Trigeminal Neurinomas: A Review of a Personal Series

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Abstract:

The surgical management issues of 157 cases of trigeminal neurinoma treated over a 20-year period were analyzed. The case records and radiologic material of these patients who were operated on in the Neurosurgery Department at King Edward Memorial Hospital and Seth Gordhandas Sunderdas Medical College, Mumbai, between the year 1989 and 2009 were retrospectively analyzed. The appropriateness of the selected surgical route is studied. Apart from the other typical presenting features of trigeminal schwannoma, 12 patients presented with the rarely reported symptom of pathologic laughter. Three approaches were found appropriate to treat these tumors: the infratemporal fossa interdural approach, the lateral and anterior basal subtemporal approach, and the retrosigmoid approach. For tumors extending extracranially, a 'reverse skull base approach' that involved basal temporal craniotomy was used. In 129 (82%) cases, total tumor excision was achieved. Two patients died during the postoperative period. During an average follow-up of 52 months, there has been a recurrence in 5 cases. Radical surgery is associated with an excellent clinical outcome and long-term tumor control. A majority of tumors, even those that are large and multicompartmental, can be removed in a single surgical stage and exposure.

Key Words: gasserian ganglion, retrosigmoid approach, subtemporal approach, trigeminal neurinoma

Trigeminal neurinomas are relatively rare tumors and represent 0.2% of all intracranial tumors. (1–12) Trigeminal neurinomas usually arise from the Schwann cells of the sensory root and can originate in any section of the fifth cranial nerve; correspondingly, a variety of symptoms and signs may develop. Improved surgical outcome and long-term growth control have been uniformly reported after the improvement in diagnosis and understanding of the anatomical intricacies of the tumor and advancement in the skull base operative techniques. (13–21) Jefferson (6) presented a classification scheme for trigeminal schwannomas that categorized these tumors according to location. Three distinct types are: middle fossa type (type A); posterior fossa root type (type B), where the tumor is in front of the brainstem; and dumbbell-shaped type with both middle and posterior fossa components (type C). Less commonly, the tumor has an extracranial extension (type D). (7) Yoshida and Kawase (21) classified the extracranial tumor into infratemporal, orbital, and pterygopalatine fossa components (Table 1). We recently reported a series of 28 cases of trigeminal neurinomas with extracranial extension. (22) There was tumor extension along the ophthalmic division of the nerve in 4 cases, along the maxillary division in 5, and along the mandibular division in 13. In 6 tumors there was diffuse extracranial extension and the exact extracranial division of nerve involvement could not be ascertained. In 10 cases, the tumor had a multicompartmental location – in the posterior fossa, the middle fossa, and the extracranial compartment. The series of trigeminal neurinomas in general and of those having an extracranial extension is the largest personal series published in the literature.

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Anatomical Considerations

After its exit from the medial aspect of middle cerebellar peduncle, the fifth cranial nerve traverses under the tentorium and later over the petrous apex, where it forms the Gasserian ganglion. The Gasserian ganglion then divides

into 3 divisions: the first and second divisions participate in forming the lateral wall of the cavernous sinus, whereas the third division exits from the cranial cavity via the foramen ovale and has a short intracranial course. As the fifth nerve travels over the petrous apex, a dural and arachnoid sheath forming the cave of Meckel covers it. There is a large subarachnoid space over the Gasserian ganglion in the cave of Meckel containing cerebrospinal fluid. Apart from this dural and arachnoid envelope, which is continuous with the corresponding layers in the posterior fossa, the fifth nerve is also covered by the 2 divisions of the middle fossa dura, which divide at the lateral border of the Gasserian ganglion. The inner (cerebral) layer continues as the lateral wall of the Gasserian ganglion and cavernous sinus, whereas the outer layer (osteal layer) continues medially and forms the inferior dural layer of Gasserian ganglion and the medial wall of the cavernous sinus. This sheath of dura is closely approximated but manually separable from the inner dural layers. The dural layers can be easily dissected off the Gasserian ganglion because of the large subarachnoid space. The dural layers merge with sheaths of the divisions of the nerve at the level of the foramen ovale, foramen rotundum, and superior orbital fissure, where the dissection of the dura from the neural tissue is relatively difficult. Despite the merging of the meninges, they continue along with the extracranial part of the nerve and are then labeled as perineurium. The superior petrosal sinus traverses in the layers of the dura superior, and the inferior petrosal sinus traverses inferior to the root of the fifth nerve at its entry in the cave of Meckel. The precavernous segment of the carotid artery is posteromedial in relation to the Gasserian ganglion; in approximately two thirds of cases, there is either only a thin shell of bone or a tough cartilaginous layer in addition to the meningeal cover of the nerves that provides a barrier between the carotid artery and the Gasserian ganglion. No matter how big the tumor becomes, it seldom actually pierces the dura and enters into the venous spaces of the cavernous sinus or engulfs the internal carotid artery in its precavernous or cavernous sinus segment. The middle fossa dura was not pierced by the tumors in any of our cases, despite the large size of the tumors. The origin of these tumors is from a segment of the nerve, and rest of the nerve is involved by displacement as a result of the growing mass. From our experience, we believe that the site of origin of these tumors is at the point where the nerve enters the cave of Meckel. Most trigeminal neurinomas, irrespective of the site of spread, have an association with this region of the nerve. The tumor grows larger and spreads in the available space.

The cave of Meckel can accommodate a large amount of the tumor, which enlarges the cave. The tumor presses the adjacent normal fifth nerve, most of which is clinically involved by direct pressure of the tumor. In the posterior fossa, trigeminal neurinomas are located intradurally. However, we have recently identified a number of posterior fossa tumors that are located entirely within the dural confines and not extending into the subarachnoid space. The presence of dura/ meninges around the tumor provides a protective barrier during the operative procedure that assists dissection from closely related cranial nerves and carotid artery (Fig.1). In general, these tumors involve the adjoining cranial nerves, blood vessels, and brain only by displacement and not by invasion. We analyzed 157 cases of trigeminal neurinoma treated surgically during the period from 1989 to 2009 (Tables 2–4).

TABLE 1. Physical characteristics of the tumor

Tumor characteristics	Number of patients	Percentage
Location of tumor		
Type A	57	36.3
Type B	14	08.9
Type C	58	36.9
Type D	28	17.9
Tumor size		
<2 cm	11	06.9
2–4 cm	57	36.9
4–6 cm	69	43.8
>6 cm	20	12.3
Nature of tumor		
Predominantly solid	24	32.8
Predominantly cystic	13	17.8
Mixed	26	35.7
Calcification	04	05.5

TABLE 2. Demographics

Clinical features	Number of patients	Percentage
Age		
0–10 years	04	02.5
11–20 years	34	21.7
21–30 years	43	27.3
31–40 years	36	22.9
41–50 years	23	14.7
>50 years	17	10.9
Sex		
Male	66	42
Female	91	58

TABLE 3. Clinical presentation

Duration of symptoms		
<1 month	19	12.1
1–6 months	74	47.1
6 months–1 year	41	26.1
1–2 years	14	8.9
>2 years	09	5.7
Presenting symptoms		
Facial numbness	121	77
Facial pain	18	11.4
Headaches	107	68.2
Gait disturbance	67	42.6
Pathologic laughter	12	7.6
Hearing deterioration	29	18.4
Diplopia	32	20.3
Visual deterioration	21	13.3
Proptosis	12	7.6
Limb weakness	9	5.7
Seizures	5	3.2

TABLE 4. Physical abnormalities

Cranial nerve involvement	Number of patients	Percentage
Trigeminal nerve		
Sensory		
V1	80	50.9
V2	136	86.6
V3	84	53.5
Motor	131	83.3
Abducens	23	14.6
Occulomotor	9	5.7
Facial	16	10.1
Hearing	24	15.2
Lower cranial nerves	11	7.0
Cerebellar signs	54	34.3
Pyramidal signs	14	8.9

Figure 1a: Line drawing showing the trigeminal nerve and its relationship.

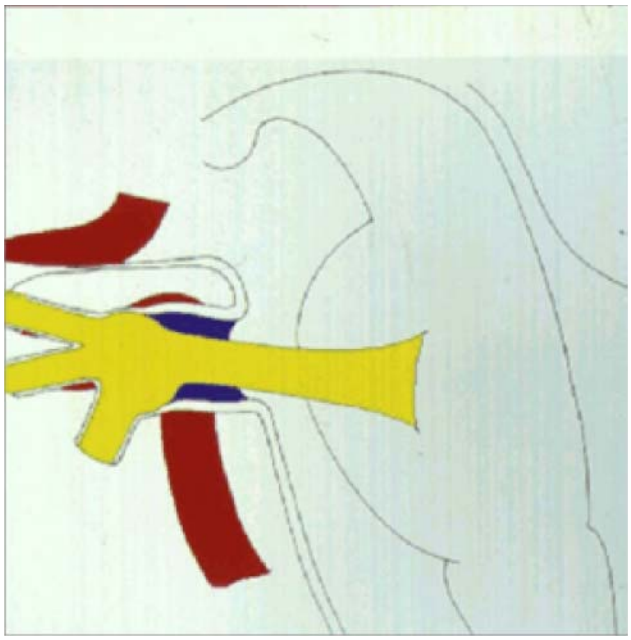


Figure 1b: Dumbbell-shaped trigeminal neurinoma. It is interdural in the cave of Meckel and intradural in the posterior fossa.

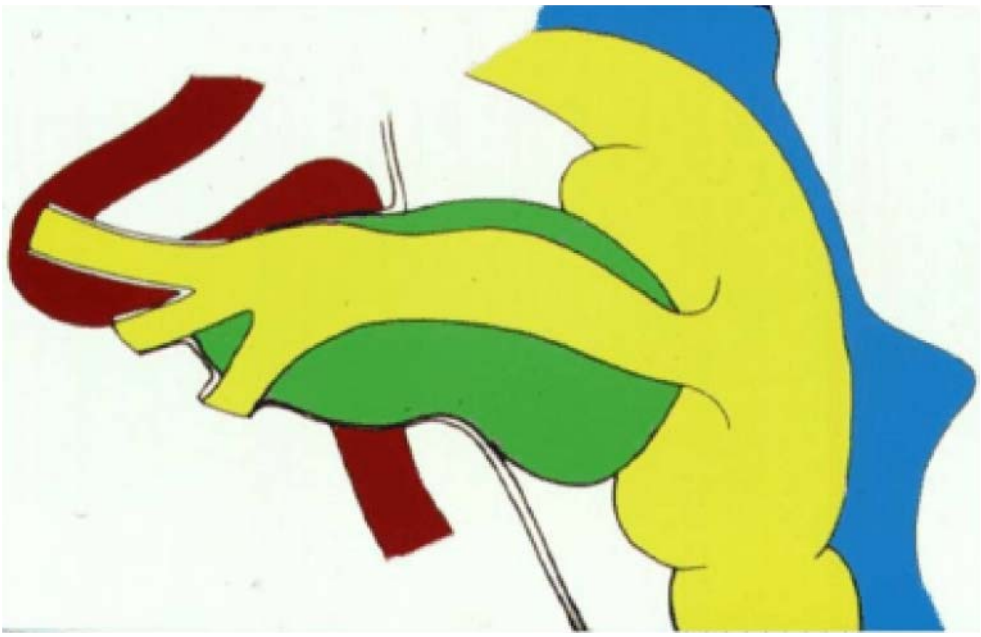
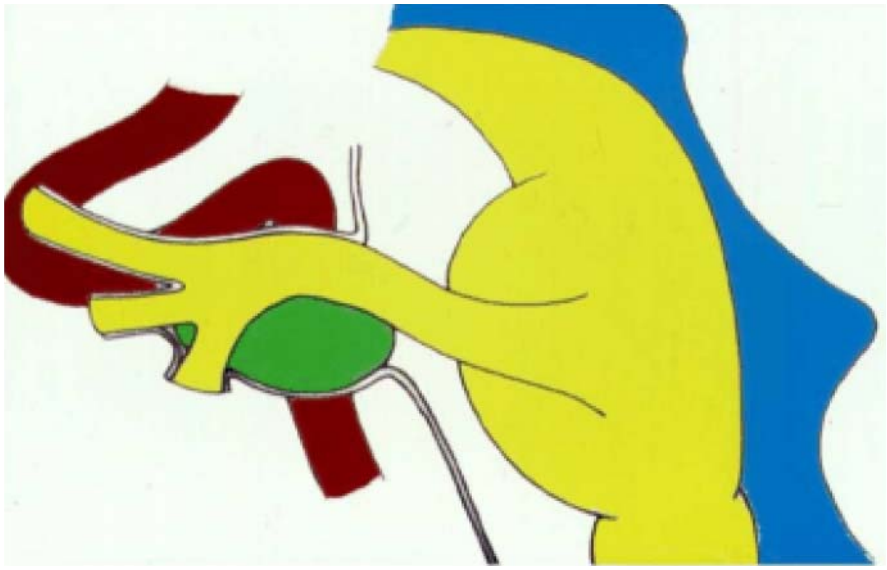


Figure 1c: Line drawing showing a relatively small sized tumor located in the middle fossa.



Clinical Presentation

Table 2 shows the presenting clinical features. There was no significant sex variation in our series, with the male-to-female ratio being 1:1.4. The age of presentation varied from 6 months to 72 years. The age at the time of presentation of 6 months is the youngest in the literature and was reported by us earlier. (23) Neurofibromatosis is less commonly associated with trigeminal neurinomas. (20, 24–27). In our series, 14 patients had features suggestive of neurofibromatosis. Because of the subtle nature of symptoms extending over long periods of time and neglect of early symptoms, probably as a result of illiteracy and ignorance in some of our cases, the majority of the tumors achieved a large size before being diagnosed. The clinical presentation was commonly in the form of paresthesiae or numbness, usually in more than 1 division of the nerve. Severe or neuralgic pain is uncommon and was seen in 9 cases. Wasting of the temporalis and pterygoids was frequent

in 83.3% cases; when present, it was usually diagnostic.

The corneal reflex was depressed or absent in 80 (51%) patients. Dense motor and sensory involvement, including complete absence of corneal sensation, is uncommon and often suggests malignant change in the tumor. Although symptoms of involvement of adjoining cranial nerves in the cavernous sinus and cerebellopontine angle have been reported frequently in the literature, they formed a predominant group, probably because of the large size of the tumors encountered in this series. Twelve patients presented with the rarely encountered symptom of proptosis. The unusual symptom of pathologic laughter was seen in 12 cases of large and dumbbell shaped tumors. (28) Although the exact physiologic cause of the pathologic laughter remains to be defined, on the basis of a literature survey, it seems that the complex combination of extra-axial displacement of the brainstem and medial temporal lobe structures could result in this symptom. It appears that pathologic laughter is an important and possibly early presenting symptom in cases of massive trigeminal schwannoma. The clinical features of slow progressive symptoms and the predominant presence of trigeminal nerve–related symptoms of numbness and wasting of muscles are usually diagnostic. In 5 cases, there was difficulty in mastication that was related to presence of lump in the mouth.

Radiologic Features

The size of the tumors in our series ranged from 8 mm to 7.8 cm (Fig. 2). The physical characteristics of the tumors are shown in Table 3. Bilateral trigeminal neurinomas have only rarely been reported. (27) In this series, there were 6 bilateral trigeminal neurinomas, all associated with features of neurofibromatosis. The contralateral tumor in cases with bilateral trigeminal schwannomas was nonsymptomatic in all cases (Fig. 3). Tumors limited to the middle fossa and dumbbell-shaped tumors occupying both the middle fossa and posterior fossa were more common than tumors located only in the posterior fossa. All tumors with extension into the extracranial compartment had a middle fossa component. Tumors extending into the orbit were seen in 4 cases, extension along the maxillary division of the nerve was seen in 5 cases, and extension along the mandibular division in 13 cases. In 6 cases the tumor was identified in the infratemporal/pterygopalatine fossae, but the exact division of the nerve of origin could not be clearly identified. In two cases the tumor extended into the orbit and probably arose from the lacrimal division of the fifth cranial nerve (Fig. 4). (29) The usual form of trigeminal neurinoma was relatively soft and moderately vascular. The predominant presence of cystic changes was seen in a number of tumors, and there were multiple cysts in 16 tumors. The presence of multiple cysts, although rarely seen in acoustic schwannomas, (30) has never been reported with trigeminal schwannomas. Fluid-fluid level within the tumor cysts was encountered in 8 cases. (Fig. 5) Calcification in trigeminal schwannoma has never been reported but was encountered in 6 tumors. The erosion of the petrous apex and smooth circumferential erosion of the foramen ovale, rotundum and superior orbital fissure on plain radiography or computerized tomography was uniformly observed in larger tumors, and these findings were of diagnostic significance. Although there have been reports of malignant trigeminal schwannomas, (6, 26, 31–34) none of the patients in our series had a malignant schwannoma. Transgression of the medial dural wall and actual invasion into the venous spaces of the cavernous sinus or encasement of the precavernous sinus and cavernous sinus–related internal carotid artery was not encountered in any case in our series but has been reported earlier. (8, 35, 36) The displacement of the internal carotid artery was characteristic and had important diagnostic value. (37) In the posterior fossa, the tumor was located anterolateral to the pons and, like acoustic neurinoma, was "extraarachnoidal" in nature in majority, but 'interdural' in some cases.

Surgical Strategy

Surgery for trigeminal schwannomas has evolved along with the evolution of skull base surgical techniques.(20,38) The anatomy of the tumor and its dural covers, anatomy of the region, and anatomy of various approach routes are now better understood. Trigeminal

Figure 2a: Axial computed tomography showing the large dumbbell- shaped trigeminal schwannoma.

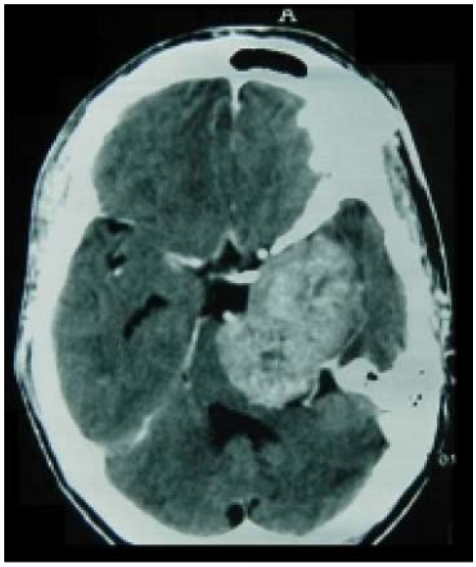


Figure 3: Coronal scan showing bilateral trigeminal neurinoma.

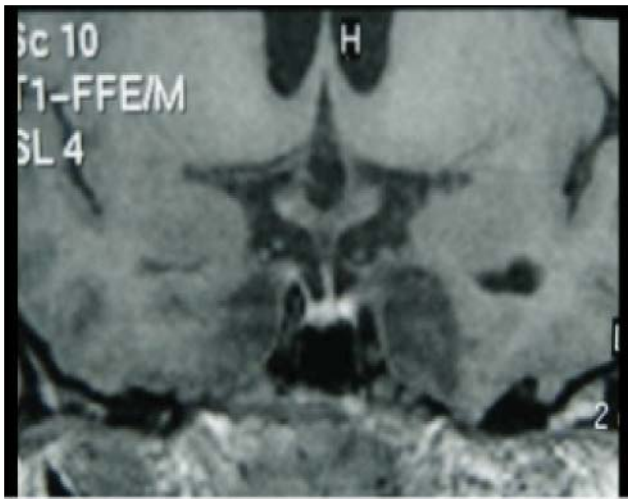


Figure 5a: Axial MRI showing the trigeminal neurinoma. The tumor has cystic necrosis and there are multiple fluid-fluid levels within the tumor.

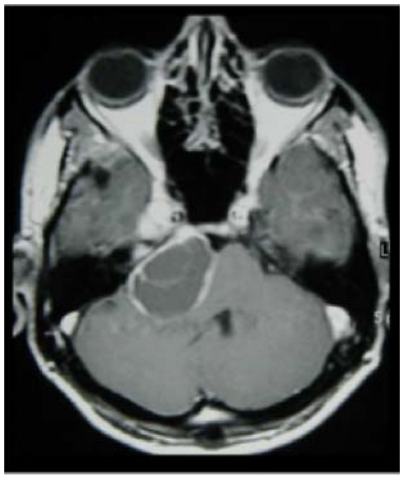


Figure 2b: Postoperative magnetic resonance imaging showing complete excision of the tumor.

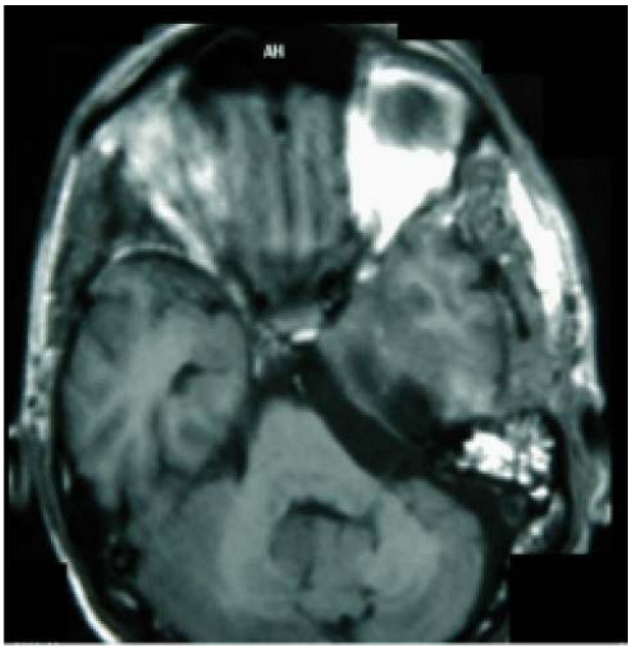


Figure 4: Sagittal T2-weighted magnetic resonance imaging (MRI) showing the hyperintense trigeminal schwannoma extending from the cavernous sinus to the orbital region.

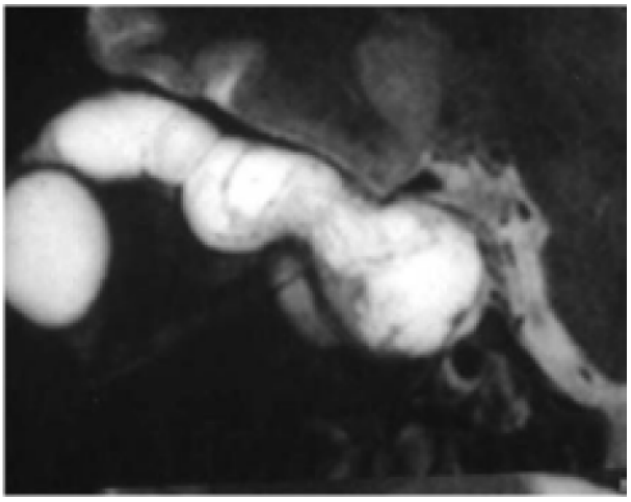
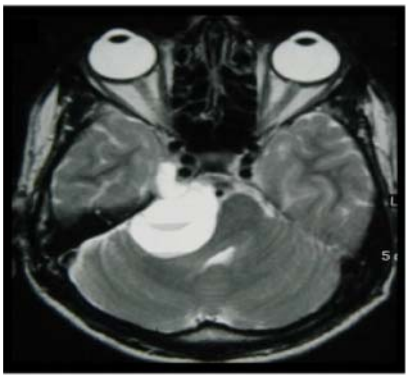


Figure 5b: Axial T2-weighted scan showing the tumor and the multiple fluid-fluid levels vividly.



neurinomas can now be removed by relatively small and straightforward exposures with minimum brain handling.

Various reports have stressed the need for radical surgery, because total resection leads to cure from the tumor and the recurrence rate for cases with partial resection, particularly of cystic tumors, is relatively higher for trigeminal neurinomas than for acoustic neurinomas. (39)

The major impediment to complete removal is an inadequate exposure. Because of their location near the midline and close proximity to vital neural and vascular structures, the ideal surgical approach should be the shortest and most direct; it must be wide and low so as to avoid the need for prolonged brain retraction. It is also necessary that the exposure part of the operation itself does not become unduly time-consuming, tedious, and complex and affect or threaten the anatomical and functional integrity of cranial nerves, major arterial or venous channels, or joints. Toward these objectives, various modifications to the classic approaches have been described.

Even a little extra exposure and working space can sometimes be crucial for radical resection of the tumor as well as for the safety of the patient. The following anatomy and characteristics of trigeminal neurinomas guide the philosophy as regards operative approaches to excise these tumors radically: 1. Trigeminal neurinomas are more frequently soft and suckable tumors. These features make surgery on these lesions relatively easy. Occasional cases of highly vascular and tough tumor may also be encountered. The physical nature of the tumor is more variegated in tumors having an extracranial extension. 2. Trigeminal neurinomas can frequently be diagnosed

on the basis of the presenting clinical and radiologic features. The characteristic location at the petrous apex, dumbbell-shaped appearance, and erosion and widening of the cave of Meckel are frequently seen. The most characteristic feature is the relation between the precavernous and cavernous segments of the carotid artery. Because of the anatomical relation between the carotid artery and the Gasserian ganglion, the precavernous carotid artery is located on the inferior surface of the tumor, whereas the cavernous carotid is displaced medially. 3. Although the carotid artery is in close approximation to the tumor, there is a well-defined dural sheath separating the carotid artery and the tumor. If one remains in the confines of the tumor capsule, there is little danger of injury to the carotid artery. As a result of this anatomical feature, there is seldom any need for perioperative control of the carotid artery. Even the extracranial component of the tumor is surrounded by a relatively tough dural layer that separates it from the adjoining critical neural and vascular structures. 4. Because of its location in the lateral wall of the cavernous sinus, the tumor displaces the venous plexuses and never invades into the venous spaces. If one remains in the tumor capsule, one may seldom encounter venous bleeding. 5. The tumor does not involve all the fibers of the nerve. Some fibers are invariably spared. These fibers can usually be preserved. Working within the confines of the tumor capsule, using blunt dissection with the help of suction or Cavitron ultrasonic aspiratory (CUSA), and avoiding coagulation as much as possible could avoid injury to these fibers. 6. In the posterior fossa, the tumor has a well-defined plane of cleavage from the brainstem, adjoining cranial nerves, and blood vessels. 7. The location of the middle fossa and extracranial components of the tumor is "interdural," whereas the posterior fossa part is "intradural." in majority and 'interdural' in some. This information is useful when dissecting the tumor. A thick dural wall in the middle fossa covers the tumor; hence, dissection is relatively safe in this portion of the tumor. 8. Intratumoral cystic and necrotic changes are common in trigeminal neurinomas, usually making the central part of the tumor soft, and such tumors can be removed with gentle suction or CUSA. 9. Some tumors are firm and fibrous, and some are vascular. Firm tumors can be excised using limited exposure, but the dissection can be relatively difficult in such tumors. One has to restrict the dissection intratumorally and debulk the lesion as much as possible before dissecting the capsule. 10. The retrosigmoid approach is useful for tumors limited to the posterior fossa. Retrosigmoid exposure of the posterior fossa portion of the dumbbell-shaped tumor can be

avoided in most instances. The lateral basal subtemporal approach can provide satisfactory exposure to the entire tumor. 11. A 2-staged approach can generally be avoided. Approaches involving sectioning of the transverse sinus can only be condemned in present-day neurosurgery. 12. Preoperative lumbar drainage of cerebrospinal fluid can be used to great advantage during surgery by means of the lateral basal subtemporal approach. The procedure relaxes the brain and helps by increasing the exposure and protection of the vein of Labbé. 13. Tumors extending into the extracranial compartment can be resected by a 'reverse skull base approach'. The strategy of limited basal temporal craniotomy and retraction of the brain to expose the extracranial component of the tumor can be effectively used for tumor resection.

Operative Approaches

Various operative approaches have been described for the surgical resection of trigeminal neurinomas. (40–42) Skull base techniques have been used to provide for a low and wide exposure and limit the need for brain retraction. This has resulted in a higher percentage of tumor resection, a low surgical morbidity rate, and a lower rate of recurrence. (8, 11, 12, 20, 24, 35, 40) The approaches preferred by the authors are discussed.

Infratemporal Fossa Interdural Approach

Indications

In smaller tumors limited to the middle cranial fossa and where the posterior fossa component of a dumbbellshaped tumor is relatively small, an infratemporal fossa interdural approach can be used (Table 5).

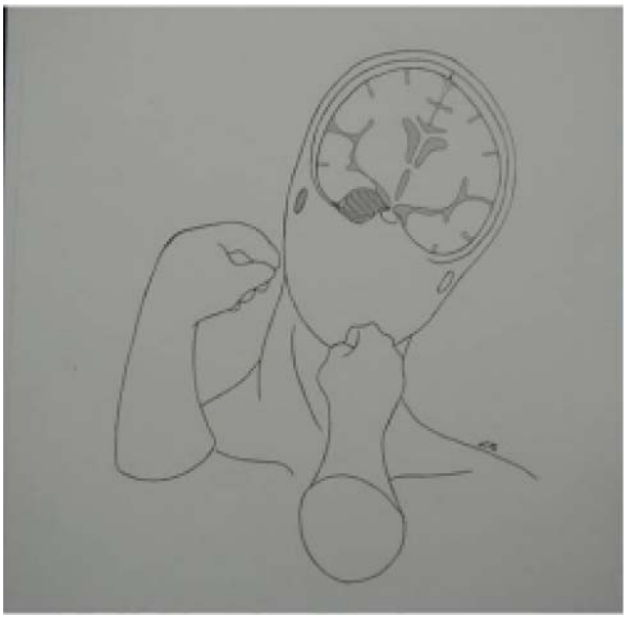
Surgical Technique

The patient is positioned so that the head is extended and turned to the contralateral side. The right-handed surgeon stands on the right side at the level of the chest of the patient irrespective of the side of the lesion and suitably alters the angle of the rotation of the head (Fig. 6). A linear incision is taken over the zygomatic arch, which is completely resected. The temporalis muscle is either split in the direction of the incision or can be reflected superiorly or inferiorly. The muscles of the infratemporal fossa are dissected off the bone by sharp subperiosteal dissection, and the foramen ovale is exposed. With a microdrill, a small craniectomy incorporating the foramen ovale and measuring approximately 3 cm × 3 cm is made in the infratemporal fossa (see Fig. 6). An incision is taken on the lateral surface of the dural sheath of the mandibular nerve at the level of the foramen ovale and extended posteriorly to the inferior and lateral surface of the dural sheaths covering the Gasserian ganglion. The dura over the Gasserian ganglion and the lateral wall of cavernous sinus are reflected, exposing the middle fossa part of the tumor (temporal lobe exposure and a middle fossa floor durotomy are avoided). The bulk of the tumor usually dilates the dural sheaths in each case, and a large exposure can be obtained. After debulking the tumor, the exposure can be further widened. The tumor is followed in the posterior fossa along the cave of Meckel. A large tumor in the region of the Gasserian ganglion is seen to dilate the cave of Meckel, thus providing sufficient exposure to the part anterior to the brainstem. The cavernous sinus part of the tumor can usually be resected by anterior angulation of the microscope. The infratemporal fossa interdural approach uses an infratemporal fossa exposure after sectioning of the zygomatic arch. The association of atrophy of the temporalis and pterygoid muscles with trigeminal neurinomas makes the dissection in the infratemporal fossa relatively easier and the exposure wider. Al-Mefty et al (43) described sectioning of the temporalis muscle at its insertion at the coronoid process and superior displacement of the muscle. The temporalis muscle can also be split in the direction of its fibers or reflected inferiorly and later used for basal reconstruction. (44) The direction of approach and the surgeon's position as regards the patient can be altered to provide direct and low access to the lesion. The route of approach is interdural, avoiding the need to expose the temporal lobe and thus limiting the extent of temporal lobe retraction to the minimum. The possibility of anatomically dissecting the layers of the dura in cases of trigeminal neurinoma has been described earlier. Although the carotid

artery at the petrous apex is not exposed because a dural sheath usually covers it, it is close in the field and can be exposed relatively easily. The tumor bulk widens the cave of Meckel, and a large window can be obtained for the resection of the posterior fossa portion of the tumor. The soft nature of the tumor can be used to circumvent the disadvantage of confronting the tumor before exposure of the brainstem. The approach to the tumor anterior to the brainstem is entirely infratentorial. In our series, the exposure obtained by such an approach was seen to be adequate for safe and complete resection of the tumor. In none of our cases was there any need to extend the exposure for tumor removal. The entire procedure could be performed in a significantly shorter time and was cosmetically appealing.

Figure 6: Line drawing with the surgeon's hands showing the direction of approach. Shaded ipsilateral zygomatic arch and infratemporal fossa bone depict the extent of bone removal for the trigeminal schwannoma shown by oblique lines. The zygomatic arch will be replaced at the end of the operation.

Figure 7: Line drawing showing the scalp incision for the lateral basal subtemporal approach.



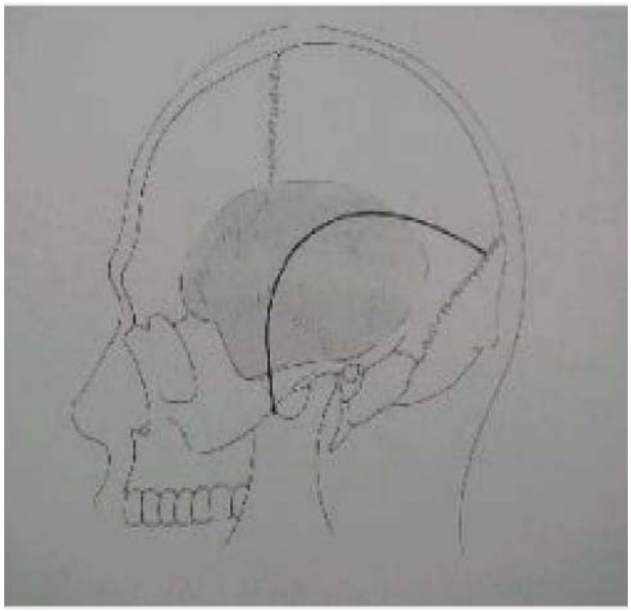
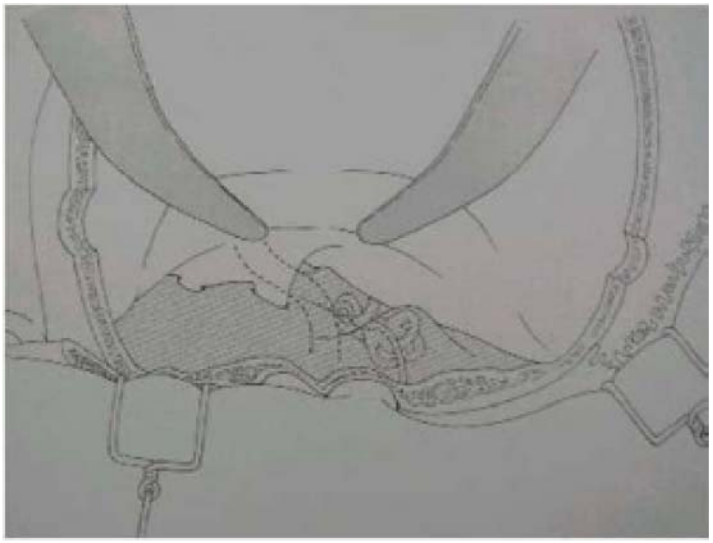


Figure 8: The condyle and superior half of the external canal are unroofed, and the superior third of the mastoid air cells has been drilled to obtain a basal extradural exposure. After elevation of the middle fossa dura, exposure of the foramen spinosum, foramen ovale, foramen rotundum, dura covering the cave of Meckel, and anterior aspect of the petrous bone has been obtained.



Basal Lateral Subtemporal Approach

Indications

For large middle fossa tumors and dumbbell-shaped tumors, a subtemporal craniotomy centered on the external ear canal was found to be suitable to deal with larger tumors located in either or both the middle and posterior cranial fossae. This approach has the advantage of being simple and relatively quick, and general neurosurgeons are familiar with it.

Surgical Technique

The patient is placed in a lateral position. A continuous external drainage of cerebrospinal fluid by lumbar subarachnoid catheter placement is set up. The scalp incision is shown in Figure 7. It starts from a point about 1.5 cm to 2 cm anterior to the tragus of ear and about 1.5 cm inferior to the zygomatic arch. The incision is anterior to the trunk of superficial temporal artery. Working deep to the deep layer of temporalis and masseteric fascia and displacing the soft tissues harboring these tiny nerves anteriorly protect the frontal and zygomatic branches of the facial nerve. The incision curves initially superiorly and then traverses posteriorly. The incision exposes the squamous temporal and posterior parietooccipital bone, posterior third of the temporalis muscle, roots of the zygomatic arch, supramastoid crest, and base of the mastoid process. The incision can be extended further posteriorly and curved inferiorly to enhance the temporal, occipital, and mastoid process exposure. The wide base of the scalp flap and preservation of all feeding arteries ensure its adequate vascularity. The posterior aspect of the temporalis muscle is mobilized in the subperiosteal plane from the temporal bone and from the sharp superior border of the zygomatic arch. The muscle is then rotated anteriorly. A low temporal craniotomy with the base centered on the external ear canal is performed. The anterior and posterior roots of the zygomatic arch, the glenoid fossa, and the lateral half of the roof of the external ear canal are removed in a single piece with the help of a power-driven saw or are resected with the help of rongeurs and a power-driven drill (Fig. 8). Removal in a single piece is frequently difficult and can result in the inadvertent opening up of the external ear canal, a procedure that could affect the sterility of the field. The external ear canal is protected by sharp subperiosteal separation of the canal from the bony roof. The external ear canal has loose fibrous connections to the bony and cartilaginous wall. It is more firmly attached to the spine of Henle, where sharp dissection may be necessary to expose the canal. The meniscus of the temporomandibular joint is exposed but not removed. In tumors limited only to the middle cranial fossa or in smaller lesions, the glenoid fossa may not be removed and mastoidectomy can be avoided. The superior third or half (about 1.5–2 cm below and medial to the supramastoid crest) of the mastoid air cells is drilled. The mastoid antrum may or may not be opened. The drilling of the mastoid process may be continued medially to expose the bony labyrinth around the superior and posterior semicircular canals. The sigmoid sinus and the region of its junction with the transverse sinus are not exposed, and a thin plate of bone is left between the sinus and the mastoid exposure. The dura can now be elevated off the middle fossa floor after sectioning the middle meningeal artery, and a basal extradural exposure is obtained as shown (Fig. 8). An entirely extradural route can remove the tumors limited to or having a larger bulk in the middle fossa after exposing the foramen ovale and dissecting the outer sheath of the dura. In large tumors and those with a significant posterior fossa component, however, an intradural exposure is preferred. By an intradural route and elevation of the temporal lobe, the middle fossa floor and the tentorium are exposed. The bulge of the tumor under the dura of the middle fossa floor is identified. A transverse incision is made over the bulge of the dura, and the tumor within the dural walls is progressively removed, saving the displaced normal trigeminal nerve fibers. An incision is made in the tentorium, which begins at its free edge at the level of posterior end of the cerebral peduncle and is then directed anterolaterally toward the lateral aspect of the superior petrosal sinus. A triangular flap of tentorium is then everted over the superior petrosal sinus, thus providing a wide window from the subtemporal view to the infratentorial structures. By this maneuver, the fourth and fifth

cranial nerve fibers and petrosal vein are protected from inadvertent injury and are exposed widely. The tentorial dural flap is then resected by cutting parallel to the superior petrosal sinus or everted over the middle fossa floor. The posterior fossa component of the trigeminal neurinoma is now exposed. Its consistency, vascularity, and extensions are examined, and the need for further exposure is assessed. Taking an incision exposes the tumor at the petrous apex over the superior dural cover of the cave of Meckel after transecting and packing or clipping the cut ends of the superior petrosal sinus. With angulation of the microscope, the tumor in the lateral dural walls of the cavernous sinus anteriorly and the

cerebellopontine angle posteriorly can be exposed. The entire approach is from the middle cranial fossa. Whenever necessary, expansion of the exposure is possible in various directions. A tumor located in the region of the cave of Meckel and extracranial tumors never transgressed the dural confines, a feature that assisted in developing a plane of dissection of the tumor from the venous spaces of the cavernous sinus and carotid artery and from structures in the infratemporal fossa. Tumor resection is begun from its lateral and inferior aspect in the region of third division of the fifth nerve and then proceeds superiorly, taking special care to avoid injury to the first division of the fifth nerve. After the procedure, the mastoid air cells are packed with bone wax, free muscle, or fat graft. The posterior third of the temporalis muscle along with its fascia can be rotated to the base for strengthening the reconstruction and for providing a vascularized pedicle. If preserved, the bone piece harboring the roots of the zygomatic arch, the glenoid fossa, and the lateral aspect of the roof of the external ear is replaced and sutured along with the craniotomy bone flap. The external auditory meatus is packed with cotton pledget to avoid cicatricial stenosis. The subtemporal or a middle cranial fossa approach has frequently been used for the treatment of vascular lesions and tumors located in the petroclival and clival regions. A common disadvantage of this approach has been damage to the temporal lobe caused by retraction, particularly when the venous drainage is interrupted. The more commonly used methods of basal expansion of a low temporal craniotomy to reduce retraction-related brain injury include a zygomatic osteotomy and inferior mobilization of the temporalis muscle and resection of the middle fossa floor. Partial unroofing of the external ear canal and inferior mobilization or resection of the condyle of the temporomandibular joint have also been recommended for enhancing the inferior angle of vision. A modified basal extension of the lateral subtemporal approach includes resection of the root of the zygomatic arch, roof of the external ear canal, and superior third of the mastoid bone. The inclusion of a mastoidectomy in the exposure adds to the advantages of the more popularly employed petrosal approach to these tumors. The basal extension of the conventional subtemporal or middle fossa approach is frequently carried out by means of a zygomatic osteotomy, which facilitates the inferior displacement of the temporalis muscle. Some reports indicate the usefulness of resection of the middle fossa floor. Such approaches are anterior subtemporal, however, and the working angle to the petrous apex is about 40° to 60° from the horizontal plane in the line of the external ear canal. The posterior subtemporal approach is performed after a mastoidectomy, usually in combination with a presigmoid approach. In such a posterior subtemporal approach, the vein of Labbé obstructs the exposure. A direct lateral approach in the line of the external ear canal, which is almost transversely oriented, is the shortest route to the petrous apex. A basal subtemporal approach has the advantage of ease of working in both the middle fossa and infratentorial compartment and is suitable for all varieties of trigeminal neurinomas. The technique of basal extension of the temporal craniotomy improves the horizontally wide exposure directly in line with the petrous apex. It does not affect hearing or involve facial nerve exposure or manipulation. The function of the temporomandibular joint is not affected, and because there is no manipulation or displacement of the condyle. there is no postoperative pain during mastication. Because the bone in the lateral aspect of the subtemporal route is removed, the operating distance to the

tumor is reduced by about 1.5 cm to 2 cm. Additional inferior and posterior space of about 1.5 cm to 2 cm is also available after a mastoidectomy and removal of the roof of the external ear canal and glenoid fossa. The external ear canal and the condyle and meniscus can be gently depressed inferiorly for this purpose whenever necessary. This space and the relaxation of the temporal lobe after drainage of cerebrospinal fluid provide sufficient exposure for manipulation of instruments and safe dissection of tissues under direct vision and a suitable angle. None of our patients developed any retraction related temporal lobe damage. For the middle fossa component, the inferolateral operative route was found to be considerably safer for preservation of the first division of the fifth nerve than an anterolateral trajectory available by means of a frontotemporal approach. The lateral route also seemed to be safer for the dissection of the tumor from the brainstem because it could be done under vision. The other advantage of the described approach is that it is technically relatively easy. There is no need to expose the superior bend of the sigmoid sinus, which is posterior in the line of the operative direction. The elaborate exposure necessary for mobilization of the sigmoid sinus (as is necessary in the petrosal approach) is avoided. This makes the exposure significantly quicker and safer. The temporalis muscle is displaced anteriorly and thus away from the operative field, and resectioning of the entire zygomatic arch for this purpose is unnecessary. Anterior, posterior, medial, and inferior expansion of the initial exposure is relatively easy. Reconstruction of the exposure is easy and compact and can be done with the help of vascularized pedicled temporalis muscle–based flaps. With the advantages listed here, a basal lateral subtemporal approach appears to be useful for the exposure of trigeminal neurinomas. An additional partial mastoidectomy enhances the scope of the basal subtemporal approach. The ease of expansion of the exposure before or during tumor resection provides versatility to the approach.

However, basal extension of the temporal craniotomy was found to be not necessary in the several cases done in the later part of the series. For over 6 years, basal extension of the temporal craniotomy was not done for resection of a trigeminal neurinoma, irrespective of their size.

For tumors located in the middle fossa and those that are relatively small in size, an anterior temporal basal exposure is useful and basal extension of the temporal craniotomy is not necessary. Extradural exposure from an anterior perspective is easier than an inferior perspective. The tumor resection should however be initiated in the inferior aspect of the tumor to avoid injury to the first division of trigeminal nerve.

For tumors having an extracranial extension, a basal temporal craniotomy was done to expose the extracranial component of the tumor. The temporal brain was retracted to provide an enhanced angle of vision. Retraction of the brain to expose the extracranial portion of the tumor was labeled as 'reverse skull base' approach. Whenever necessary, in addition to basal temporal craniotomy, zygomatic osteotomy can be done.

Summary of our Surgical Experience

In the early part of the series, a frontotemporal pterional approach with or without an orbitozygomatic osteotomy and petrosal approaches were employed, but in the later part of the series, these approaches were not used. The main operative approaches preferred and used after the period from 1992 through 2009 to resect these tumors were the infratemporal fossa interdural approach,(39) lateral basal subtemporal approach, (45) , anterior basal subtemporal approach and retrosigmoid approach. The operative approaches and outcomes after surgery are shown in Table 4. A sitting position was adopted for the retrosigmoid approach, whereas the remaining patients were operated on in a lateral position.

Lumbar cerebrospinal fluid drainage was done during surgery in the lateral position. In 7 cases, the operation was done in 2 stages, comprising either the frontotemporal or lateral basal subtemporal approach and the retrosigmoid approach. In the later part of the series, all tumors were removed in a single stage and all dumbbellshaped tumors were resected

by means of the lateral basal subtemporal approach (Figs. 9 - 17). The principal operative finding was that the part of the tumor located in the middle fossa was always confined within the dural walls of the cave of Meckel. Transgression of the medial dural wall and invasion into the venous spaces of the cavernous sinus or encasement of the precavernous or cavernous carotid artery were not seen in any case. In the posterior fossa, the basilar artery and its branches and the adjoining cranial nerves were displaced and never encased by the tumor (Figs. 10, 11). Although most tumors were soft, necrotic, and relatively avascular, some were firm and fibrous and some were vascular. Firm tumors could also be excised using a limited exposure, but the dissection was relatively difficult in such tumors. All tumors were well encapsulated. Even in the extracranial compartment, the tumors were well defined and had a thin capsule, which was continuous with the dura in the region of the cave of Meckel (Figs. 18-20). An attempt was made in all cases to restrict the dissection within the mass of the tumor and to debulk the lesion as much as possible before dissecting the capsule. On the basis of the presenting clinical features and characteristic radiologic signs, a diagnosis of trigeminal neurinoma could be made in the majority of cases. Such a diagnosis was crucially important in planning the surgical strategy for the cavernous sinus–related lesion. The relation between the dura and adjoining structures, which is characteristic in trigeminal neurinomas, may not exist for other tumors in the location.

Total tumor resection was achieved in 129 (82%) cases. More than 5% of tumor residue was not left behind in any case. All these patients in whom subtotal excision was achieved were operated on in the early part of the series. Seven patients underwent a 2-stage surgery, which included a combination of a retrosigmoid approach and a subtemporal approach. In the later part of our series (over the last 9 years), total excision was achieved in the majority of cases; when a "near-total excision" was done, only small residual tumor was left behind. We believe that these improved surgical results of resection in the later part of our series are the result of surgical experience, better imaging facilities, and the addition of skull base techniques. During surgery, the fourth cranial nerve was inadvertently cut in 2 patients, the ipsilateral sixth nerve was damaged in 1 patient, and the facial and eighth nerve complex were damaged in 2 patients. Two patients died in the postoperative period. One of these patients died on the first postoperative day. Postmortem examination showed brainstem infarction. One patient died suddenly on the fifth postoperative day even though he was doing well in the immediate postoperative period and the postoperative scan had shown complete tumor resection. Postmortem examination was not done in this case. Among the complications, cerebrospinal fluid leak from the wound was encountered in 1 case and was treated by lumbar drainage of cerebrospinal fluid. One patient developed an acute extradural hematoma that required emergency evacuation. Postoperative meningitis was seen in 1 case and was treated uneventfully with antibiotics. Osteomyelitis of the bone flap requiring bone flap excision was seen in 1 case. Histologic examination of all tumors showed a benign appearance containing both Antoni type A and B tissue. After surgery, 2 patients developed additional sixth nerve paresis and 2 patients had worsened facial weakness and hearing and developed facial palsy and deafness. Sixth and eighth nerve function did not recover, whereas facial function improved significantly but incompletely on follow- up in both patients. One patient (who developed a postoperative sixth nerve deficit) developed keratitis and permanent corneal opacity 2 months after the surgery. There was no other major morbidity in the series. All patients with preoperative facial numbness remained with some degree of trigeminal hypesthesia. Facial sensation subjectively improved after surgery in majority of patients; however, it was not possible to quantitatively ascertain this fact. No patient with preoperative normal facial sensation developed numbness in the face. Thirteen patients worsened in sensation over the face and developed complete anesthesia in 1 or more divisions of the fifth nerve. Majority of patients had at least subjective improvement in the power of the temporalis and masseter muscles. Wasting of the muscle did not improve in any case. All 12 patients with pathologic

laughter were relieved of this symptom immediately after surgery. On follow-up ranging from 6 months to 19 years (with the average being 52 months), all surviving patients have shown improvement in their preoperative symptoms and functional ability, have resumed their occupation, and are independent. There was a nonsymptomatic recurrence in the region of the cavernous sinus 8 years after surgery in a patient with a massive dumbbell-shaped multicystic tumor. The recurrence is being clinically and radiologically observed. Four cases having extracranial extension had recurrence. Two of these patients were reoperated. Of the cases with bilateral tumors, only the symptomatic tumor on 1 side was treated surgically. No tumor growth has been observed on the nonoperated side during the follow-up period in any case. We found that cystic tumors operated on earlier by only cyst evacuation and small resectioning of the tumor capsule were more prone to recurrence.

Huang (46) evaluated the role of stereotactic radiosurgery for trigeminal neurinomas. He found it to be an alternative primary or adjuvant strategy that controlled tumor growth, did not

Figure 9a: T2-weighted image of an 18 year old boy showing a large dumbbell shaped trigeminal neurinoma.

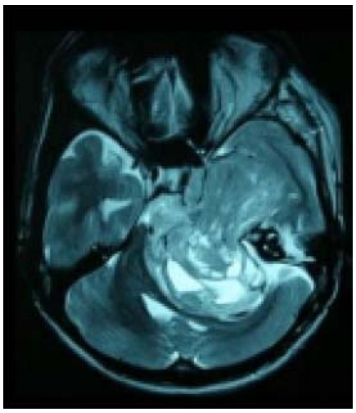


Figure 9b: Sagittal image showing the extracranial extension of the tumor.

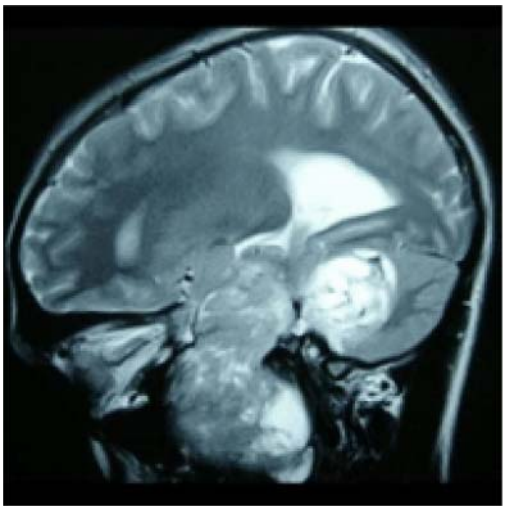


Figure 9c: Coronal image showing the multi-compartmental nature of the tumor.

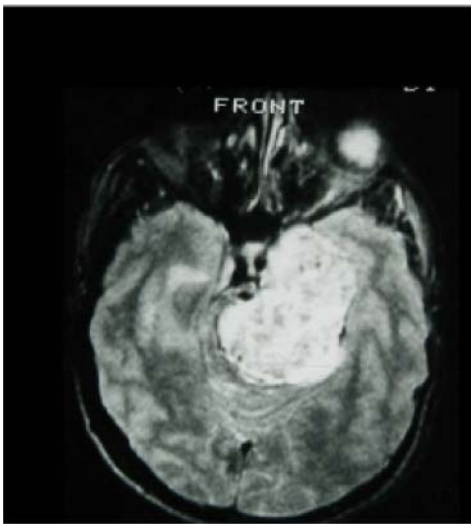


Figure 10a: MRI showing the large dumbbell shaped tumor.

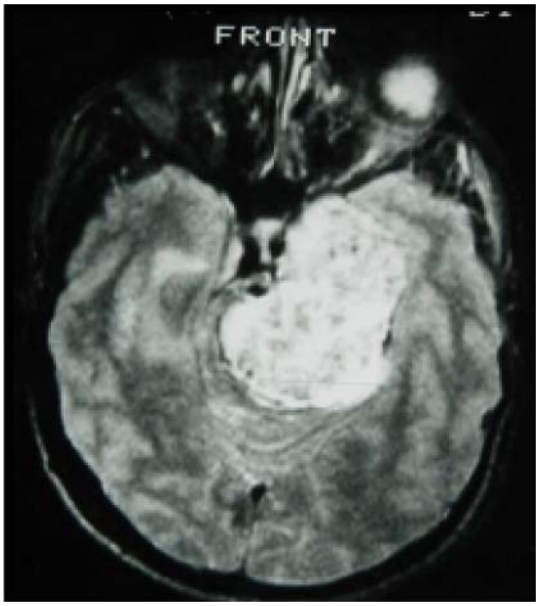


Figure 10b: T1-weighted image showing the tumor.

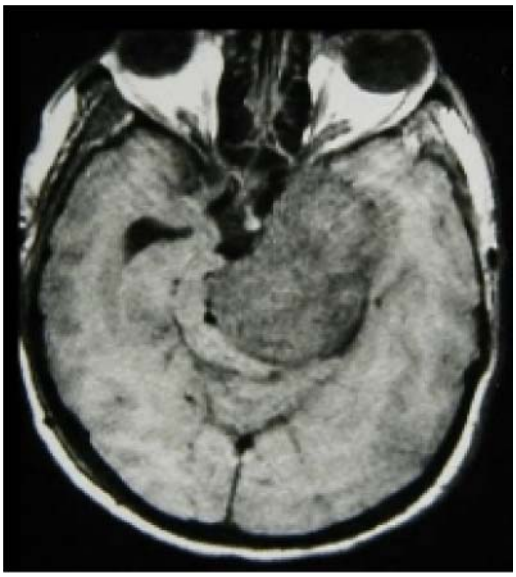


Figure 10c: Sagittal scan showing the extensions of the tumor.

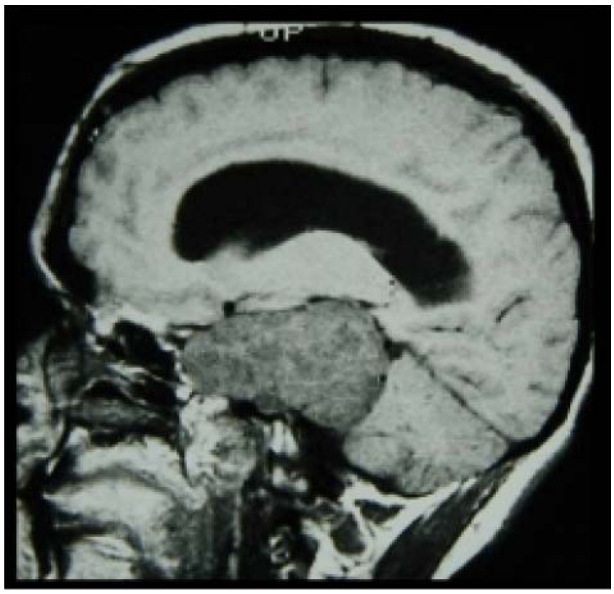


Figure 10d: Postoperative scan showing the tumor resection. The resection was done in two stages.

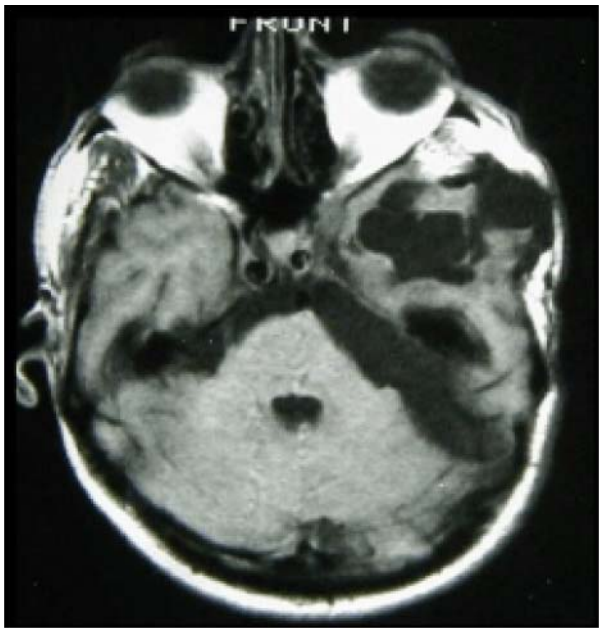


Figure 11a: Sagittal T1-weighted MRI showing the large multi-compartmental tumor in a 36-year old female patient.

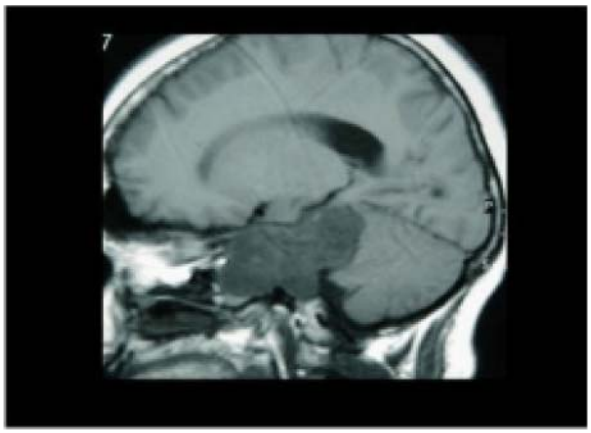


Figure 11b: Coronal image showing the tumor.

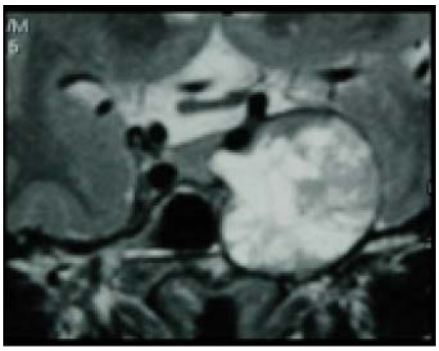


Figure 11c: postoperative scan showing the tumor resection.

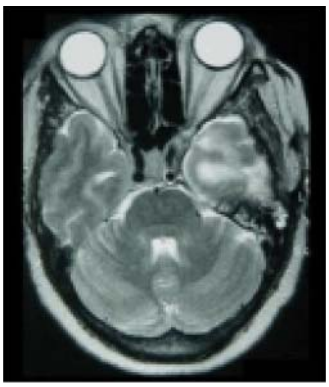


Figure 11d: Coronal scan showing the tumor resection.

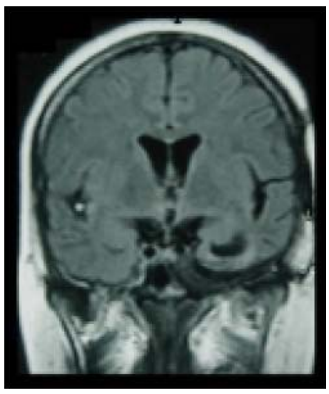


Figure 12b: T1-weighted MRI showing the tumor.

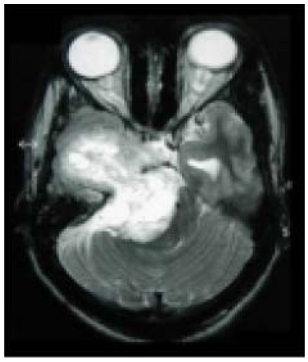


Figure 12d: T2-weighted scan showing the tumor resection

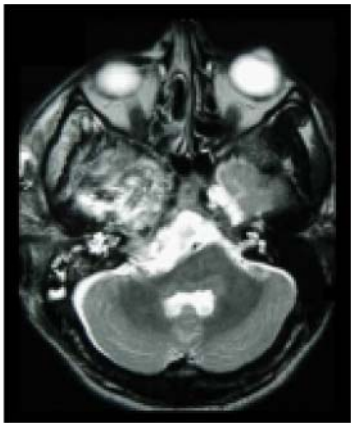


Figure 12a: Axial T1-weighted MRI showing the large dumbbell shaped tumor.

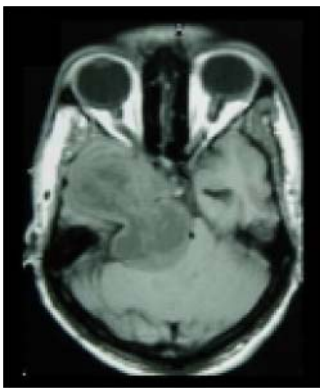


Figure 12c: T1-weighted postoperative scan showing the tumor resection.

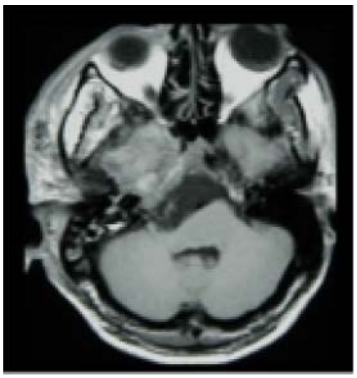


Figure 13a: Axial T1-weighted magnetic resonance imaging (MRI) showing the large dumb-bell shaped tumer.

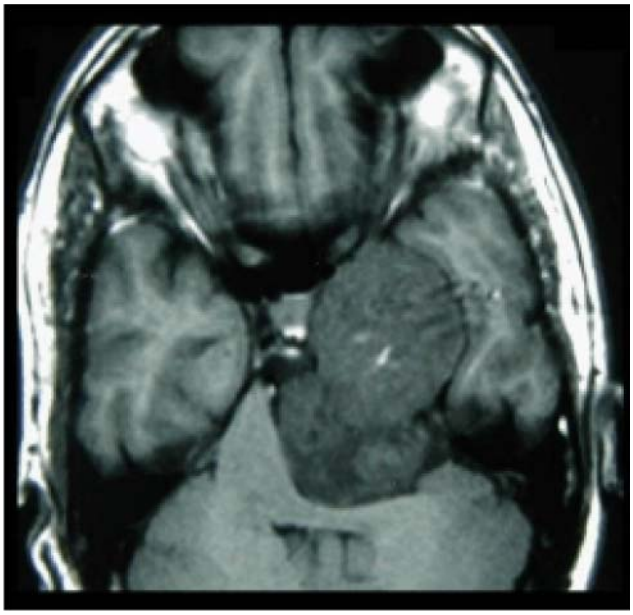


Figure 13b: Axial contrast-enhanced MRI showing the intense enhancement of the dumbbell-shaped trigeminal schwannoma.

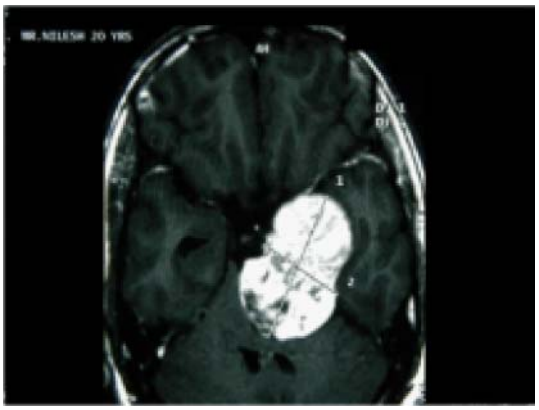


Figure 13c: Coronal image showing the intensely enhancing tumor.

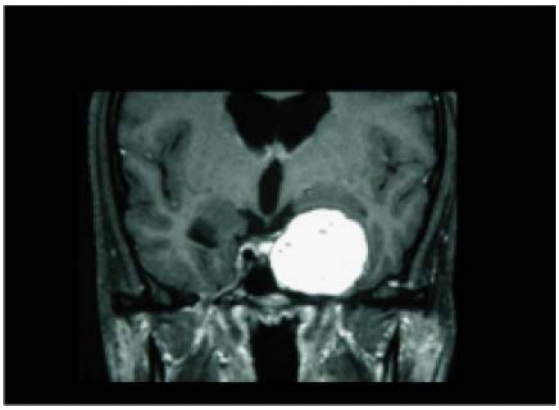


Figure 13d: Postoperative T2-weighted MRI showing complete excision of the tumor.

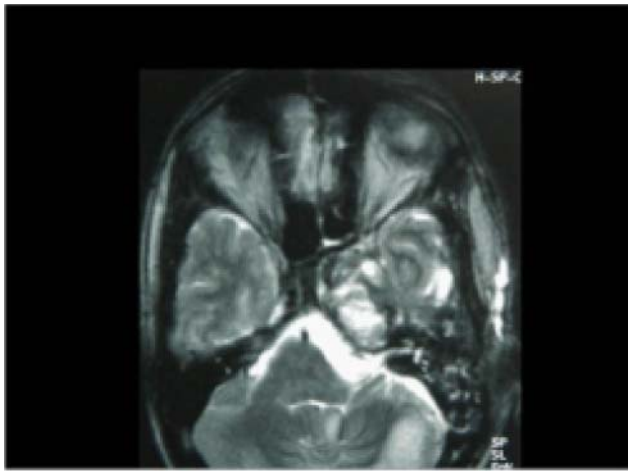


Figure 14a: Axial T1-weighted magnetic resonance imaging (MRI) showing the large predominantly trigeminal schwannoma extending into the orbit, sella, and middle fossa.

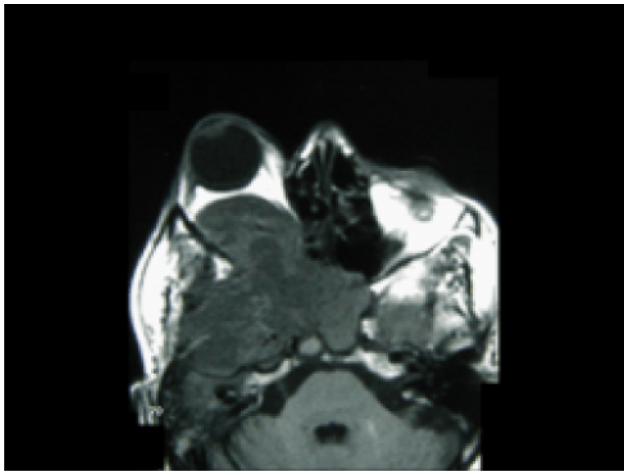


Figure 14b: Axial T2-weighted MRI showing multiple hyperintense cystic areas within the tumor.

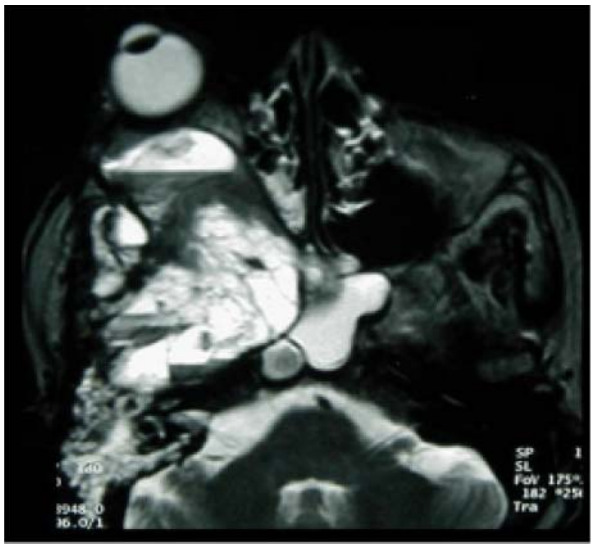


Figure 15a: Axial MRI showing a massive dumbbell shaped trigeminal neurinoma.

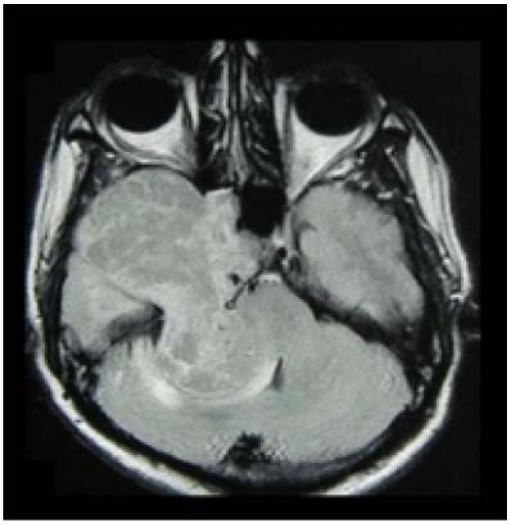


Figure 15b: T2-weighted axial MRI showing the tumor and its extensions. Postoperative scan showing te tumor resection.

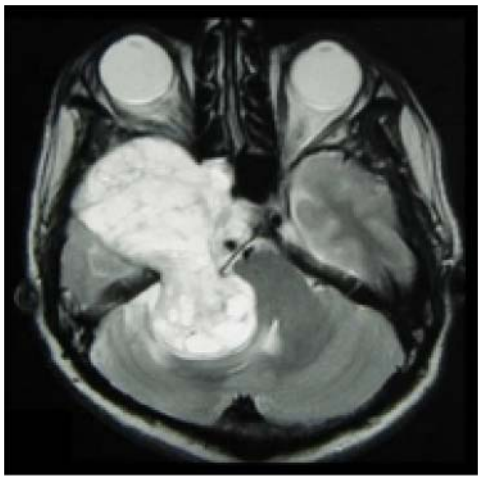


Figure 16a: MRI of a 19-year old girl with NF2. It shows multiple neurinomas that include fourth, fifth and bilateral seventh and eighth neurinomas.

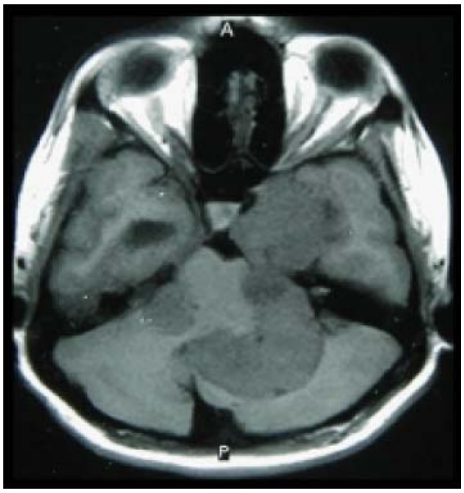


Figure 16b: Contrast enhanced scan showing the lesions vividly.

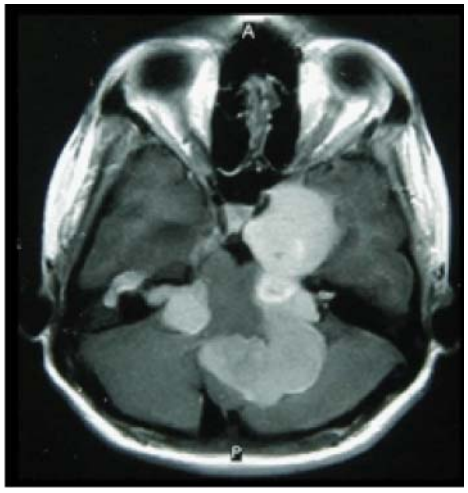


Figure 17: Axial MRI showing a relatively small trigeminal neurinoma.

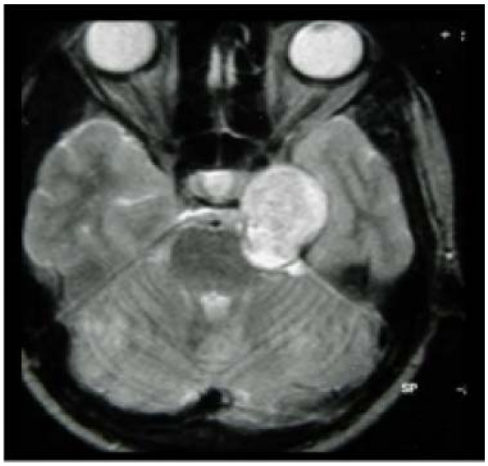


Figure 18a: Coronal T2 weighted MR image showing an extracranial trigeminal neurinoma extending into the pterygopalatine fossa along the foramen rotundum.

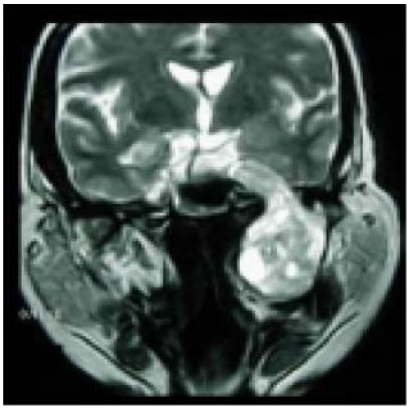


Figure 18b: Axial T2- weighted image showing the tumor and its extension along the maxillary division of the nerve.

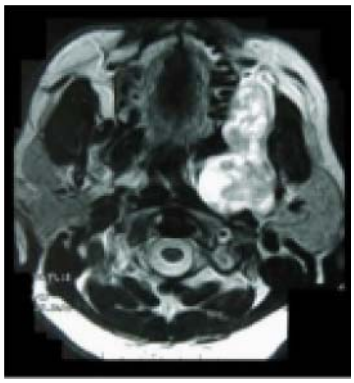


Figure 19b: Sagittal T2 –weighted MR image showing the tumor.

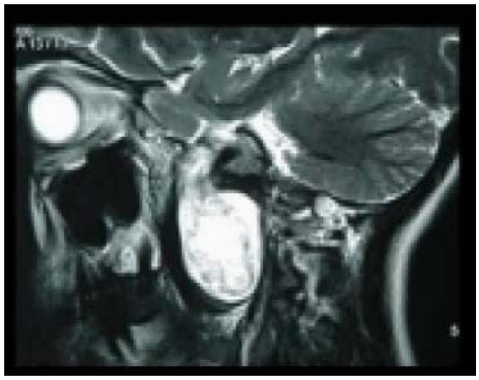
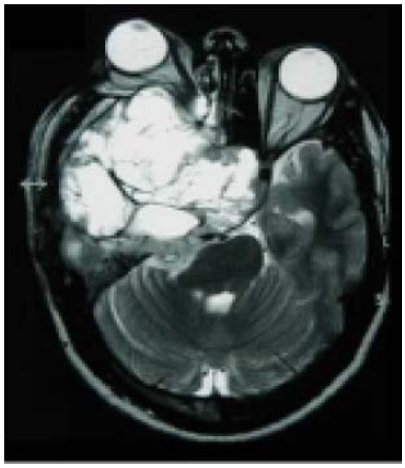


Figure 20b: Axial T2-weighted image showing the marked necrotic nature of the tumor.



cause new deficits, had improved rates of cranial nerve preservation, and often improved presenting symptoms. This therapy was not employed in our series, even in cases with demonstrated residue. On the basis of our experience, we believe that radical tumor

Figure 19a: Coronal T1-weighted MR image obtained in a 42 year old woman showing the trigeminal neurinoma extending into the infratemporal fossa along the mandibular division of the nerve.

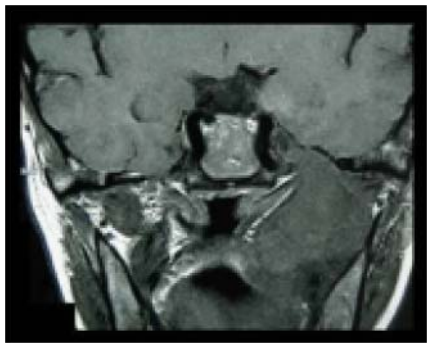
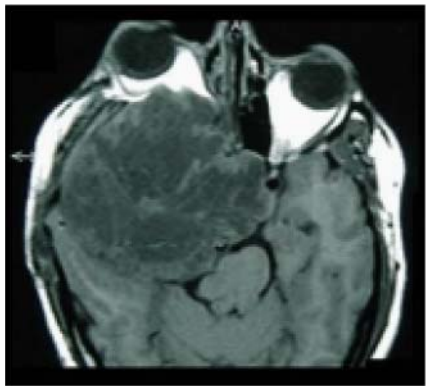


Figure 20a: Axial T1-weighted MR image obtained in a 30-year-old man showing a large trigeminal neurinoma extending up to the temporal convexity and into the orbit. Note the proptosis.



surgery for trigeminal neurinomas is safe and that excellent neurologic outcome and long-term tumor control can be expected.

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