibulocochlear nerve, the schwannoma seems to born in a transitional area, called the Obersteiner-Redlich zone, the "junction zone" of glia cells and Schwann sheaths of the cranial nerves. That is why the most common site of development for a vestibular schwannoma is the cerebellopontine angle (CPA) and the internal auditory canal (IAC).

The physiopathology of trigeminal schwannomas is less known, and a schwannous proliferation can appear in the CPA, in the cavernous sinus, in the infratemporal fossa, or in 1 of the 3 branches of the trigeminal nerve after the gasserian ganglion. We present a case of trigeminal schwannoma developed in the CPA, revealed by facial pain, and removed via retrosigmoid intradural procedure.

CASE REPORT

A 46-year-old man presented with an isolated and regressive paroxysmal sharp facial pain. The pain lasted up to 10 minutes and then quickly stopped. During this acute facial pain, increased lacrimation and nasal congestion were present. The following day, the patient had different facial pain combined with left inferior facial palsy and hypoesthesia on the third sensitive root of the left trigeminal area. This clinical presentation occurred over 7 days. The patient denied hearing loss, tinnitus, or vertigo.

On physical examination, the patient was in good health. Medical examination revealed no vestibular or cerebellous symptoms. Results of otologic and neurologic examinations were normal. An audiologic examination showed no hearing loss, and the short latency brainstem auditory evoked potential showed significant increases of the I-III and I-V intervals.

Magnetic resonance imaging revealed a CPA mass, measuring 24 mm by 20 mm, next to the internal auditory canal without extension inside and with displacement of the brainstem (Figure 2). This lesion appeared as a hypointense mass on T1-weighted images and as an isointense mass on T2-weighted images. After injection of gadolinium, the lesion was heterogeneously enhanced (Figure 1).

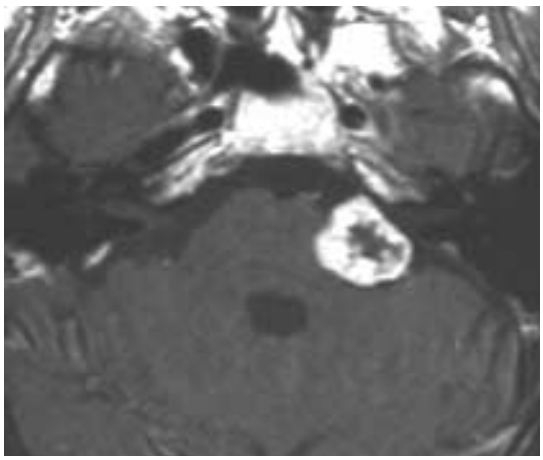

Figure 1: MRI: Axial gadolinium-enhanced T1 sequence. Well-circumscribed and heterogeneous lesion is shown compressing the left pons.

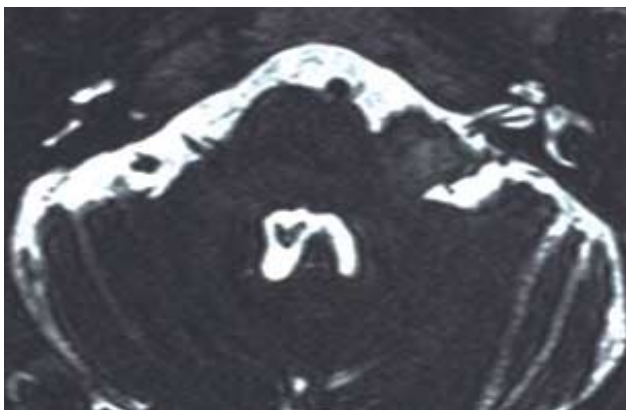

Figure 2: MRI: Axial T2 sequence. The lesion developed next to the inner auditory canal without extending into it. The cochlear and vestibular nerves can be seen.
MRI = magnetic resonance imaging.

Surgery utilizing a left retrosigmoid intradural route with craniotomy of a 4-cm skull section was performed under facial-nerve electromyographic control. The voluminous lesion presented with a central cyst and was very adherent to the brainstem. After a partial debulking, a diagnosis of trigeminal nerve schwannoma originating from the inferior fascicle of the trigeminal nerve root was confirmed. The surgical procedure

allowed us to separate the tumor from the facial and acoustic nerves, and the lesion was totally removed with no complications and with more than 90% nerve-fascicle preservation. Histologic examination confirmed tumor type as Antoni type A schwannoma.

After surgery, the patient described a partial sensitive left mandible. He was discharge from the hospital after 1 week.

DISCUSSION

The trigeminal nerve is less frequently involved in schwannomas compared with vestibulocochlear and facial nerves. Schwannomas can occur at any age, but they are more predominant in the fourth and fifth decades, with a long and insidious preoperative course. They often grow to significant size before their presence is recognized. Our case was uncommon because of the patient's quick evolution to painful facial palsy.

Jefferson presented the first detailed description of trigeminal neurinoma in 1955 and first classified the tumors' relationship to their principal location.[3] Type I includes schwannoma of the roots of the posterior fossa and CPA. Type II concerns schwannoma that develop in the gasserian ganglion. Type III includes schwannoma of the trigeminal branches.[4] The differentiation between type I and type II can be made through the schwannoma's extension into the Meckel's cave, representing the limit of the middle and the posterior fossa. Because of its unique anatomic structure and location, the Meckel's cave represents a common avenue for the spread of disease from the posterior to the middle fossa and vice versa.[5]

A fourth level (type IV) was added to this classification system that included tumors originating from a major division of the trigeminal nerve in the gasserian ganglion and extending extracranially.[6] In this case, the surgical procedure is more difficult because it requires the use combined approaches.

In our patient, the tumor was located at the CPA with no extension into the Meckel's cave. At this location, the differential diagnosis may be difficult because these lesions commonly present with irritating or sensitive dysfunction in the trigeminal territory: ataxia, facial nerve dysfunction, hearing loss, tinnitus, or nystagmus, all due to cranial nerve involvement.[7]

Vestibular schwannomas, by far the most common cerebellopontine neoplasms, most commonly present as a combined intracanalicular/CPA lesion.[8] Large vestibular schwannomas are likely to cause trigeminal nerve symptoms. The IAC is rarely enlarged by trigeminal schwannomas, which distinguishes these tumors from acoustic neurinoma.[9] In our case, we followed the vestibular nerve from the brainstem to the IAC via magnetic resonance imaging.

A posterior petrous meningioma is a possible differential diagnosis, depending on the form of the lesion, but, in our patient, the angle between the tumor and the dural tail wasn't a classic symptom for meningioma.

The lesion in our patient corresponds to facial nerve schwannomas in order of frequency, even after imaging. Vestibular, facial, and trigeminal schwannomas of the CPA may be impossible to differentiate prior to surgery because their imaging characteristics are identical.[10] An extension to the middle fossa can help us to distinguish a trigeminal schwannoma from another type of tumor.

The primary parenchymal neoplasms, such as childhood medulloblastoma, astrocytoma, and ependymoma, show an irregular cerebellous or brainstem extension, so we could exclude them from our differential diagnosis.

In our case, the information provided by magnetic resonance imaging allowed us to diagnosis schwannoma,[11] but its origin at the trigeminal, facial, or vestibulocochlear nerve roots was difficult to determine before surgery.[12]

The surgical procedure to remove trigeminal schwannoma must be defined by the volume and the location of the lesion. Schisano and Olivecrona reported a 1-year mortality rate of 41% when they reviewed the literature prior to 1956.[13] In recent years, however, surgical results have improved,, and there is little mortality. Considering the biologically benign behavior and the tendency toward noninvasiveness of these neoplasms, the complete surgical excision of the tumor with minimal morbidity is the first, if not sole, surgical target.[14]

Recent techniques of neuroimaging and electrophysiologic monitoring can help us reach this target, aiding selection of the best surgical procedure. The choice of optimal surgical approach, including basal approaches, should be based on the correct evaluation of the tumor's location and its relationship to surrounding brain structures. When the Meckel's cave is invaded, the surgical approach can be epidural, epiduro-transduro-transtentorial, or transpetrosal.[15-18] In contrast, when the tumor is located strictly in the posterior fossa of the CPA, the most effective way to remove this lesion is posteriorly, via the retrosigmoid intradural course with its good exposition and permanent hearing preservation. This approach allows us to separate the facial and the vestibulocochlear nerve from the tumor with good control, and we can completely resect the tumor even with brainstem adhesion.

CONCLUSION

In our patient with facial and trigeminal symptomatology, diagnosis was difficult prior to surgery. Considering the biologically benign behavior and the tendency toward noninvasiveness of CPA trigeminal schwannoma, complete surgical excision of the tumor with minimal morbidity is the first, if not sole surgical target. The retrosigmoid intradural approach, which has very low morbidity and preserves hearing function, is an effective and safe procedure for removal of CPA trigeminal schwannoma, even when large, provided the Meckel's cave isn't invaded.[19]

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