# The Surgical Management of Trigeminal Schwannomas

# 21

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## Contents



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## 21.1 Introduction

Schwannomas arising from the trigeminal nerves are the second most common type of intracranial schwannomas, representing 0.8–8 % of all intracranial schwannomas [5, 34, 42, 52]. These tumors are usually benign, isolated, and slow growing. They may occur in multiple sites when associated with neurofibromatosis type 2 (NF2). Trigeminal schwannomas (TSs) tend to occur in middle-aged patient; the highest incidences are between the ages of 38 and 40 years and are more common in women [28, 29, 51]. Patients with TSs frequently complain of trigeminal nerve-related symptoms, but they may also be asymptomatic. In a recent review of the literature, Samii et al. [47] reported trigeminal nerve symptoms in 51 % of patients, followed by headache (16 %) and diplopia (11 %). Prior to the advent of computed tomography (CT) and magnetic resonance (MR) imaging, diagnosis of TS was difficult. The treatment of choice remains resection. In 1918, Frazier [16] reported the first removal of TS. Very high mortality has been associated with surgical treatment prior to the late 1950s [49]. Since then, in addition of a large series of 111 patients [29], several small series [9, 13, 17, 18, 32, 34, 44, 47] have demonstrated positive results and low mortality and morbidity rates associated with complete resection. Different surgical approaches may be used depending on the location and extension of the lesion.

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## 21.2 Surgical Anatomy

Recent advances in understanding the microsurgical anatomy of skull base structures are hallmarks of modern neurosurgery. Findings reported in the literature have proven, specifically in trigeminal nerve schwannomas, that a precise 3D knowledge of the dural, venous, and nerve anatomy of the cavernous sinus acquired through dissections of laboratory specimens is mandatory [11, 14, 48]. Moreover, clinical surgical experience in resecting tumors within such a complex region is necessary to remove the maximum amount of the tumor safely [12, 15].

The trigeminal nerve emerges from the ventrolateral surface of the pons and runs anteriorly ~1–2 cm [6] through the cerebellopontine cistern to reach the petrous apex. Vascular structures, such as the petrosal vein and the superior cerebellar artery, lie close to the trigeminal nerve. Over the petrous apex, ~7 mm of distance from the medial lip of the internal acoustic meatus [19], the Gasserian ganglion is enveloped by a dural deflection forming the Meckel's cave, laterally to the cavernous sinus and carotid artery. As it leaves the Meckel's cave, the trigeminal nerve is divided into 3 branches: the ophthalmic (V1), maxillary (V2), and mandibular (V3) branches. These three nerves run under the medial fossa dura and leave the temporal bone through the lateral wall of the cavernous sinus (for V1), foramen rotundum (for V2), and foramen ovale (for V3).

The trigeminal nerve can also be surgically classified into three segments: cisternal (from the brainstem to the petrous apex), intracranial-extradural (from Meckel's cave to the foramina), and extracranial (V1, V2, and V3).

Functionally, the trigeminal nerve has two portions: the "pars compacta," which constitutes the triangular portion and comprises the primary afferent fibers that are responsible for the special sensibility of the face, and the motor root, which carries the branchiomotor fibers to the muscles of mastication. The motor root runs practically separated from the "pars compacta" but together with the cranial portion of the nerve. At the level of the Meckel's cave, it is oriented medially and leaves the skull together with the maxillary nerve.

The intracranial-extradural portions of V2 and V3 are surgically identified using the *foramen spinosum* as an anatomical landmark, which is located at the sphenoid bone and contains the middle meningeal artery. The foramina ovale and rotundum are located ~2–5 mm superoanteriorly and 10–12 mm superomedially to the foramen spinosum, respectively [31].

## 21.3 Classification of Tumor Extension

TSs may originate from the root, the ganglion, or the peripheral branches of the trigeminal nerve. Jefferson [26] initially divided these tumors into four groups depending on their anatomical location: posterior fossa (root type), combined posterior fossa-middle fossa (dumbbell type), middle fossa (ganglion type), and peripheral (division type). Samii et al. [47] classified the tumor extension based on radiological findings: Type A, intracranial tumor predominantly in the middle fossa; Type B, intracranial tumor predominantly in the posterior fossa; Type C, intracranial dumbbell-shaped tumor in the middle and posterior fossa; and Type D, extracranial tumor with intracranial extensions.

In our institution, we prefer to use the classification of Samii et al. [47], with the following modifications [44]: Type A, predominantly an extracranial tumor with small extension in the middle fossa (Fig. 21.1); Type B, an intracranial tumor predominantly in the middle fossa with extracranial extension (Fig. 21.2); Type C, a tumor in the middle fossa (Fig. 21.3); Type D, a tumor in the posterior fossa (Fig. 21.4); Type E, a tumor with middle and posterior fossa extensions (Fig. 21.5); and Type F, a tumor with extracranial, middle, and posterior fossa extensions (Fig. 21.6).

In our opinion, these modifications are important to assure the best surgical approach for each type of lesion. In the experience of the senior author (R.R.), each subgroup of this modified classification presents a different surgical challenge. Tumor types are ordered according to the level of difficulty as A, C, D, B, E, and F. The

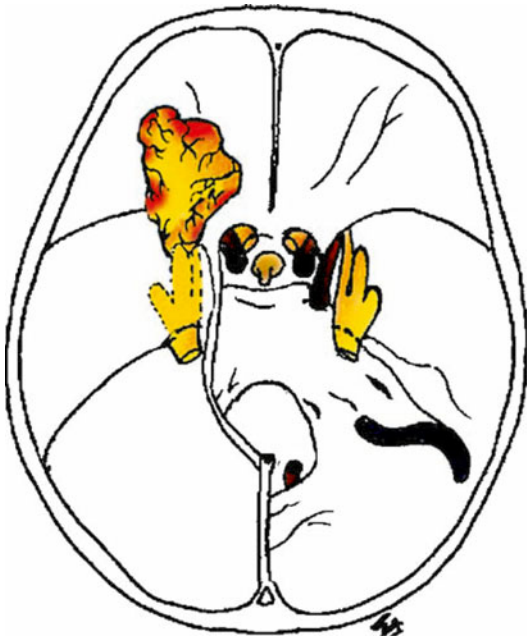

**Fig. 21.1** Large extracranial trigeminal schwannoma with small middle fossa extension (type A)

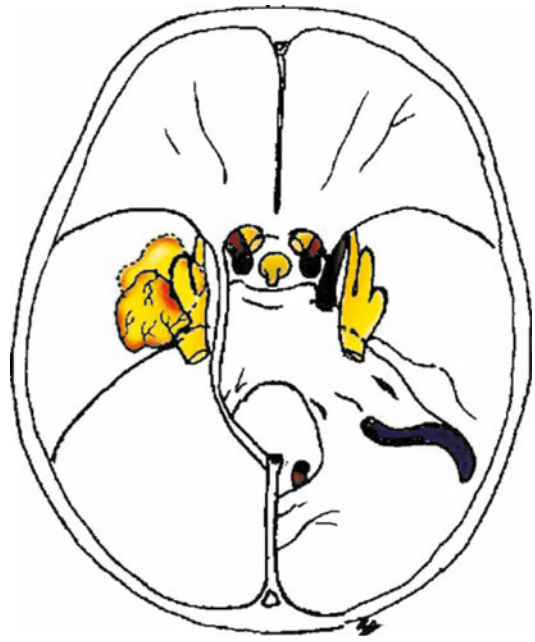

**Fig. 21.3** Middle fossa trigeminal schwannoma (type C)

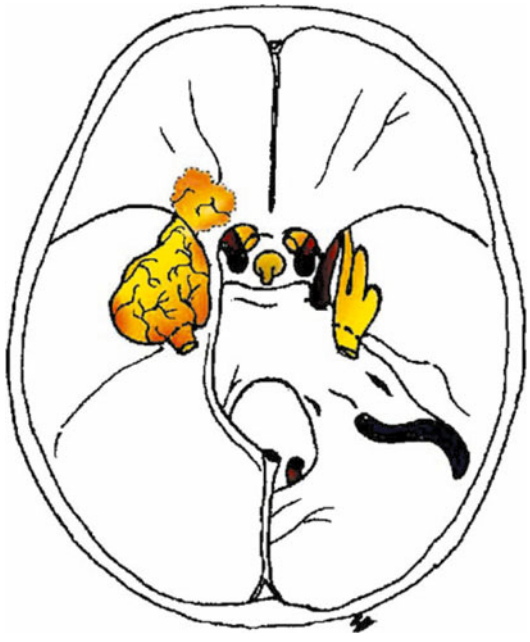

**Fig. 21.2** Drawing of trigeminal schwannoma with main portion in the middle fossa and small extracranial extension (type B)

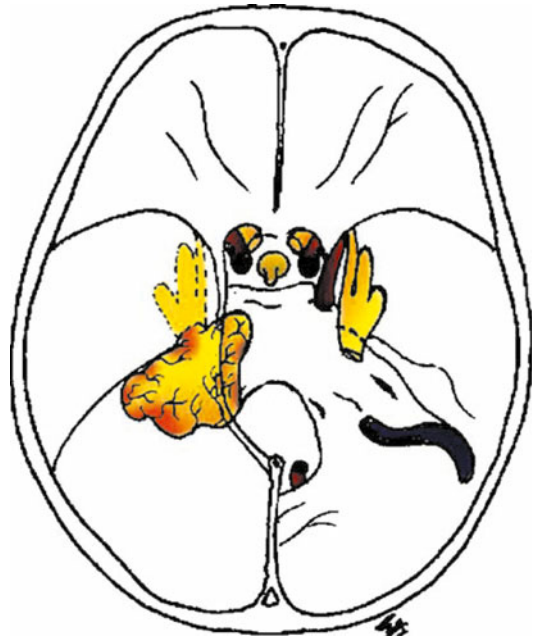

**Fig. 21.4** Schematic drawing showing a posterior fossa trigeminal schwannoma (type D)

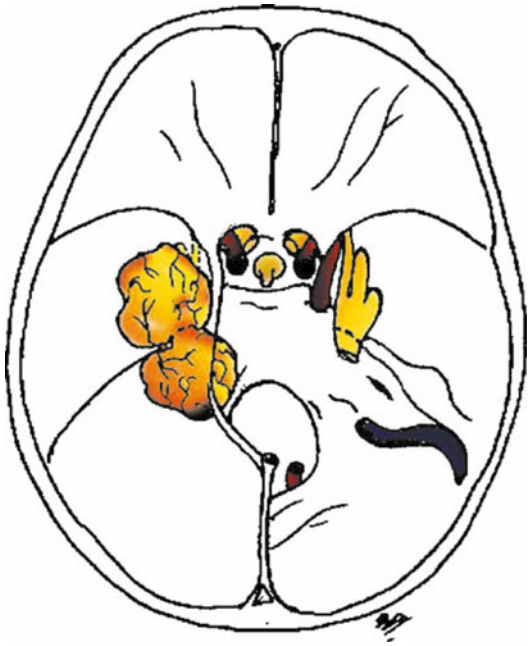

**Fig. 21.5** Drawing of trigeminal schwannoma with middle and posterior fossa extensions (type E)

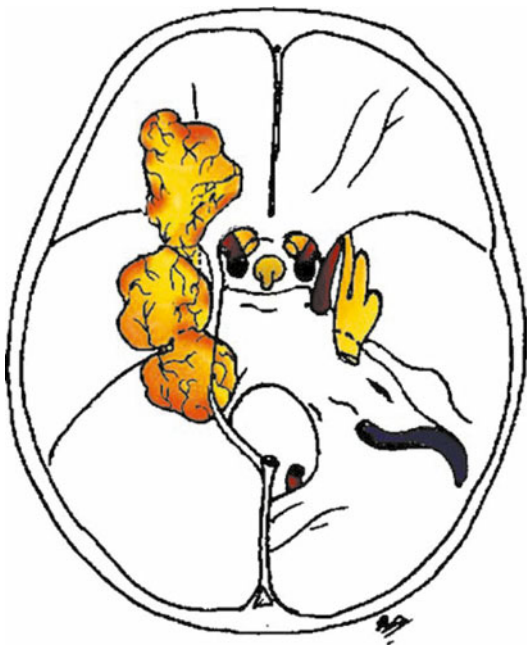

**Fig. 21.6** Schematic drawing showing a trigeminal schwannoma with extracranial, middle and posterior fossa extensions (type F)

**Table 21.1** Types of tumors and surgical approaches in 21 patients with TSs

| Level of difficulty | Type of tumor | No. of patients (%) | Approach |
|---|---|---|---|
| Easiest | A | 3 (14.3) | Extradural (TM or TM + EMF) |
| | B | 2 (9.5) | Intradural (IMF) |
| | C | 8 (38.2) | IMF or EMF |
| | D | 2 (9.5) | Retrosigmoid |
| | E | 4 (19) | Presigmoid or 2-staged retrosigmoid/ middle fossa |
| Hardest | F | 2 (9.5) | Combination of all of the above |

*EMF* extradural middle fossa, *IMF* intradural middle fossa, *TM* transmaxillary

degree of difficulty in radical surgical removal depends on the particularities of each case, but according to this modified classification, Type A tumors have the lowest level of difficulty and Type F tumors, on the contrary, the highest.

## 21.4 Clinical Material

This series includes 21 patients who were surgically treated between 1987 and 2012 at our institution. Two patients received a diagnosis of NF2. One patient presented with a posterior communicating artery aneurysm on the same side as the TS. There were 3 Type A, 2 Type B, 8 Type C, 2 Type D, 4 Type E, and 2 Type F tumors (Table 21.1). The cavernous sinus was involved in 16 cases. The sizes of the lesions were small (<3 cm) in 5, medium (between 3 and 4 cm) in 6, large (>4 cm) in 6, and giant (> 5 cm) in 4 cases.

The clinical symptoms in these patients included trigeminal hypesthesia (10 patients), facial pain (9 patients), headaches (6 patients), hearing impairment (3 patients), seizures (3 patients), diplopia (4 patients), ataxia (2 patients), exophthalmos (1 patient), and hemiparesis and increased ICP with papilledema (1 patient) (Table 21.2). Prior to 1990, postoperative CT imaging studies confirmed the extent of resection.

**Table 21.2** Preoperative clinical symptoms in 21 patients with TSs

| Symptom | No. of patients (%) |
|---|---|
| Trigeminal hypesthesia | 10 (47.6) |
| Facial pain | 9 (42.9) |
| Headache | 6 (28.6) |
| Diplopia | 4 (19) |
| Seizure | 3 (14.3) |
| Hearing symptoms | 3 (14.3) |
| Ataxia | 2 (9.5) |
| Exophthalmos | 1 (4.8) |
| Hemiparesis and increased ICP with papilledema | 1 (4.8) |

**Table 21.3** Results of surgical procedures in 21 patients with TSs

| Op outcomes | No. of patients (%) |
|---|---|
| Total resection | 20 (95.2) |
| Near-total resection | 1[a] (4.8) |
| Recurrence | 2 (9.5) |
| Op-related death | 0 |

[a]Tumor adhered to the cavernous sinus in a patient with NF2

After 1990 all patients underwent pre- and postoperative CT and MR imaging studies. In our series, postoperative imaging examinations were routinely performed 3 days after surgery. Postoperatively, imaging examinations are performed every 6 months for 2 years, then once a year for 5 years, and after that every 2 years. Recurrence was defined as a new lesion identified on routine MR imaging in a patient considered to have had total resection. Progression was related to tumor growth in a patient considered to have had subtotal or partial resection, as demonstrated by immediate postoperative MR imaging. Total tumor excision was possible in 20 patients, whereas total removal was not achieved in 1 patient with NF2 due to adherences within the cavernous sinus. There were no surgery-related deaths (Table 21.3). Nine patients developed postoperative anesthesia in at least one branch of the trigeminal nerve. Trigeminal motor function was preserved in 8 patients. Two individuals developed a CSF leak and were successfully treated by lumbar CSF drainage. Only 1 of the 9 patients who reported facial pain preoperatively remained symptomatic postoperatively. Two patients exhibited minimal facial palsy that resolved during the follow-up period. The patient with preoperative hemiparesis showed progressive improvement after surgery. There were two recurrences: one in a patient with NF2, which occurred 5 years after a radical removal of a Type C tumor, and one in a patient with a Type A tumor 6 years after a radical resection. In this patient recurrence occurred in other portion of the trigeminal nerve (tumor Type C). In both cases, the recurrent tumor was removed, with an uneventful postoperative outcome.

## 21.5 Surgical Approaches

According to the aforementioned classification (Table 21.1), Type A tumors are resected via a transmaxillary (Fig. 21.7) approach (TM) or TM associated with an extradural middle fossa approach (EMF) if there is broad extension to the middle fossa. Type B lesions are removed through an intradural middle fossa approach (IMF); large extradural extensions, however, may require a transmaxillary exposure (Fig. 21.8). Type C tumors are extirpated via a middle fossa approach, either intradurally or extradurally (Figs. 21.9 and 21.10). Resection of Type D tumors is best achieved via the retrosigmoid approach. Tumors Type E are extirpated using a presigmoid approach or 2-staged retrosigmoid and middle fossa approaches [4] (Figs. 21.11 and 21.12). As stated before, Type F tumors are considered the most challenging lesions and may demand a combination of all previously described approaches (Fig. 21.13).

### 21.5.1 The Transmaxillary Approach (Caldwell-Luc Procedure)

This procedure is usually performed in association with the middle fossa approach to remove

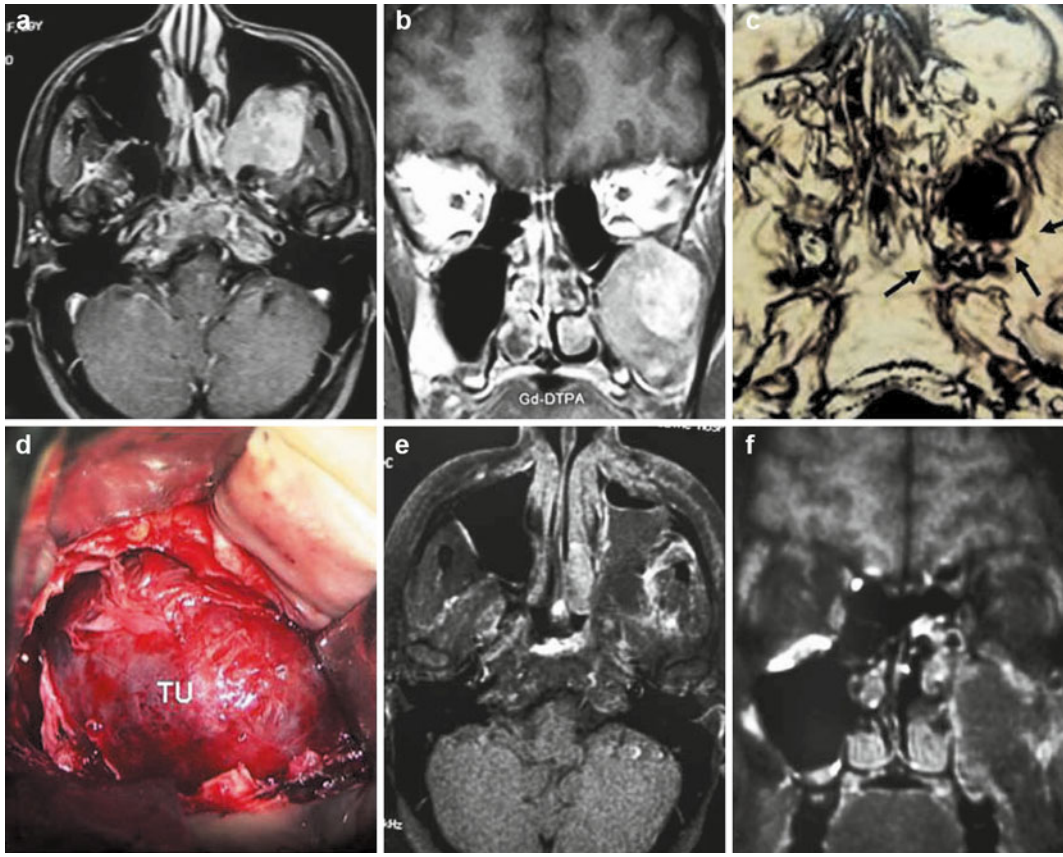


**Fig. 21.7** (**a**, **b**) Magnetic resonance imaging (MRI) scan of a large trigeminal schwannoma with main portion in the maxillary sinus (type A). *Gd-DPTA* Gadopentetate. (**c**) Computed tomography (CT) scan showing enlargement of the foramen rotundum (*arrows*). (**d**) Surgical picture: transmaxillary approach exposing the tumor (*TU*). (**e**, **f**) Postoperative MRI

large TSs. In our series, it was used to remove a large schwannoma within the maxillary sinus with small extension to the middle fossa and the cavernous sinus. After induction of general anesthesia, the gingiva is infiltrated with lidocaine 1.0 % with 1:100,000 epinephrine solution. A 3-cm incision is made over the upper canine and the upper first premolar tooth. To improve the suturing, 1 cm of gingival tissue is left intact above the dentition. The soft tissue and periosteum are elevated from the anterior wall of the maxilla. The infraorbital nerve is identified as it exits the infraorbital foramen (midpupillary line). The anterior wall of the maxillary sinus is opened. The tumor is identified and microsurgical intracapsular debulking is performed. Total removal of the maxillary, middle fossa, and cavernous sinus portions is accomplished with this approach. The incision is closed with 4–0 absorbable sutures.

### 21.5.2 The Middle Fossa Zygomatic Approach

The patient is placed supine with the head fixed in a 3-pin device and rotated ~30° to the opposite side. A C-shaped skin incision over the *pterion* is performed. The temporalis muscle fascia is incised to protect the frontal branch of the facial nerve, and the scalp-fascia flap is lifted anteriorly. The temporalis muscle is retracted inferiorly and posteriorly. A pterional craniotomy is performed and enlarged to the base of the temporal

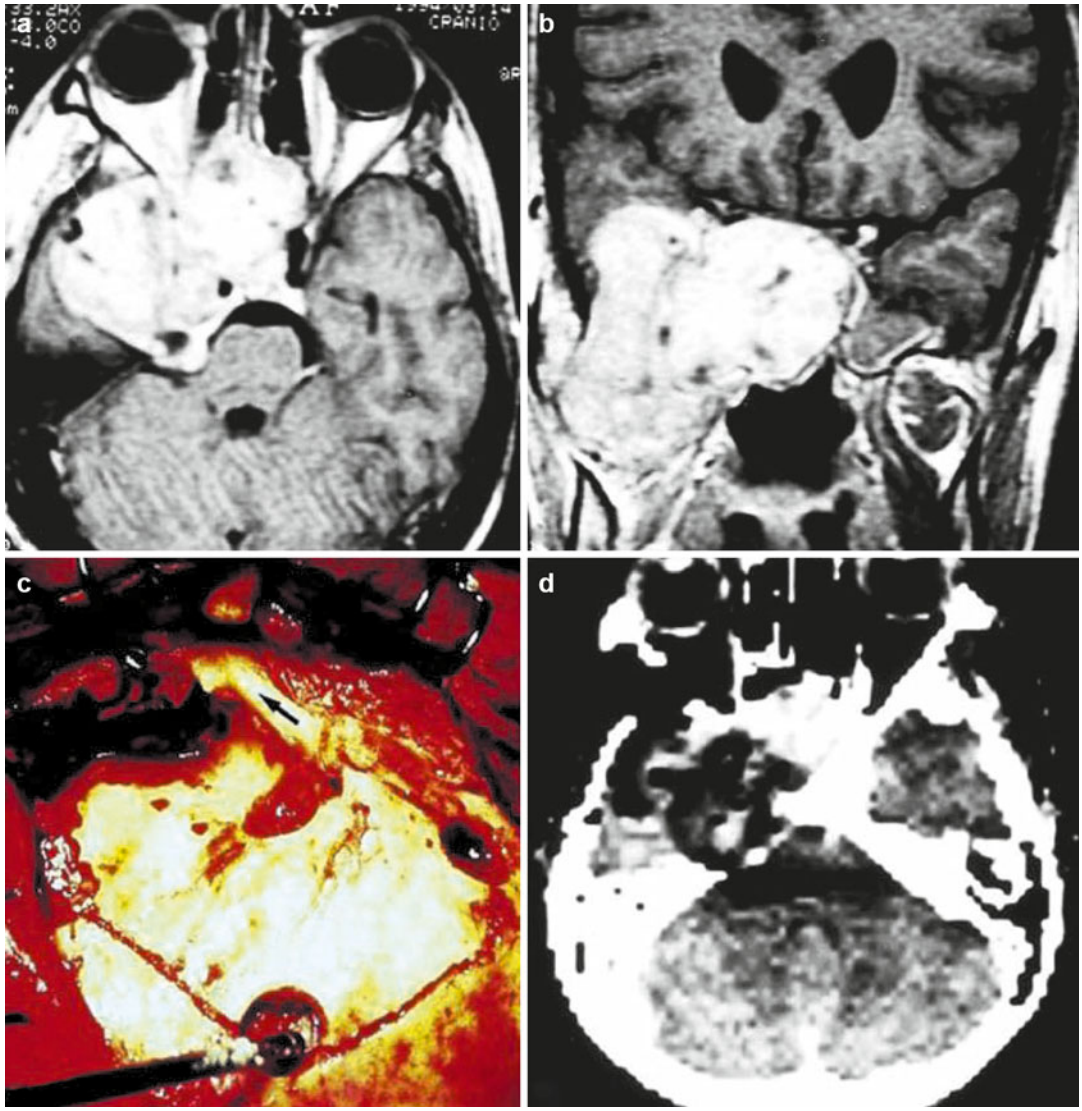


**Fig. 21.8** (**a**, **b**) MRI of a large trigeminal schwannoma with middle fossa and extracranial extension (type B). (**c**) Middle fossa craniotomy. Zygomatic arch (*arrow*). (**d**) Postoperative CT scan

fossa by removing bone with a rongeur to obtain a flat viewing angle across the floor of the middle fossa. The dura mater is then dissected and elevated from the middle fossa, exposing the superior orbital fissure. The middle meningeal artery is coagulated and transected. The second and third branches of the trigeminal nerve are identified. The foramen ovale and rotundum are exposed using a drill. The dura mater is elevated from the lateral cavernous sinus wall, revealing the tumor extradurally and the branches of the trigeminal nerve. The tumor is debulked and its capsule is dissected from the intact branches of the trigeminal nerve, which allows radical removal of the lesion. The tumor may be approached intradurally by opening the Sylvian fissure and exposing the optic nerve, the internal carotid artery, and the third cranial nerve. The lateral cavernous sinus wall is exposed via a temporopolar approach, and the tumor is removed. The

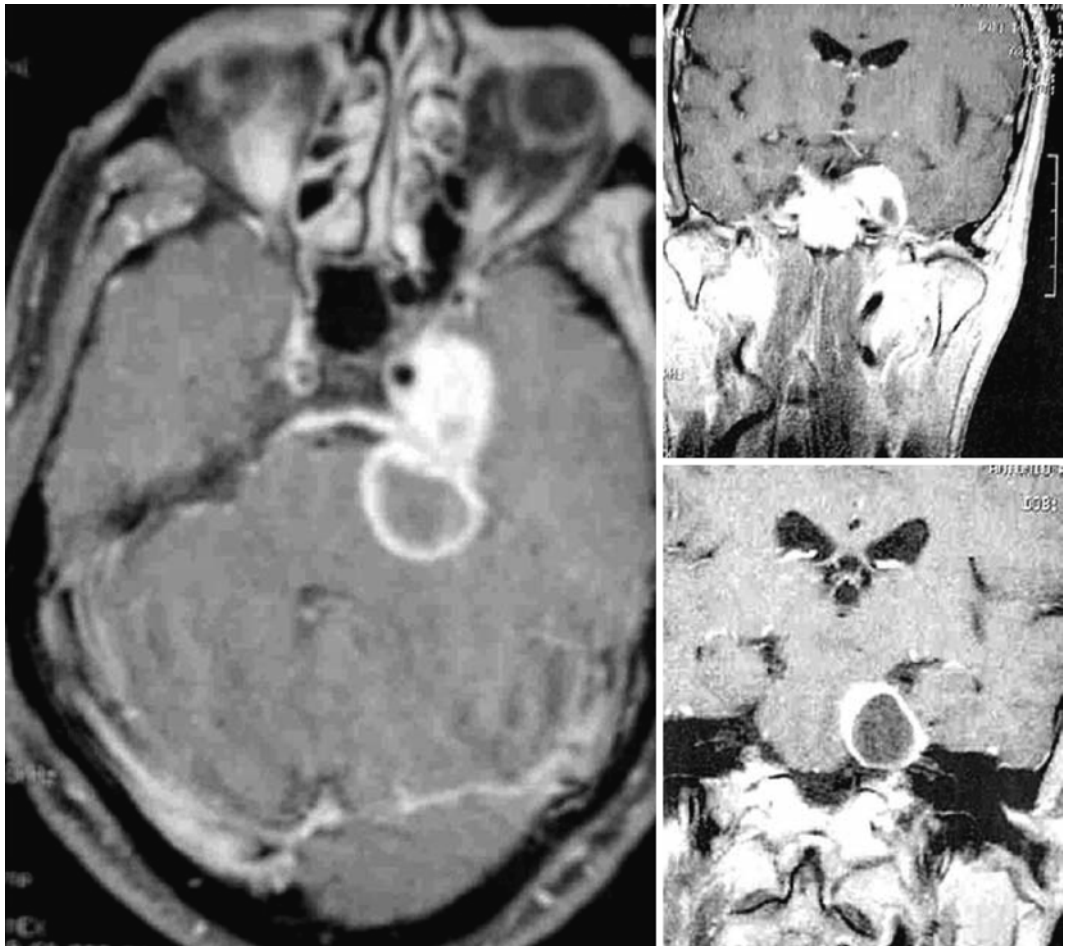

**Fig. 21.9** MRI studies showing middle fossa trigeminal schwannoma with cystic posterior fossa portion (type C)

trigeminal fibers are preserved as much as possible. The petrous apex is drilled, and the petroclinoidal ligament is cut to expose the posterior fossa portion, allowing complete excision of the remaining tumor. Neuronavigation can provide valuable intraoperative information during tumor debulking as the location of vascular structures is distorted by the tumor [7].

### 21.5.3 The Retrosigmoid Approach

This approach is performed by placing the patient in the dorsal (mastoid) position with the head turned to the opposite side and the ipsilateral shoulder elevated. A linear incision is placed 4 cm behind the external auditory canal. The asterion is exposed to determine the junction of the transverse and sigmoid sinuses. MR neuronavigations are also utilized for localization of the transverse and sigmoid sinuses, reducing risk of venous sinuses injuries [8]. A craniotomy 4 cm in diameter is performed, with the superior and anterior margins bordering the transverse and sigmoid sinuses, respectively. The dura mater is opened parallel to the sigmoid sinus, CSF is drained from the cerebellomedullary cistern, and CNs VII–XI are identified. The tumor is thereby exposed near the tentorium margin. After intracapsular tumor debulking, microsurgical radical removal is accomplished. A watertight dural closure is completed, and the bone flap is fixed in

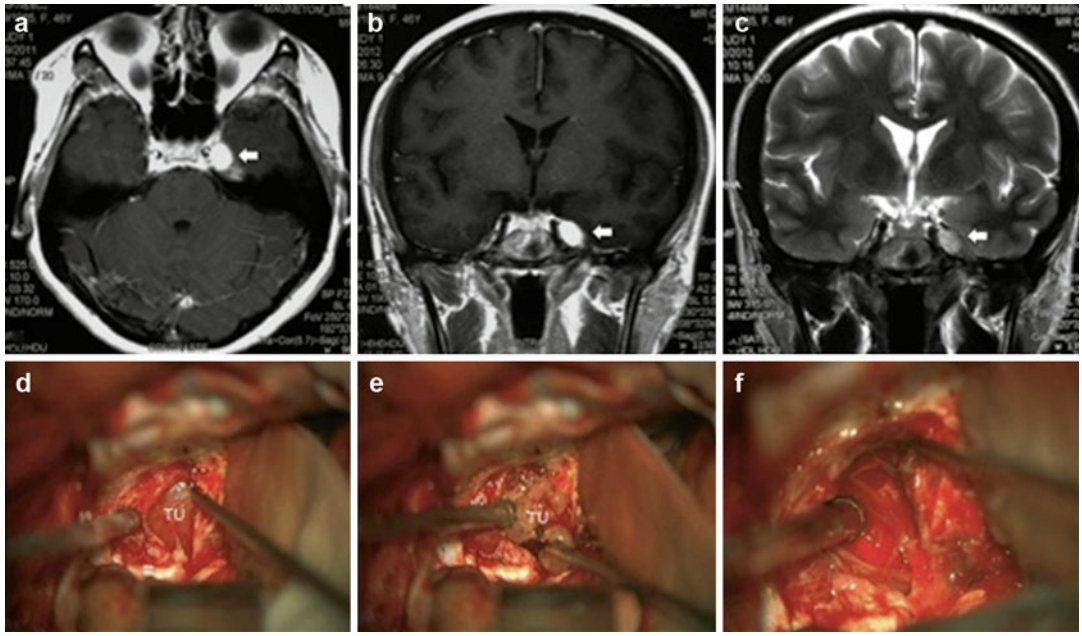


**Fig. 21.10** Axial (**a**) and coronal (**b**, **c**) MR imaging study of a Type C TS with cavernous sinus involvement (*arrows*). (**d**) Intraoperative photograph showing tumor exposure (TU) in a middle fossa intradural approach. The lateral wall of cavernous sinus was opened. (**e**) Intraoperative photograph showing dissection of the tumor capsule (TU) and preservation of trigeminal fibers after total removal (**f**)

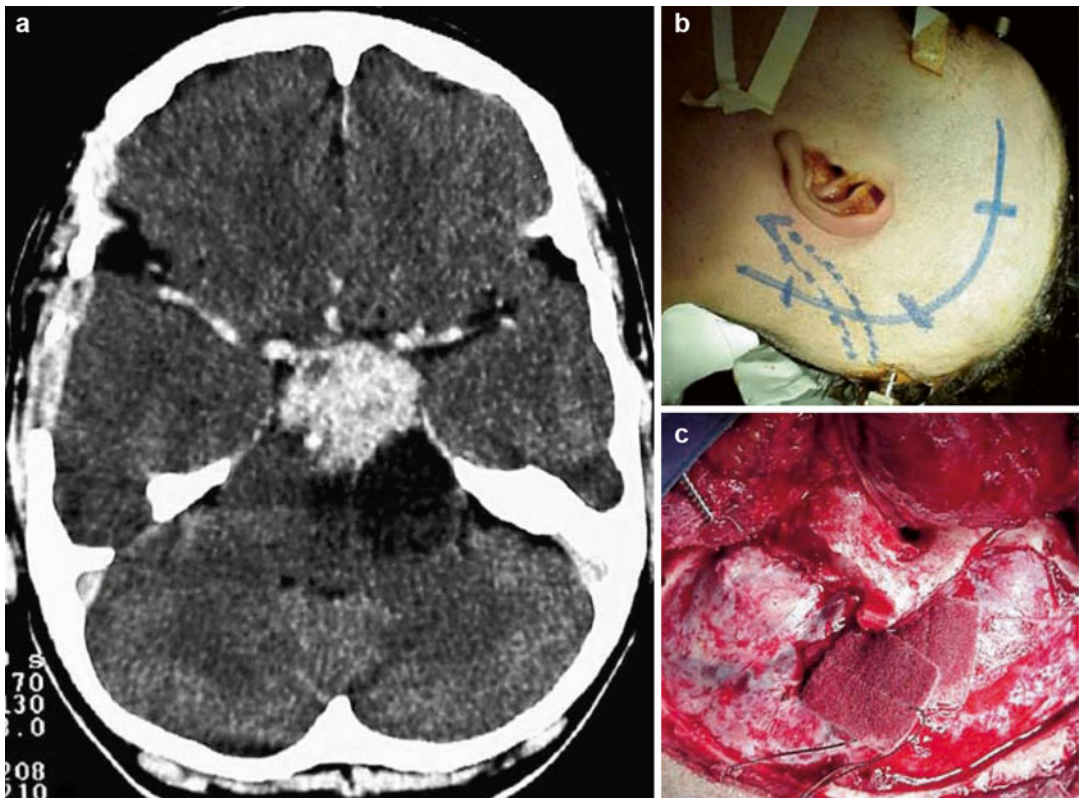


**Fig. 21.11** (**a**) Preoperative MRI showing a posterior fossa trigeminal schwannoma (type D) with cystic portion. (**b**) Skin incision. (**c**) Presigmoid craniotomy

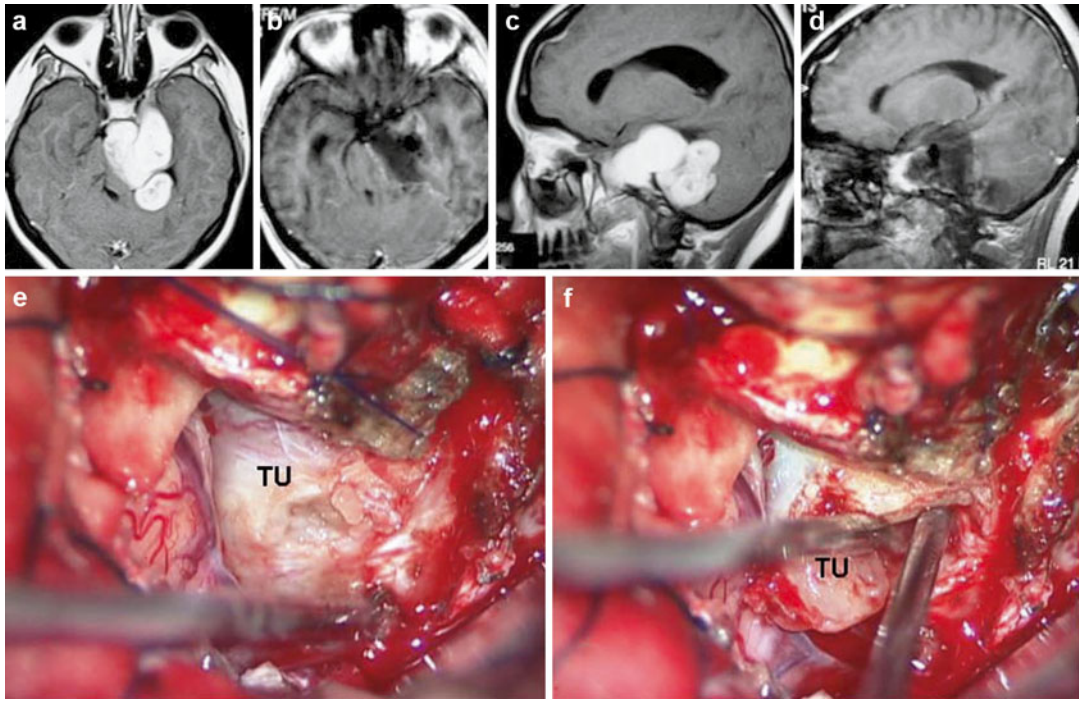


**Fig. 21.12** Pre- (**a**, **c**) and postoperative (**b**, **d**) MRI scans of a large trigeminal schwannoma (type E) with extension within the middle and posterior fossa. (**e**) Surgical picture of presigmoid approach for removal of the tumor. (**f**) Removal of the tumor capsule from the brainstem

place. All opened mastoid cells are sealed with bone wax to help prevent CSF leaks.

### 21.5.4 The Presigmoid Approach

The patient is placed in the dorsal (mastoid) position. A C-shaped skin incision from the middle fossa to the mastoid tip is performed. Two burr holes are placed anteriorly and 2 are placed posteriorly at the intersection of the transverse and sigmoid sinuses. The temporal and retromastoid dura mater is exposed through a craniotomy. Mastoidectomy with preservation of the labyrinth block and the facial nerve canal is the next surgical step. Parallel dural incisions to the middle fossa floor are made anteriorly to the sigmoid sinus. The superior petrosal sinus is then ligated and transected. At this point, the inferior temporal lobe and the lateral portion of the cerebellum are slightly retracted so that the vein of Labbé is preserved. The tumor is identified and the seventh and eighth cranial nerves are usually displaced inferiorly. The tumor within the cavernous sinus is removed after opening its lateral wall. After complete excision of the tumor, the dura mater is closed in a watertight fashion and the skull base is reconstructed with myofascial flaps [43].

## 21.6 Discussion

Although TSs are the second most common intracranial schwannoma, there is no typical clinical syndrome caused by TSs. Nevertheless, the most frequently noted symptoms related to trigeminal nerve dysfunction manifest as decreased sensation over the involved trigeminal nerve branch distribution, the whole face, and eventually corneal reflex alteration [34, 47]. Instead of pain, sensory disturbance is mostly described as numbness and paresthesia. Some authors reported that ~10 % of patients will not present with trigeminal dysfunction as an initial symptom

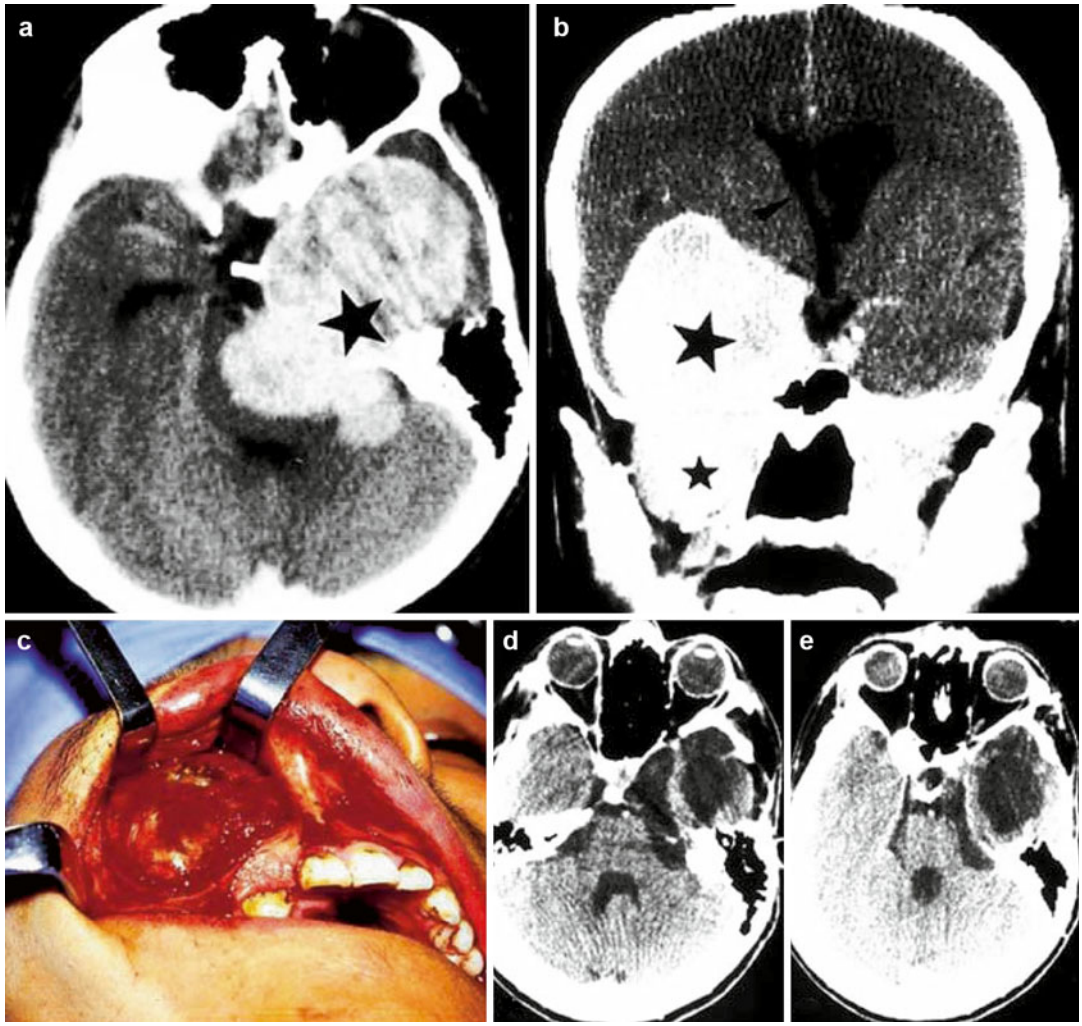


**Fig. 21.13** CT scan of a large trigeminal schwannoma (type F) with extensions in the posterior fossa, middle fossa and maxillary sinus (**a**, **b**; *stars*). (**c**) Transmaxillary tumor exposure. (**d**, **e**) Postoperative CT scans

[32, 42]. When present, however, facial pain can affect one or more trigeminal branches. Intensity may vary from light to lancinating pain. It may occur in paroxysms, usually without trigger zones, or may last hours. According to Day and Fukushima [9], the majority of patients with TSs have paroxysmal lancinating facial pain in episodes that tend to last longer than classic trigeminal neuralgia and do not respond to carbamazepine. Some authors [10, 33, 36, 49] reported that trigeminal pain may be absent with tumors originating distally in the divisions, whereas it is more common in tumors arising from the ganglion. Other reported symptoms are headaches, hearing impairment, hemifacial spasm, hemiparesis, ataxia, diplopia, signs of cerebellar involvement, increased intracranial pressure, and dysfunction of oculomotor, trochlear, and abducens nerves [3, 32, 34, 36, 51]. Although rare, malignant tumors can present with involvement in all three branches [24, 26].

Adequate preoperative neuroradiological evaluation is fundamental for establishing a correct diagnosis and treatment. Bone anatomy of the skull base is better evaluated with CT scanning and plain radiographs, which generally

reveal sharp amputation of the petrous apex, anterior-medial erosion of the petrous bone, and enlargement of the foramen ovale [25, 35, 38]. On CT scans, TSs usually appear iso- to hyperintense compared with the surrounding brain, with clearly defined margins that strongly and uniformly enhance after intravenous contrast administration [20, 37]. Cystic changes may also be observed. An MR imaging study gives the best information concerning the localization and extent of the tumor in addition to revealing displacement of neighboring structures and involvement of the cavernous sinus and vessels. On MR imaging, TSs appear hypointense in T1-weighted and hyperintense on T2-weighted images, with marked and homogeneous enhancement after intravenous Gd administration [45]. The differential diagnosis includes meningiomas, epidermoid cysts, metastasis, chondrosarcomas, chordomas, chondromas, vestibular schwannomas, and maxillary sinus tumors.

Management strategy for TSs involves clinical observation followed by MR imaging follow-up for incidental tumors, surgical removal, and, alternatively, radiotherapy or radiosurgery [2, 18, 23, 29, 30, 39, 54]. Complete or near-total surgical removal can be achieved in >70 % of the patients by means of skull base approaches and microsurgical dissection. Involvement of the cavernous sinus is one cause of subtotal resection. In most cases, a clear plane of cleavage between the tumor capsule and the cavernous sinus structures can be found, allowing complete dissection and total resection. In the modern neurosurgical era, recurrence of TS is rare and the outcome is usually favorable; the most frequent symptom after surgery is trigeminal hypesthesia, which is transient in most cases. Facial pain may persist after surgery, but most patients report improvement or total relief during follow-up. Diplopia, CSF leakage, meningitis, and hydrocephalus have been also described as possible complications. Most new cranial nerve deficits present resolution within 4–6 months.

Several reports have demonstrated that radiosurgery is a safe and effective treatment for TSs, with tumor control rate between 78 and 100 % [1, 21–23, 27, 39–41, 46, 50, 53]. However, Phi et al. [41] reported that 46 % of their patients showed tumor expansion after radiosurgery, but these cases did not require additional surgical resections. Hasegawa et al. [23] reported that 14 % of their patients did require additional surgical treatments after radiosurgery because of tumor enlargement or uncontrollable facial pain. Sheehan et al. [50] also reported that two of 26 patients harboring TSs underwent surgical resection after radiosurgery.

Although good results have been reported with radiosurgery, this technique is reserved for small, nonresectable, and residual tumors within the cavernous sinus. Long-term follow-up of patients treated with this method is still needed to evaluate the exact role of radiosurgery in the late control of these lesions. It is evident, however, that no patient will ever be cured of this benign tumor with radiotherapy or radiosurgery.

Several contemporary series have demonstrated no deaths or major surgical complications with radical removal of TSs [9, 13, 17, 18, 44, 47]. In a classic series of 44 patients reported by Dolenc [13], total resection was achieved in 100 % of the patients, including 5 who had undergone incomplete resection elsewhere and underwent another surgery to excise the remain of the tumor. The authors recommend an epidural approach to schwannomas originating in the fifth cranial nerve peripherally to the Gasserian ganglion and either an epidural-transdural or an epidural-transdural-transpetrous approach to lesions originating in the Gasserian ganglion or in the root of the fifth nerve.

Patients who had experienced preoperative atypical trigeminal pain reported a resolution of their symptoms after surgery. No additional treatment such as radiosurgery, other forms of irradiation, or chemotherapy was necessary.

Based on our experience and published data, we believe that the best treatment for TSs is complete microsurgical removal of the lesion and that this treatment should be considered the gold standard therapeutic modality for the majority of cases when performed in selected, well-established, skull base neurosurgical centers.

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