

Surgical treatment of trigeminal schwannomas

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Received: 16 December 2006 / Revised: 22 March 2007 / Accepted: 28 April 2007 / Published online: 4 August 2007
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Abstract Schwannomas that arise from the trigeminal nerve are rare, but this nerve is the second most frequent intracranial site of schwannoma occurrence next to the vestibular nerve. The advent of microsurgical techniques and skull-base approaches has greatly enhanced the surgical management of these tumors, and outcomes have improved markedly. This report documents 18 cases of histologically verified schwannomas that arose from the trigeminal nerve and were treated surgically in our clinic between January 1992 and July 2005. The patients were ten women and eight men of age 39.7 years (range, 22–62 years). The tumor was located in the middle fossa (type A) in five cases, in the middle and posterior fossae (type C) in nine cases, in the posterior fossa (type B) in two cases, and in the branches of the trigeminal nerve (type D) in two cases. Total excision was achieved in 17 cases, and there was no mortality in the series. Our results indicate that trigeminal schwannomas, regardless of type, can be removed via skull-base approaches. We present an algorithm for surgical management of trigeminal schwannomas based on our experience and information from the literature.

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Keywords Cavernous sinus · Schwannoma · Surgery ·
Trigeminal

Introduction

Schwannomas are benign neoplasms of peripheral nerve sheaths that arise distal to the oligodendroglial-schwann myelination junction. Trigeminal nerve schwannomas are rare, but this nerve is the second most common intracranial site of occurrence after vestibular nerve origin. Trigeminal schwannomas account for 0.07% to 0.28% of all intracranial tumors and 0.8% to 8% of all intracranial schwannomas [7, 11, 16, 17, 22, 30]. As noted, these tumors can arise anywhere between the root and the distal extracranial branches of the trigeminal nerve. They produce a variety of symptoms and signs that depend on tumor location and the direction and extent of tumor growth.

The aim of surgery in cases of trigeminal schwannoma is tumor extirpation with preservation of cranial nerve function. Introduction of microsurgical techniques and advanced skull-base approaches has led to markedly improved outcomes for surgically treated trigeminal schwannomas [6, 8, 17, 22, 26, 29–31]. This report documents our experience with surgical treatment of these tumors.

Patients and methods

Eighteen patients with histologically verified trigeminal schwannomas underwent surgical treatment in our clinic between January 1992 and July 2005. Detailed general, ophthalmological and neurological examinations were done in each case.

The preoperative neuroimaging evaluations included magnetic resonance (MR) imaging (all cases), computed tomography (CT) scanning (ten cases only), and cerebral angiography (five cases only). Each tumor was classified according to Day and Fukushima's modified version of the Jefferson classification system. Jefferson defined three types of trigeminal schwannomas: type A, located mainly in the middle fossa; type B, located mainly in the posterior fossa; type C, with significant components in both the middle fossa and the posterior fossa [17]. Day and Fukushima [6] identified a fourth group (type D) comprised of schwannomas that arise from the branches of the trigeminal nerve. At our center, the Day and Fukushima classification system essentially dictates which surgical approach is used.

All 18 patients underwent surgical excision aimed at total tumor removal. Intraoperative monitoring was done in five cases, and this included recording of brainstem auditory-evoked responses and somatosensory-evoked potentials with median nerve stimulation.

Two of the tumors were located within the posterior fossa alone, and the classical retromastoid suboccipital approach was used in both these cases. In the other 16 cases, the skull-base cavernous sinus approach described by Dolenc (fronto-temporal intra- and/or extradural approach) was performed [8]. The extradural route was used in 14 cases, and the extra-intradural route was used in 2 cases.

Postoperative evaluation and follow-up checks involved general and neurological examinations. In each case, a CT scan was performed within first 6 h after surgery, and MR imaging was done within the first 24 h to assess the extent of tumor removal. MR imaging was repeated approximately

6 months after the operation and annually thereafter to assess for recurrence. Anticonvulsants were only given for 3 months. Steroids were given for 7–10 days postoperatively. In all cases, pathological evaluation included intraoperative evaluation of frozen sections and examination of formalin-fixed specimens with routine and special staining.

Results

The patients were ten women and eight men of mean age 39.7 years (range, 22–62 years). Table 1 lists the clinical findings in the 18 cases.

Neuroimaging findings

As noted, preoperative CT was done in ten cases, and this revealed bone erosion in seven patients. Five of the ten tumors enhanced with contrast.

Preoperative MR imaging was performed in all cases. Eight tumors were isointense and 10 were slightly hypointense on T1-weighted images, 15 were hyperintense on T2-weighted images, and 17 showed homogeneous enhancement after administration of contrast agent. The images revealed that seven of the masses had a cystic component. The distribution of tumor sizes is shown in Table 2.

The CT, MR imaging, and surgical findings helped us accurately diagnose the trigeminal schwannomas and clearly identify each tumor's location, extent and origin. The tumor was located in the middle fossa (type A) in five cases (Figs. 1 and 2), the posterior fossa (type B) in two

Table 1 Clinical findings in the 18 cases of trigeminal schwannoma

Case	Age (years)/sex	Initial symptom	Trigeminal deficit		Additional deficit	
			Sensory	Motor	Cranial nerve	Other
1	48/M	Facial hypoesthesia	Yes	No	No	No
2	54/F	Facial hypoesthesia	Yes	No	No	No
3	52/M	Weakness on left	No	No	No	Hemiparesis
4	35/F	Facial hypoesthesia	Yes	No	No	No
5	24/F	Headache	No	No	No	Ataxia
6	36/F	Facial hypoesthesia	Yes	No	No	No
7	47/M	Diplopia	No	No	VI	No
8	62/F	Headache	No	No	VIII	No
9	31/F	Diplopia	No	No	VI	No
10	35/F	Facial hypoesthesia	Yes	No	No	No
11	49/F	Headache	No	No	No	Ataxia
12	32/M	Orbital pain	No	No	III and VI	No
13	38/M	Facial hypoesthesia	Yes	No	No	No
14	60/F	Facial hypoesthesia	Yes	No	No	No
15	29/M	Diplopia	No	No	VI	No
16	22/M	Facial hypoesthesia	Yes	Yes	No	No
17	35/F	Headache	Yes	No	No	No
18	39/M	Headache	Yes	No	No	No

Table 2 Distribution of the tumors according to size

Size	No. of cases
<2 cm	2
2–4 cm	7
4–6 cm	7
>6 cm	2

cases (Fig. 3), the middle and posterior fossae (type C) in nine cases (Fig. 4) and along branches of the trigeminal nerve (type D) in two cases (Fig. 5).

Operative results

The 18 patients underwent a total of 21 operations (Table 3). Two-staged surgery was performed on one type C tumor. Another patient required surgery for recurrence at

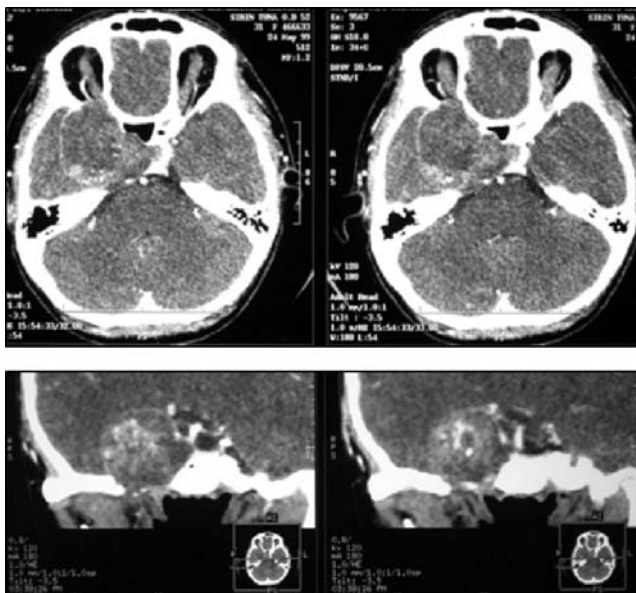


Fig. 1 Preoperative CT and 3-dimensional CT images show bone erosion caused by a giant trigeminal schwannoma

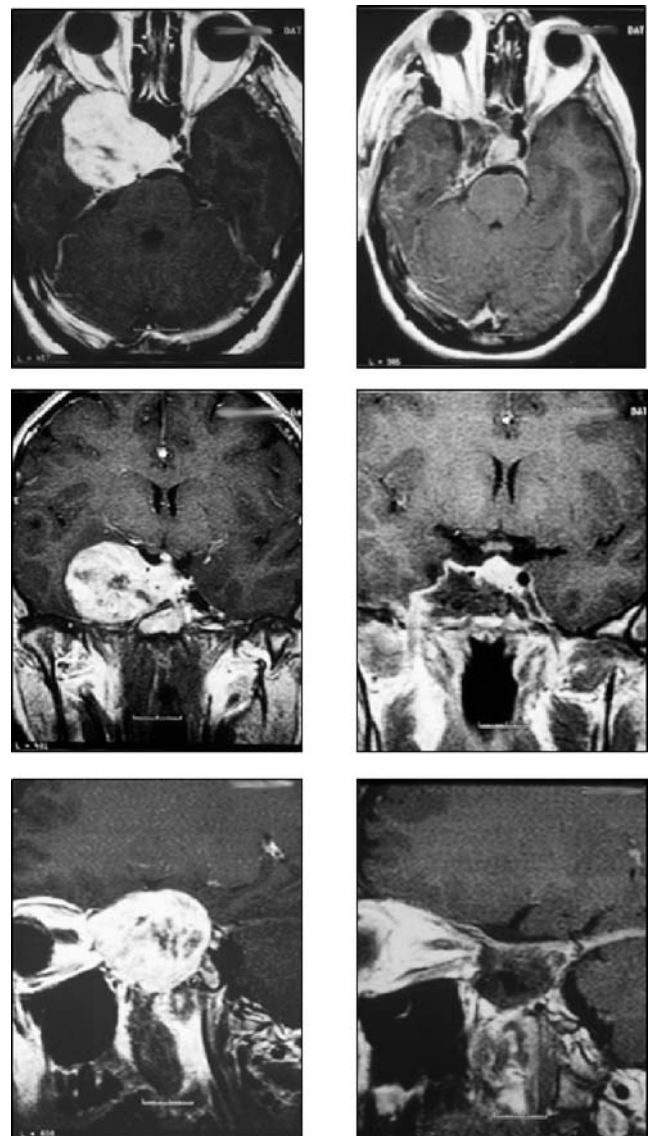


Fig. 2 Preoperative (left column) and postoperative (right column) MR images show a giant type A trigeminal schwannoma. Total excision was achieved via a fronto-temporal epidural approach

96 months after the first operation. Another needed repeat surgery immediately after tumor removal to address an epidural hematoma. Total excision was achieved in 17 cases, and only subtotal resection was possible in one case. The tumor was hard in this case. Other tumors were all soft and suckable. Gamma-knife radiosurgery was performed on the residual tumor tissue in the latter case.

Histopathological results

On histopathological examination, only 3 tumors were with Antoni A type and the remaining 15 tumors were Antoni B type.

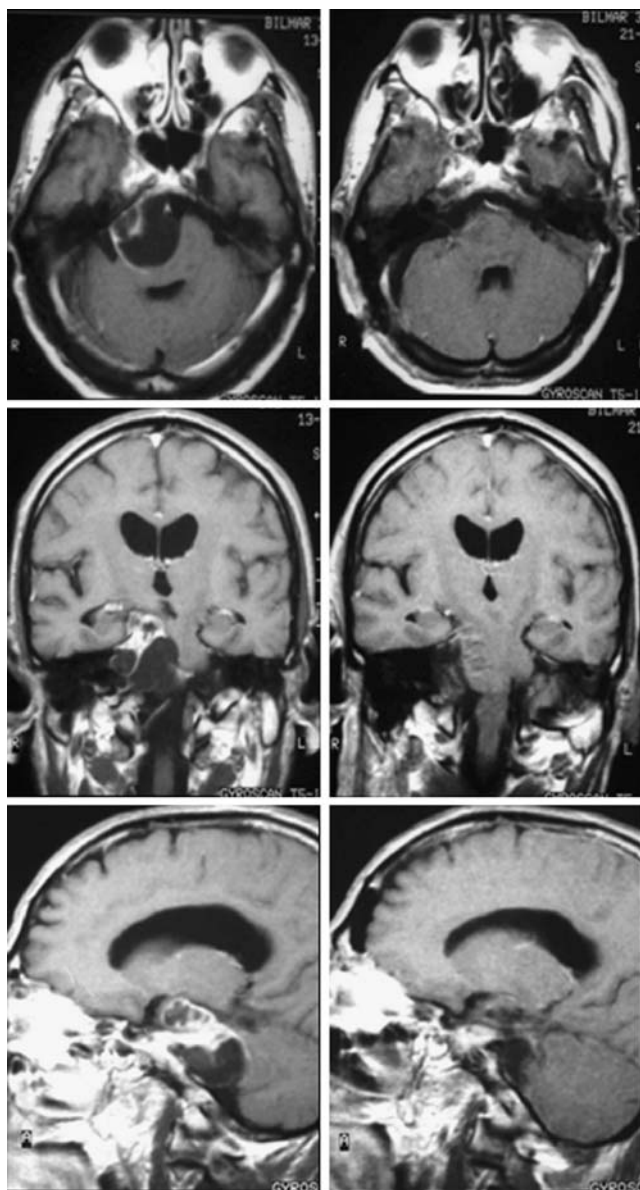


Fig. 3 Preoperative (left column) and postoperative (right column) MR images show a type B trigeminal schwannoma. Note the cystic degeneration and displacement of the pons caused by tumor compression

Outcome

Outcome after surgery was strongly correlated with extent of tumor excision. The mean follow-up time for this study was 134 months (range 14–192 months). Sixteen of the 17 patients who had their tumors surgically excised remained free of clinical symptoms and showed no MR imaging evidence of recurrent disease during follow-up. Only one of the patients who underwent complete tumor excision (according to postoperative MR imaging findings) experienced tumor recurrence, and this was detected 96 months after the initial surgery. As noted, a second surgery was done to remove the recurrent mass.

Postoperative neurological function

Fifteen of the 18 patients exhibited abnormal fifth nerve function immediately after surgery, and these deficits turned out to be permanent in three cases. Six patients showed transient deficits in cranial nerves other than the fifth nerve after surgery. Only two of these were permanent deficits (both abducens nerve palsy).

Complications

One patient developed cerebrospinal fluid leakage from the wound site, and this was treated with lumbar drainage. As

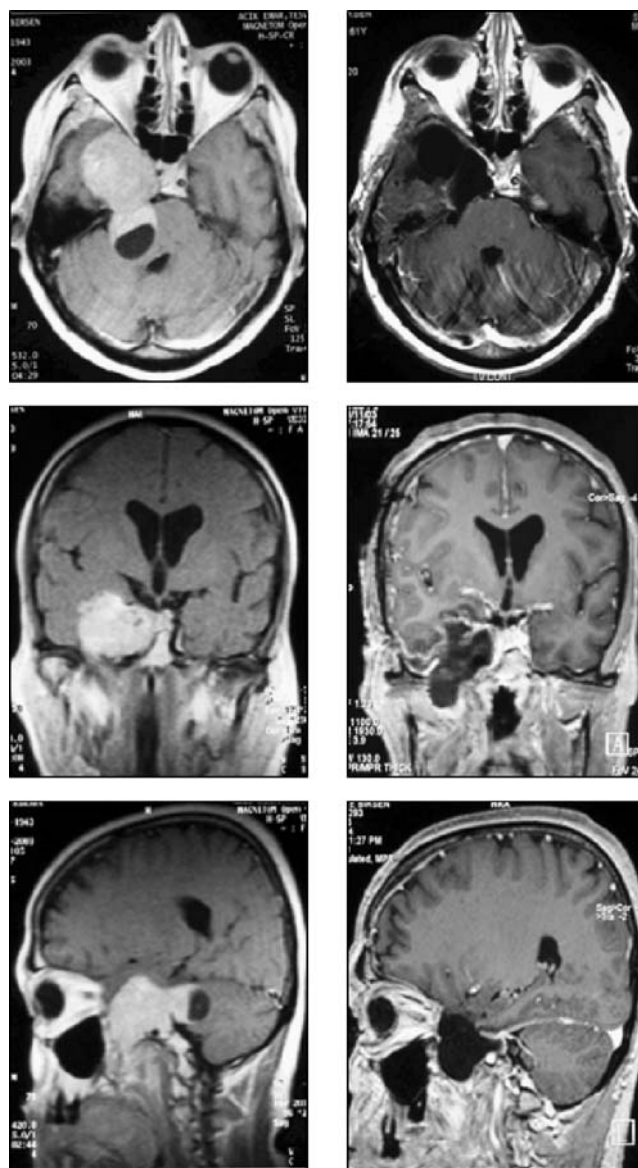


Fig. 4 Preoperative (left column) and postoperative (right column) MR images show a type C trigeminal schwannoma. The tumor has a larger component in the middle fossa than in the posterior fossa, and also features a cystic component

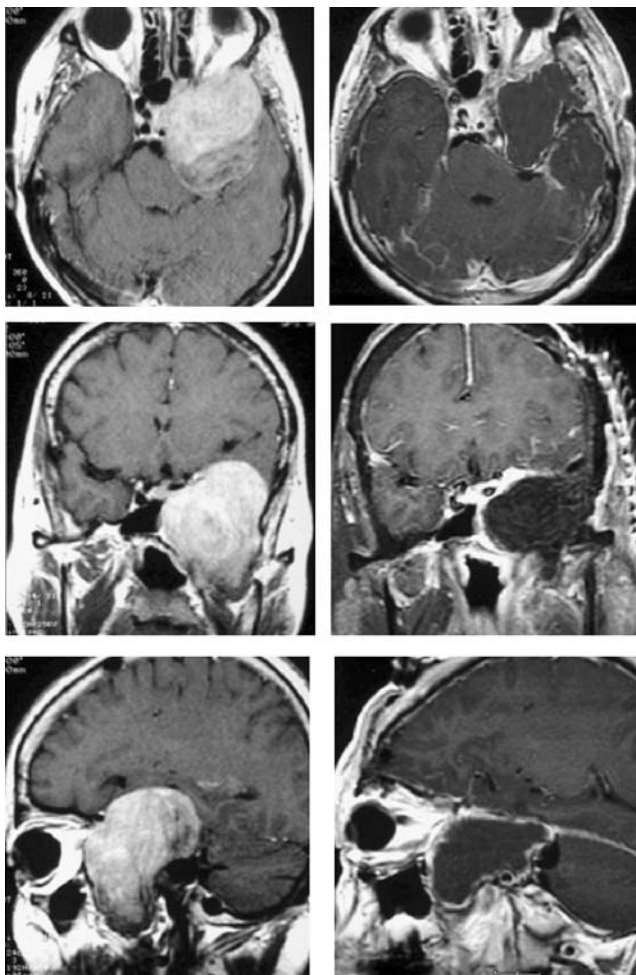


Fig. 5 Preoperative (left column) and postoperative (right column) MR images show a type D trigeminal schwannoma that extends to the infratemporal fossa

noted, one patient developed an epidural hematoma that required immediate surgical evacuation. Another individual developed postoperative meningitis and was successfully treated with antibiotics.

Discussion

Clinical presentation

Our 18 patients with trigeminal schwannomas ranged from 22 to 62 years of age (mean, 39.7 years), and there was no significant sex bias in the group (male-to-female ratio 1:1.3). There were no cases of bilateral trigeminal schwannoma, which may be associated with neurofibromatosis [11], and none of the patients showed symptoms of neurofibromatosis. The early symptoms of trigeminal schwannoma can be mild and may exist for long periods of time before they are investigated. This was the situation with all our cases, and most of the tumors reached large size before they were diagnosed. Most of the 18 patients presented with paresthesia or numbness in the area innervated by one or more branches of the trigeminal nerve. None of the patients was experiencing severe or neuralgic pain at presentation. Only one individual showed wasting of the temporalis muscle. All 18 patients had normal corneal reflexes. Five presented with deficits of cranial nerves other than the trigeminal nerve, and this relates to the large size of the tumors in our series (Table 1). Cerebellar symptoms, pyramidal system symptoms, and problems related to increased intracranial pressure were

Table 3 Tumor locations and operative approaches for the 18 cases of trigeminal schwannoma

Case No.	Tumor location	Tumor type	Operative approach	Extent of removal
1	Middle and posterior fossa	C	FT extradural	Total
2	Middle and posterior fossa	C	FT extradural	Total
3	Middle and posterior fossa	C	FT extradural	Total
4	Infratemporal fossa	D	FT extradural	Total
5	Posterior fossa	B	Suboccipital retromastoid	Total
6	Middle fossa	A	FT extradural	Total
7	Middle fossa	A	FT extradural	Total
8	Infratemporal fossa	D	FT extradural	Total
9	Middle and posterior fossa	C	FT extradural	Total
10	Middle and posterior fossa	C	FT intra-extradural	Total
11	Posterior fossa	B	Suboccipital retromastoid	Total
12	Middle and posterior fossa	C	FT extradural	Subtotal
13	Middle fossa	A	FT extradural	Total
14	Middle and posterior fossa	C	Two-staged surgery (Retromastoid+FT extradural)	Total
15	Middle fossa	A	FT extradural	Total
16	Middle fossa	A	FT extradural	Total
17	Middle and posterior fossa	C	FT intra-extradural	Total
18	Middle and posterior fossa	C	FT intra-extradural	Total

FT: Fronto-temporal

observed in one case each, and these were all attributed to large tumor size as well. Symptoms of proptosis and pathologic laughter are reported to be rare in cases of trigeminal schwannoma, and none of our patients exhibited these abnormalities [4, 11]. Clinically, predominance of symptoms related to the trigeminal nerve and slow progression of these symptoms may assist in diagnosing trigeminal schwannoma.

Radiological features

Preoperative radiological evaluation is essential to establish the precise location of a trigeminal schwannoma and its relationship to neighboring structures. These features help guide the surgical strategy. MR imaging has revolutionized the management of these tumors, but other modalities, such as plain radiography and CT scanning, are still useful for assessing bone architecture and skull-base anatomy.

On plain X-ray films of the skull, trigeminal schwannomas of the middle fossa and dumbbell-shaped lesions that traverse the foramen ovale generally produce a sharp-edged bone defect in the floor of the middle fossa [3, 18, 21, 26, 27]. These lesions usually also erode the foramen ovale and the foramen spinosum, as well as the anteromedial portion of the apex of the petrous bone. Trigeminal schwannomas that extend medially and anteriorly from the cavernous sinus typically erode the lateral aspect of the sella turcica, the anterior clinoid process, the dorsum sellae, or the superior orbital fissure [7, 18, 21, 26, 27].

When investigating large trigeminal schwannomas, cerebral angiography is necessary because it is essential to determine the tumor's vascularity and assess for vessel displacement prior to surgery. Displacement of the internal carotid artery is characteristic of trigeminal schwannomas and, thus, is highly diagnostic. Transgression of the medial dural wall of cavernous sinus and invasion of the venous spaces of the cavernous sinus, encasement of the pre-cavernous sinus and encasement of the cavernous sinus portion of the internal carotid artery have been reported previously, but none of our 18 cases had these features [17, 26]. However with the advent of MR angiography, there is a little role for angiography today.

In cases of trigeminal schwannoma, high-resolution CT clearly demonstrates tumor location, extent of tumor growth and any bony changes that have occurred. Most of these neoplasms appear isodense or slightly hyperdense on CT and enhance uniformly with intravenous contrast [12, 19].

MR imaging has added a new dimension to preoperative diagnosis and assessment of trigeminal schwannomas. The advantages of this modality include highly sensitive detection of contrast differences, the ability to obtain sagittal views, the absence of streak artifacts caused by bone and the ability to visualize vascular structures in

relation to the lesion [25]. In addition, displacement of normal structures and lesions with altered signal intensities are better delineated on MR images than on CT. Trigeminal schwannomas appear isointense or slightly hyperintense on T1-weighted images and show significant enhancement after contrast injection.

Surgical strategy

Early attempts at surgical removal of trigeminal schwannomas were associated with significant morbidity and mortality. Schisano and Olivecrona [27] investigated a series of 19 cases in 1960 and reported operative mortality of 5.3%. The same authors noted a 1-year mortality rate of 41% for the 39 operative cases of trigeminal schwannoma that were reported in the literature prior to 1956. Arseni and Camenita [3] reported 25% postoperative mortality for all cases published up to 1970.

Introduction of the operating microscope and improved anesthetic and microsurgical techniques have significantly reduced the morbidity and mortality risks associated with operative treatment of trigeminal schwannomas (Table 4). Even malignant trigeminal schwannomas can be seen; these neoplasms are almost always biologically benign; therefore, complete surgical excision with minimal morbidity is the primary goal of surgery. The most important feature of our series was that extradural surgical approaches were used to resect the majority of the tumors, even though the trigeminal schwannomas were of different types.

Type A trigeminal schwannomas are located in the interdural space and are enveloped by the inner membrane of the cavernous sinus. This membrane is composed of the perineurium of cranial nerves within the sinus. Subtemporal, fronto-temporal intradural and fronto-temporal epidural approaches have been described for removing these tumors [8, 16, 26]. Early descriptions detailed an extradural surgical route for approaching type A trigeminal schwannomas. This method involved extradurally exposing the gasserian ganglion complex, followed by a subtemporal intradural or fronto-temporal intradural approach. In both of the latter, the tumor was exposed after the dura was opened. These steps provided sufficient exposure, but required significant brain retraction and usually demanded sacrifice of bridging veins at the temporal tip [25]. The extradural strategy described by Dolenc [8] offers the advantage of more direct access to the region with less brain retraction required. As well, if more exposure is needed, the brain tissue can be retracted while being protected by overlying dura, which helps preserve venous structures. In our series, we achieved excellent surgical and clinical results with the fronto-temporal epidural approach for type A tumors. We found that this method allowed for easy dissection of the tumor from surrounding structures, and there were no

Table 4 Operative series of trigeminal schwannomas in the literature

Authors	No. of cases					Total removal	Perm. morbidity	Mortality
	Type A	Type B	Type C	Type D	Total			
Conventional approaches								
Schisano & Olivecrona [27]	11	4	0	0	15	5 (33%)	60%	0%
de Benedittis et al. [7]	5	1	3	0	9	4 (44%)	11%	0%
McCormick et al. [17]	6	5	2	1	14	6 (43%)	78%	0%
Pollack et al. [22]	6	4	5	1	16	12 (75%)	6%	0%
Skull-base approaches								
Yasui et al. [30]	2	2	3	1	8	8 (100%)	100%	0%
Dolenc [8]	NA	NA	NA	NA	40	40 (100%)	25%	0%
Samii et al. [26]	5	1	5	1	12	10 (83%)	0%	0%
Konovalov et al. [14]	42	26	30	13	111	86 (77%)	87%	3%
Day & Fukushima [6]	18	9	9	3	38	30 (79%)	79%	0%
Yoshida & Kawase [31]	4	5	10	8	27	20 (74%)	74%	0%
Al-Mefty et al. [1]	6	0	17	2	25	25 (100%)	40%	0%
Goel et al. [11]	29	7	30	7	73	51 (70%)	7%	2%
Mariniello et al. [16]	5	0	0	0	5	5 (100%)	0%	0%
Present study	5	2	9	2	18	17 (94%)	28%	0%

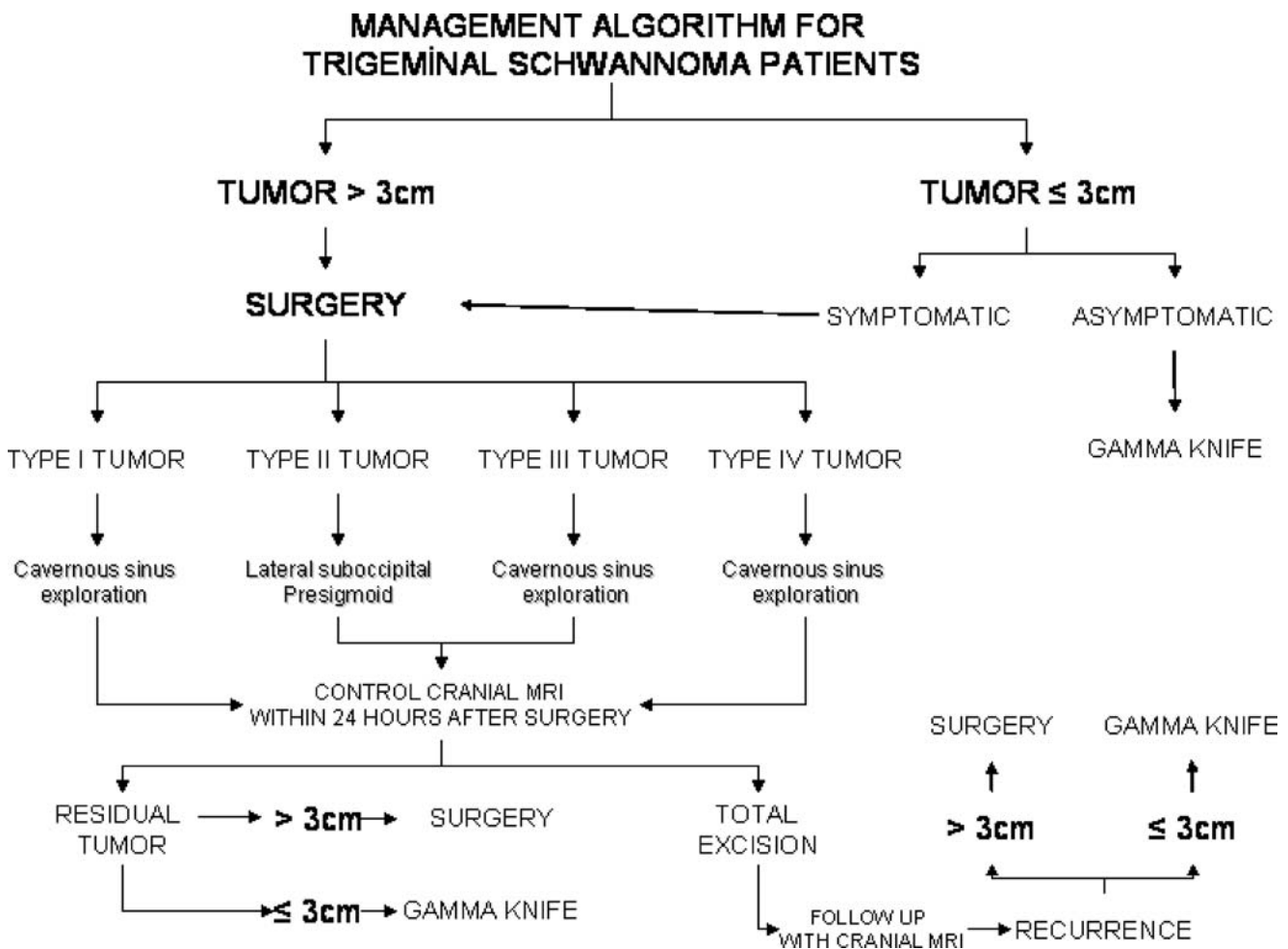


Fig. 6 An algorithm for surgical management of trigeminal schwannomas

complications related to brain retraction. With this approach, it was as easy to expose large tumors as it was to expose small tumors.

Type B trigeminal schwannomas are located in the posterior fossa and originate from the trigeminal nerve root. The literature discusses the use of suboccipital or transpetrosal transtentorial approaches for this type [6, 11, 17, 30]. Our series featured two type B trigeminal schwannomas, and we used the classical suboccipital retromastoid route for these patients. Total excision was achieved in both cases (Fig. 2). With this approach, tumor debulking and dissection should be done as recommended for vestibular schwannomas.

Type C trigeminal schwannomas have significant components in both the middle fossa and the posterior fossa, and are known to be the most challenging type to remove. The clinical and surgical considerations for this type are distinct. Bordi et al. and McCormick et al. stated that they prefer to approach these tumors via a subtemporal transtentorial route [5, 17]. This approach provides a limited view of the underside of the neoplasm and its relationship to the vessels and cranial nerves below. Samii et al. have suggested that it is more difficult to totally remove a type C trigeminal schwannoma when the retrosigmoid and subtemporal approach is used [26]. Yasui and colleagues [30] and Al-Mefty et al. [1] reported total removal of type C tumors in 4 and 19 cases, respectively. Yasui and co-workers used a transpetrosal transtentorial approach or an orbitozygomatic infratemporal approach for their cases, and performed a two-staged operation with suboccipital/frontopterional approaches in one case. Al-Mefty and colleagues used the extradural zygomatic middle fossa approach in 14 of 19 type C cases. Al-Mefty et al. also described removing the posterior fossa component of these tumors through the expanded Meckel's cave without sectioning the tentorium or drilling the petrous apex. In our series, we operated on nine type C tumors. Total resection was achieved in eight cases, and the residual tissue in the case of subtotal resection was treated with gamma-knife radiosurgery. Of the eight cases of total resection, five were performed via a fronto-temporal extradural approach and three were performed via a fronto-temporal extra-intradural approach. One of the eight patients underwent two-staged surgery, with the first operation done via retromastoid route and the second performed via a fronto-temporal extradural approach. In our experience with nine type C trigeminal schwannomas, we found that the fronto-temporal approach provided better access to the posterior fossa through the enlarged Meckel's cave. This allowed easier excision. In contrast, the posterior fossa approach provides limited access to the portion of the tumor located in the middle fossa.

Type D trigeminal schwannomas arise from the peripheral portion of the trigeminal nerve. These tumors can

extend to the infratemporal fossa via the foramen ovale and/or foramen rotundum, or can erode the base of the middle fossa. Type D trigeminal schwannomas can be accessed via an entirely extradural approach [6, 26]. Our series featured two type D tumors, and we successfully excised these via a cranioorbital extradural approach and a fronto-temporal extradural approach, respectively.

Radiosurgery for trigeminal schwannomas

Reports indicate that stereotactic radiosurgery, as an alternative or adjunct to surgery, yields excellent long-term outcomes for patients with trigeminal schwannomas [2, 9, 10, 13, 15, 20, 23, 24, 28]. For large symptomatic trigeminal schwannomas that are causing mass effect, surgical removal is the preferred initial management strategy. However, when a tumor cannot be excised completely and the residual tumor is less than 3 cm in diameter, radiosurgery is an excellent secondary strategy. This method is also very valuable for treating recurrence. The goal for managing recurrent and residual trigeminal schwannomas should be to control tumor growth. Radiosurgery seems ideal for treating small residual and primary tumors located in the cavernous sinus. We suggest that patients who have asymptomatic primary, residual or recurrent tumors smaller than 3 cm diameter should be offered the option of radiosurgery instead of resection.

The findings from our series indicate that all types of trigeminal schwannomas can be removed via skull-base approaches. Based on our experience and what is documented in the literature, we created an algorithm for surgical management of trigeminal schwannomas (Fig. 6). According to this system, tumor size and symptoms are two major factors in the management of primary trigeminal schwannomas. Regardless of size, symptomatic tumors require surgery. Small asymptomatic trigeminal schwannomas may be treated with stereotactic radiosurgery. For residual and recurrent tumors, size is an important factor when considering management options; small masses (those less than 3 cm diameter) can be effectively treated with radiosurgery.

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Comments

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Pamir and coworkers present their experience in the surgical treatment of trigeminal schwannomas in 18 cases. They should be congratulated for reporting an excellent outcome with a long duration of follow-up and no mortality. None of the patients have trigeminal motor dysfunction, which is interesting. They have been able to achieve total resection of the tumor in 17 cases using skull base approaches, and present a useful algorithm for management of trigeminal schwannomas. However, in spite of majority of the tumors being Antoni B type, which represents a biologically aggressive nature of the tumor, the recurrence rate has been surprisingly low. Trigeminal schwannomas have characteristic clinical presentation and radiological features. They usually present with a trigeminal motor or sensory neuropathy and frequently have the epicenter in the Meckel's cave or the Gasserian ganglion. The tumor does not completely destroy the trigeminal nerve and few fibres can be preserved, especially the first division of the trigeminal nerve. Amongst the various skull base approaches, we find the lateral basal subtemporal extradural approach most suited to tackle most of the small, the medium-sized as well as large trigeminal schwannomas. However, selection of an appropriate approach should be strictly individualized. As our experience in the management of this condition evolved over a period of years and better understanding of the anatomy, the two-staged approach has been infrequently used, especially for the type C tumors. Large-sized tumors can be resected completely in experienced hands, with acceptable morbidity rates and long-term recurrence-free intervals. Radiosurgery is a useful option for recurrent tumor less than 3 cm in size, but the long-term side effects and dosage standardization still need to be addressed.