

# Trigeminal schwannoma

## Classification and surgical approaches

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### RESUMO

#### **Schwannomas do trigêmeo. Classificação e abordagem cirúrgica**

*Este trabalho revisa e amplia a classificação de Jefferson em relação à localização dos schwannomas do nervo trigêmeo.*

*Os autores apresentam uma série de abordagens cirúrgicas mais adequadas a serem utilizadas para a remoção desses tumores. Utilizando técnicas microcirúrgicas na base do crânio, os autores demonstram as abordagens que, de forma mais direta, eficiente e segura, permitem a remoção dos cinco diferentes tipos de schwannomas. Três casos ilustrativos são também apresentados.*

### PALAVRAS-CHAVE

*Schwannoma do trigêmeo. Classificação. Abordagem cirúrgica.*

### ABSTRACT

*This paper reviews and expands the Jefferson's Classification regarding the localization of trigeminal schwannomas.*

*The authors present a series of specific surgical approaches best suited to remove these tumors. Using skull base neurosurgical techniques, the authors present the most direct, efficient and safety surgical approaches to remove each of five different types of trigeminal schwannomas. Three illustrative cases of their own are also presented.*

### KEYWORDS

*Trigeminal schwannoma. Classification. Surgical approaches.*

## Introduction

Primary tumors of the gasserian ganglion were first described by Smith in 1836, but the first reported attempt at removal of a tumor in this location did not occur until the turn of the century<sup>7,20</sup>.

Trigeminal schwannomas are very uncommon tumors originating within the base of the skull. They account from 0.1 to 0.4% of intracranial tumors and 1 to 8% of the intracranial neurilemmomas<sup>5,16,21,24</sup>.

Trigeminal schwannomas are slow-growing tumors. In our cases pre-admission clinical history varied from 1 to 6 years. Konovalov et al.<sup>10</sup> studied 111 patients having trigeminal schwannoma and discovered that tumors located in the posterior fossa exhibit a shorter clinical history (1 to 3.2 years, av. 2.2 years). Patients with tumors located in the middle fossa have a pre-

admission history of 1.5 to 6 years, average 3.4 years. Tumors located in the branches of the trigeminal nerve had a longer history ranging from 1.5 to 6.2 years, average 3.8 years.

Trigeminal schwannomas are more common in the fourth decade of life and with predominance in female<sup>5,24,27</sup>.

The association with neurofibromatoses type II is present in 10% of patients. Usually the most common symptoms and signs are related with dysfunction of the trigeminal nerve. Facial pain is found in 36% of patients at the time of the diagnosis. Typical trigeminal neuralgia is found in 5 to 10% of patients. Sensory dysfunction within the face is present in 87% of the cases. Masticatory deficit is found in 42% of patients. Disturbance of the others cranial nerves is less common, with the auditory and facial nerves being

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most affected. Tumor extension into the cavernous sinus wall or to the petrous apex can cause a dysfunction of the abducent nerve<sup>28</sup>. In cases with large intracranial extensions, hemiparesis, ataxia, and other neurological signs can occur<sup>5, 24, 27</sup>.

Plain films of the skull demonstrate erosion of the anteromedial portion of the petrous pyramid in 70% of patients. This appearance is not pathognomonic of trigeminal schwannoma. It can also be found in epidermoid tumors, meningiomas, chordomas, chondrosarcomas.

Computed tomography (CT-scan) is used to display tumor masses located in the parasellar area or, cerebellopontine angle, with extension to the petrous apex. CT-scan is a good method to differentiate trigeminal schwannoma and acoustic neurinoma. When using CT-scan, erosion of the internal auditory meatus is most often the only difference identifying acoustic neurinoma and trigeminal schwannoma. After contrast injection the neurinomas commonly enhance homogeneously, however cystic component is not uncommon<sup>17</sup>.

Magnetic resonance imaging (MRI) is the diagnostic method of choice for trigeminal schwannoma. Using MRI, one can observe the extensions of schwannoma following the normal anatomical pathways of the fifth cranial nerve and its divisions. At T1-weighted images, schwannomas are isointenses with an homogeneous enhancement after gadolinium injection. The T2-weighted images displays a hyperintensity mass. Using either T1 or T2 weighted images, relationships within the cavernous sinus and internal carotid artery can be clearly defined<sup>22, 30</sup>.

Cerebral angiography is rarely performed nowadays for trigeminal schwannoma diagnosis. The angiographic findings include a medial displacement of the carotid siphon, displacement of arterial structures and a low prominent tumor blush<sup>27</sup>.

The growth patterns and the locations of these tumors offer the surgeon a variety of approaches for their resection<sup>24, 16</sup>. These tumors can be intracranial intradural; intracranial extradural; a combination of intracranial intradural and extradural; and extracranial. The surgical approach must be tailored for each case.

The authors present a modified classification for these tumors based upon the anatomy of the fifth cranial nerve. The surgical approach best suited for each type of trigeminal schwannoma is suggested.

## Modified Jefferson's classification

In 1953, Jefferson classified trigeminal schwannomas into three types based on his study of 32 patients. In this

classification, type 1,2 and 3 tumors typically arise from the root and or the ganglion of the trigeminal nerve<sup>9, 11, 23</sup>.

Trigeminal schwannoma can also arise from any of the three divisions of the fifth cranial nerve. Based on this, two new types should be added to Jefferson's Classification. Type 4 includes tumors arising on V1 and extending into the orbit. Type 5 includes tumors arising from either or both V2 and V3 and extending into the infratemporal fossa. Either type 3 or type 5 can have a dumbbell shape (Figure 1 and 3).

Therefore each type of trigeminal schwannoma requires a different surgical approach.

**Table 1**  
**Trigeminal Schwannoma – Classification**

|        |  |
|--------|--|
| Type 1 | Tumors located mainly in the middle fossa                |
| Type 2 | Tumors located mainly in the posterior fossa             |
| Type 3 | Tumors extending in the posterior and middle fossa       |
| Type 4 | Tumors with extension to the orbit (V1)                  |
| Type 5 | Tumors located in the infratemporal fossa (V2 and or V3) |

### *Type 1 trigeminal schwannoma*

These tumors are located mainly within the middle fossa. They arise from the ganglion or from the intracranial divisions of the trigeminal nerve. These tumors are totally extradural and are located in the lateral wall of the cavernous sinus. True invasion of the cavernous sinus is not very common. A thin inner wall of the lateral wall of the cavernous sinus generally prevents invasion of the sinus. The internal carotid artery (ICA) is typically displaced medially and it is not invaded by the tumor. In rare cases, true invasion of the cavernous sinus does occur and the lateral wall of the cavernous sinus becomes a clear cleavage plane between the tumor and the ICA.

### *Type 2 trigeminal schwannoma*

These tumors are located in the posterior fossa arising from the root of the trigeminal nerve. They are located within the subarachnoid space and a clear arachnoid plane is usually found between the tumor and the surrounding cranial nerves and brain stem (Case 2).

### *Type 3 trigeminal schwannoma*

These type generally arise from the gasserian ganglion and can extend from the middle to the posterior fossa. They are considered to be extradural within the middle fossa an intradural within the posterior cranial fossa (Case 3).

### *Type 4 trigeminal schwannoma*

These tumors are found arising from the V1 division and extending along its course through the superior orbital fissure into the orbit.

### *Type 5 trigeminal schwannoma*

Usually originate from the V2 and/or V3 divisions of the fifth cranial nerve and extend into the infratemporal fossa with variable extension to the neck. Generally this type of tumor presents a clear dissection plane with the adjacent neurovascular structures of the neck (Case 1).

## Surgical treatment

The only curative treatment of trigeminal schwannoma is radical total removal. Surgical mortality has decreased significantly during the last three decades<sup>5,7,23</sup>. First, because of the development of microsurgical techniques and second, because the introduction of effective skull base approaches<sup>25,26</sup>. These two factors associated with a better knowledge of the skull base anatomy make the surgery a safe and efficient treatment.

Each type of trigeminal schwannoma requires a different skull base approach (Table 2).

**Table 2**  
**Trigeminal Schwannoma – Surgical approaches**

|        |  |
|--------|--|
| Type 1 | Zygomatic-extended middle fossa approach |
| Type 2 | Standard retrosigmoid approach           |
| Type 3 | Petrosal approach                        |
| Type 4 | Supraorbital-pterional approach          |
| Type 5 | Zygomatic infratemporal fossa approach   |

### *Zygomatic-extended middle fossa approach for type 1 schwannomas*

The “zygomatic” approach is ideally designed to reach tumors that invade the cavernous sinus extradurally. In some cases, we use this approach to remove minor tumor extensions into the posterior fossa. For the most part, we are currently using this approach to treat trigeminal schwannomas limited to the middle fossa and cavernous sinus. This approach can also be used as an alternative to the classical subtemporal approach to avoid severe brain retraction. The surgical technique is simple and easily performed. The limits of this approach for extradural lesions are: inferiorly, the height of the jugular bulb; anteriorly, the superior orbital

fissure; and superiorly, the superior petrosal sinus. Intradural access can be obtained by opening the dura of the temporal fossa or by coagulating and cutting the superior petrosal sinus and tentorium.

The patient is placed supine with the ipsilateral shoulder elevated and the head turned to the opposite side. A curvilinear preauricular skin incision is made, extending behind the hairline from the anterior border of the tragus to the superior temporal line. In some cases, the skin incision may be extended more inferiorly. A subfascial dissection is performed to preserve the frontal branches of the facial nerve. The zygomatic arch is cut at the frontal and temporal processes and displaced downward; the inserted masseter muscle is held in place at the inferior face of the zygomatic arch. We prefer to displace the temporalis muscle inferiorly, to allow us to rotate the vascularized muscle caudally to close the cavity. The displacement of the zygoma provides the visualization of the origin of the temporal muscle on the coronoid process of the mandible. A small cranio-orbital or low temporal craniotomy is performed. Approaches to the middle fossa are extradural.

The foramen spinosum is the most constant landmark in the middle fossa. Surgical resection of tumors in the middle fossa follows the middle meningeal artery medially toward the base of the petrous bone. The foramen ovale and the greater superficial petrosal nerve (GSPN) are identified. The intrapetrous segment of the ICA is commonly located inferomedial to the GSPN and the tumor is resected. For tumors located in the cavernous sinus, an extradural route should be used. The outer dural layer of the lateral wall of the cavernous sinus is carefully dissected, starting over the third division of the V nerve and extending anteriorly and cranially, exposing the second division and the inferior border of the first division of the fifth cranial nerve. A wide extradural corridor is created from the superior orbital fissure to the V2-V3 complex. Tumors extending into the infratemporal fossa, the sphenoid sinus, the pterygopalatine fossa, and the petrous apex can also easily be followed and resected<sup>6, 8, 18, 24, 26, 29</sup>.

### *Standard retrosigmoid approach for type 2 schwannomas*

This approach is indicated because type 2 trigeminal schwannomas are primarily located in the posterior fossa with minimal or no extension into the middle fossa. Patients are placed in semi-sitting position, a linear skin incision is located 3 cm behind the external auditory meatus. A posterior fossa craniotomy is performed with its lateral extension exposing the posterior border of the sigmoid sinus. The superior limit of this craniotomy should expose the inferior border of the transverse sinus.

We do advocate the use of a craniotomy instead of a craniectomy on the posterior fossa because replacement of the bone flap will provide better protection for the posterior fossa structures and a better cosmetic result<sup>12</sup>. Additionally, bone flap replacement will make a second approach easier in case of tumor recurrence<sup>12</sup>. The duramater is opened following the border of the sigmoid and transverse sinuses. After the CSF has been released from the lateral medullo-pontine cisterns, the cerebellum is dissected along its supero-lateral angle to expose the tumor. The petrosal vein is commonly identified and preserved. The tumor is first debulked and removed in a piecemeal fashion. After that, the tumor is carefully dissected from the arachnoid, taking care to preserve the adjacent neurovascular structures. These tumors can be totally removed without additional neurological deficits. The preservation of the fifth cranial nerve depends of the degree of tumor invasion. Anatomical and physiological preservation is possible, however, it should not preclude the total removal of the mass. The motor branch of the trigeminal nerve should be preserved in all cases.

### *Petrosal approach for type 3 Schwannomas*

The petrosal approach is indicated for lesions located in the posterior fossa and extending into the middle fossa. The limits of this approach are bounded by: the jugular bulb inferiorly; the clivus anteriorly; and the brain stem posteriorly. The size of the mastoid process is inversely related with the advantages of the petrosal approach. In cases where the mastoid is very well developed, the removal of it and the posterior displacement of the sigmoid sinus provide a wide surgical corridor. In cases which the hearing is absent or useless, the petrosal approach can be combined with the transtemporal or translabyrinthine approaches to create an even wider surgical corridor.

The patient is placed supine with the head at the foot-end of the operating table. The table is flexed 20° to 30° to allow elevation of the head and trunk. The patient's ipsilateral shoulder is slightly elevated. The head is turned to the opposite side, and tilted toward the floor. Special care should be taken to avoid compression of the contralateral jugular vein. During the surgical procedure, the table can be rotated from side to side or up and down. A reverse question-mark incision is made starting at the zygoma in front of the ear, circling 3 cm above the ear and descending 2 cm behind the mastoid process. The skin flap is elevated and retracted anteriorly and inferiorly. Fascia of the temporal muscle is dissected from the muscle and displaced posteriorly and inferiorly contiguous with the suboccipital muscles that insert in the superior and

inferior nuchal line. The posterior half of the temporal muscle is dissected from the temporal squama and displaced anteriorly and inferiorly. At this stage in the procedure, the posterior end of the zygomatic arch, the temporal and the posterior fossa can be identified.

The bony work starts with the demarcation of the burr holes. Essentially four burr holes are drilled. Two superior and two inferior to the transverse-sigmoid sinus. The first burr hole is made just under the asterion; the second is performed 2 cm posteriorly to the former; both holes will open into posterior fossa and under the transverse-sigmoid sinuses. The third burr hole is placed in the most anterior point of the mastoid suture and will open in the middle fossa. The last burr hole is placed 2 cm superior to the third and will also open into middle fossa. The posterior fossa craniotomy is performed by connecting the first and the second burr hole with a craniotome. A bone cut in the posterior fossa is made in a curvilinear fashion and as caudal as possible. The middle fossa craniotomy is performed connecting the third and the fourth burr hole. The bone cut in the temporal fossa should be made as anterior and basal as possible in order to provide a flat surface with the base of the temporal fossa. Because the high risk of damage to the venous sinuses, the craniotome should never be used for cutting over the area of transverse-sigmoid sinuses. For safety, we recommend to connect the first and third burr holes and the second and fourth burr holes using a thin rongeur or a high-speed drill with a diamond cutting burr. Commonly severe dural adherence occurs at the junction of the transverse and sigmoid sinus requiring very careful dissection while elevating the bone flap. The single bone flap is elevated, exposing the transverse and sigmoid sinus and the dura of both the middle and the posterior fossa. In cases when a mastoidectomy is necessary a high-speed drill is used in conjunction with adequate irrigation. Initially, a cutting bit can be used, but once the dissection nears important vascular or bony structures a diamond drill should be used.

Dissection of the temporal bone is performed in a sequence of steps. The first step is the localization and skeletonization of the sigmoid sinus, sinodural angle of Citelli and the jugular bulb. The step two is identification of the bony structures of the superior, posterior, and lateral semicircular canals. Usually a large mastoid cell called the antrum orientates toward the elements of the inner ear. During this dissection a diamond bit and proper irrigation must be used to avoid inadvertent damage. The step three consists in a careful removal of the thin layer of bone left over the sigmoid and the superior petrosal sinuses (located in the sinodural angle of Citelli). Now the dura located anteriorly to the sigmoid sinus and in the basal aspect of the middle fossa is visible. In the posterior fossa, a dural opening is performed along the posterior margin

of the sigmoid sinus. In the middle fossa, the dural incision extends from the anterior border of the sigmoid sinus and along the floor of the middle fossa. At this point special care should be taken to avoid damage to the vein of Labbé, which enters the dural sinus at the level of the transverse-sigmoid junction.

The next step is the cutting and coagulation of the superior petrosal sinus and tentorium. Before the tentorial incision opening, CSF is released from the lateral medullary cisterns for brain relaxation. The dural incision is continued on the tentorium parallel to the pyramid and extended through the incisura. Before the border of the tentorium be incised, the IV cranial nerve must be identified and preserved. The posterior leaf of the tentorium is elevated and fixed superiorly by the brain spatula, a maneuver that allows the enlargement of the surgical field and the protection of the vein of Labbé. At this point in the procedure, the third cranial nerve, posterior cerebral artery and superior cerebellar artery can be easily identified. The tumor is debulked and carefully removed to protect surrounding structures. After the removal, the dura-mater is closed watertight. The remaining cavity resulted from the bone removal is covered by the posterior third of the temporal muscle which is rotated inferiorly and fixed to the suboccipital muscles. The temporal muscle fascia, displaced inferiorly fixed to the suboccipital muscle is anchored anteriorly at its original place. The skin flap is closed as usually<sup>3</sup>.

#### *Supraorbital-pterional approach for type 4 schwannomas*

The orbito-cranial approach has been used in the treatment of cavernous sinus, parasellar, petrous apical and also orbital lesions. With the orbital roof removal one can approach easily and safely type 4 trigeminal schwannomas.

The scalp flap is turned after a bicoronal incision is made behind the hairline. The incision extends from the level of the zygomatic arch on the side of the tumor to as far as the superior temporal line on the other side. Every effort is made to preserve the superficial temporal artery and the frontalis branches of the facial nerve. The temporalis muscle is detached from its insertion anteriorly to as far down as the zygomatic arc; the muscle then is retracted posteriorly and inferiorly, exposing the junction of the zygomatic, sphenoidal and frontal bones. The periosteum of the frontal bone is incised posteriorly, dissected forward, and reflected over the anteriorly scalp turned flap. The intact base of this periosteum, in continuation with the periorbita, is dissected free from the margin of the roof and lateral wall of the orbit. The supraorbital nerve is freed from the supraorbital notch by drilling around the notch with a high speed air drill.

Three burr holes are drilled. The first is made in the frontal bone above the nasion. This hole should be kept as small as possible for cosmetic reasons. The second burr hole (MacCarty keyhole) is made in the temporal fossa at the frontosphenoidal junction just behind the zygomatic process of the frontal bone. The upper half of this burr hole will expose the dura mater, while the lower half exposes the periorbita, the two membranes being separated by the roof of the orbit. The third burr hole is made posteriorly near the floor of the temporal fossa. The bone between burr holes is severed. The paramedian frontal burr hole and the posterior temporal burr hole are connected by the craniotome; its blade passes through the frontal and temporal bone about 5 cm above the supraorbital rim. The posterior temporal burr hole and the keyhole are likewise connected with the craniotome passing just above the floor of the temporal fossa. Then the paramedian frontal burr hole and the keyhole are interconnected. The orbital roof can be cut with Gigli saw or chisel. The contents of the orbit are protected with a brain spatula during this process. Particular attention should be given to keeping the periorbita intact. The craniotomy flap is freed by fracturing it loose at the sphenoid base. The removed and preserved bone flap thus includes the superior and lateral orbital rim, the anterior portion of orbital roof, and the adjacent frontal and temporal bones. This approach provides excellent wide exposure of the middle fossa and orbit<sup>1,2,4,14</sup>.

#### *Zygomatic infratemporal fossa approach for type 5 schwannomas*

This approach is a modification of the zygomatic-infratemporal fossa approach and is indicated to reach tumors with inferior extracranial extension to the infratemporal fossa. The skin incision extends inferiorly just anteriorly to the tragus and running in the anterior border of the sternocleidomastoid muscle. The wider incision allows the anterior mobilization of the skin flap and better exposure of the infratemporal fossa. The tumor can be removed following caudally the nerves through the skull base foramen or superiorly through the natural routes of the neck<sup>6,8,18,24,26,29</sup>.

## **Illustrative cases**

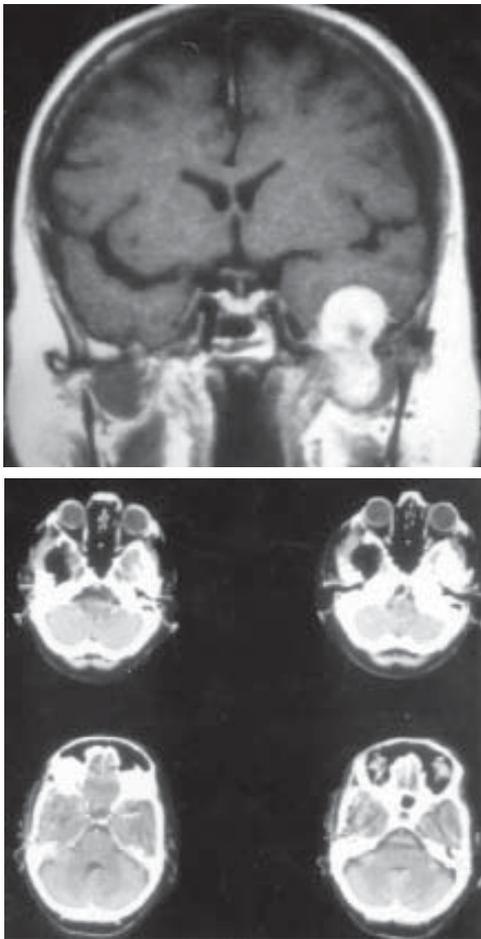
Three patients with different types of trigeminal schwannoma were recently admitted and treated at the Evangelic University Hospital of Curitiba, Parana, Brazil.

*Case 1*

A 55-year-old female was admitted with a 3 months history of right peripheral facial palsy associated a tinniuts. The neurological examination showed a right facial palsy and hyposthesia in the area of the trigeminal nerve. The corneal reflex was present. The patient denied facial pain or diplopia.

A MRI depicted a tumor mass located in the base of the middle cranial fossa with extension to the infratemporal fossa (Figure 1).

This patient underwent a surgical removal through a zygomatic-extended middle fossa approach. The tumor was totally extradural, arising from the third division of the trigeminal nerve. The geniculate ganglion, exposed in the middle fossa was compressed by the tumor. Total removal was achieved and the tumor cavity obliterated with fat. The postoperative course



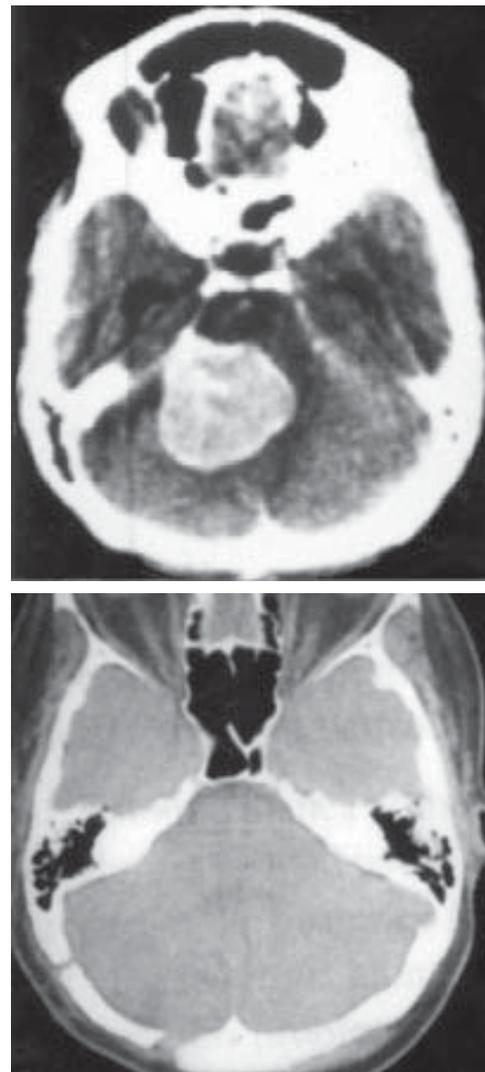
**Figure 1 (Case 1/Type 5) – Upper – preoperative MRI showing a type 5 trigeminal schwannoma. The tumor extends from the middle to infratemporal fossa. Lower – postoperative CT-scan showing the gross total removal of the lesion through a infratemporal zygomatic approach.**

was uneventful except by the persistent of preoperative deficits. A 6 months follow-up indicated an improvement of the facial palsy. Postoperative CT-scan showed gross total removal of the lesion (Figure 1).

*Case 2*

A 45 year-old female was admitted with a 6 months history of facial pain, right hearing loss and headache. Her neurological examination was marked by the presence of hyposthesia in the right hemiface and hearing impairment.

A CT-scan depicted the tumor located in the posterior fossa with attachment to the petrous ridge and porus trigeminus (Figure 2).



**Figure 2 (Case/Type 2) – Upper – preoperative CT-scan showing a type 2 of trigeminal schwannoma. The lesion is limited within the posterior fossa. Lower – postoperative CT-scan showing the tumor removal and the bone flap in place after a retrosigmoidal posterior fossa craniotomy.**

A standard retrosigmoidal surgical approach was performed with the patient placed in a sitting position. Tumor removal was achieved with the fifth cranial nerve partially preserved. The post-operative course was unremarkable and a follow-up CT-scan demonstrated total tumor removal (Figure 2).

### Case 3

A 26-year-old male was admitted with a 2 months history of facial pain and diplopia. The neurological examination was normal with exception of a slight deficit of the right sixth cranial nerve.

MRI depicted a large mass invading the posterior compartment of the cavernous sinus within the middle fossa, and extending posteriorly across the petrous apex and into the posterior fossa (Figure 3).

A petrosal approach was used to remove the tumor. In the posterior fossa, the tumor was intradural and it

extended anteriorly through porus trigeminus intradurally into the middle fossa and posterior compartment of the cavernous sinus. At a one month follow-up the patient was clear of the 6th nerve palsy and a CT scan confirmed complete removal of the tumor mass (Figure 3). Interestingly a similar case in regard of clinical manifestation has been reported in the literature<sup>28</sup>.

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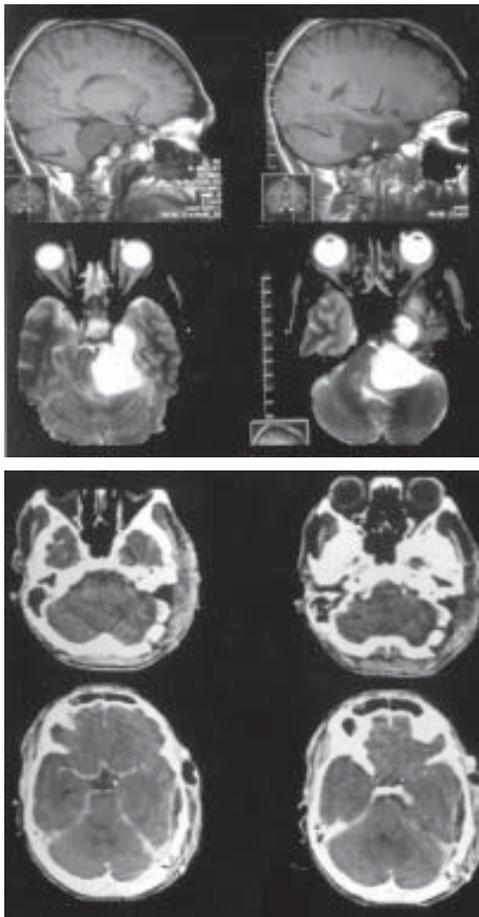


Figure 3 (Case 3/Type 3) – Upper – MR images showing the lesion within two compartments: middle and posterior fossa. Lower – postoperative CT-scan showing total tumor removal through a petrosal approach.

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