Trigeminal Nerve Tumors of the Lateral Skull Base

Abstract—A retrospective review of 8 trigeminal nerve tumors centered in the lateral skull base was carried out. Clinical manifestations, radiographic features, and outcome of management are detailed. Six patients in this series presented for primary surgery, and 2 patients had prior surgical interventions with incomplete resections.

Surgical exposure via the infratemporal fossa (ITF) approaches resulted in complete tumor extirpation in all but 1 of the reoperated cases. Limited postoperative cranial neuropathies and the lack of intracranial complications are particularly notable in this series.

A mean follow-up of 3 years revealed no evidence of recurrent disease. In the case of incomplete resection, there appears to be no appreciable growth of the small tumor remnant in the region of the cavernous sinus after 5 years.

Trigeminal tumors of the lateral skull base with intracranial components below the level of the posterior clinoid process are ideally suited for the ITF techniques. The importance of securing the petrous carotid artery through a wide exposure is emphasized. (*Skull Base Surgery*, 4(3):151–158, 1994)

Schwann cell tumors tend to localize in one of three regions along the nerve fiber.¹ They are (1) the neurolemmal-glial junction, (2) the sensory ganglion, and (3) distal nerve endings. Consequently, tumors of the trigeminal nerve arise most often intracranially from the ganglion region in Meckel's cave and from the proximal root, and involve the middle and posterior cranial fossae. The incidence of extracranial trigeminal nerve tumors is considerably lower, but are estimated to represent 15% to 20% of all extracranial neurogenic tumors.^{2–4} Their origins are often obscure and are presumed to arise from the peripheral nerve endings of the trigeminal divisions by virtue of their locations.

Infratemporal and pterygopalatine fossae as primary sites of involvement are particularly rare.^{5–12} Tumors that develop in these regions have a predilection to extend into the orbital apex and parapharyngeal space, often extensively.

In this study eight trigeminal nerve tumors of the lateral skull base are reviewed. They involve primarily the infratemporal fossa. The specific problems encountered in the management of these lesions are discussed.

MATERIAL AND METHODS

Between June 1979 and October 1991, 8 patients with tumors of the trigeminal nerve involving the lateral skull base were treated at our institution. The patient population consisted of 5 females and 3 males, ranging in age from 13 to 53 years (mean = 42 years).

Six patients presented for primary surgery, while 2 had prior surgical interventions. Our series included 3 schwannomas, 2 solitary neurofibromas, 2 neurofibromas associated with Von Recklinghausen's disease, and 1 case of malignant schwannoma.

Radiographic assessment included computed tomography, and magnetic resonance imaging was added for those treated after 1986. Tumors that demonstrated significant uptake of contrast or showed extensive infiltration were subjected to angiographic studies. This information helped to determine the status of the carotid artery and the need for embolization or carotid occlusion (see Table 1).

Frozen section analysis at the time of surgery was always performed. Immunohistochemistry was a vital

Table 1. Clinical Presentations and Surgical Results

#	Histology	Epicenter of Tumor	Tumor Extension	Approach	Adjuvant Rx	New Deficits
1.	Neurofibroma	ITF	Meckel's cave Cavernous sinus	C	—	CHL
2.	Schwannoma	ITF	Sphenoid sinus Meckel's cave Cavernous sinus Orbital apex Epipharynx	C	Embolization Carotid PBO	CHL VII paresis (transient)
3.	Neurofibroma	PPF	Orbital apex	D	—	—
4.	Neurofibromatosis type I	ITF	Parapharyngeal space	C	Embolization	CHL VII paresis (transient)
5.	Malignant schwannoma	PPF	Anterior ITF Parapharyngeal space Posterior maxillary wall Meckel's cave	D	Post-op radiotherapy	—
6.	Neurofibromatosis type I	ITF	Parapharyngeal space Parotid gland Meckel's cave	B	VII nerve resection and nerve graft	CHL Facial paralysis
7.	Schwannoma	ITF	Orbital apex Meckel's cave	C	—	CHL
8.	Schwannoma	ITF	Meckel's cave Parapharyngeal space	B	—	CHL

ITF = infratemporal fossa, CHL = conductive hearing loss, PBO = permanent balloon occlusion, PPF = pterygopalatine fossa.

part of the permanent histological assessment. Schwannomas and neurofibromas were differentiated by their morphologic features, supported by immunostains—specifically, S-100 and neurofilament stains. We have found that preoperative histological confirmation is often extremely difficult to establish, particularly if spindle cell or nonepithelial small cell tumors are encountered. Unless an alternative treatment plan is contemplated due to surgical contraindication, a preoperative biopsy of infratemporal fossa (ITF) lesions is usually not performed.

Surgical access was obtained with a type B or type C ITF approach as described by Fisch,¹³ depending on the origin and the extension of the tumor (Table 1). The extent of surgical exposure afforded by these techniques is schematically illustrated in Figure 1 and detailed in previous publications.^{14,15} In two cases (cases 3 and 5), a preauricular modification of the type C approach (designated as type D)^{16,17} was used to obtain access to the pterygopalatine region and the anterior ITF. The location of these lesions permitted such a preauricular technique, whereby the middle ear and the eustachian tube functions were preserved.

RESULTS

The length of time between the initial symptoms and diagnosis in our series ranged between 3 months and 8 years (average = 27 months). Trigeminal nerve dysfunctions were common but inconsistent (Table 2), ranging from dysesthesia, mild facial pain, to frontotemporal headaches. Conductive hearing loss (7/8) was the most frequent presentation as the result of eustachian tube ob-

struction, leading to middle ear effusion, tympanic atelectasis, or perforation. Another consistent finding was the mass effect produced by the tumor as manifested by facial asymmetry (3/8) and soft tissue asymmetry of the oropharynx and nasopharynx (6/8). Other presentations included 2 cases of proptosis, 1 case of diplopia (sixth nerve palsy) from cavernous sinus involvement with concomitant ipsilateral hemionopsia, and 1 Horner's syndrome possibly related to sympathetic neuropathy due to petrous carotid invasion.

Two patients (cases 1 and 2) were referred to our department following repeated attempts at subtemporal-transcranial resections. Patient 2 developed multiple cranial neuropathies (III-VI) and temporal lobe epilepsy subsequent to these attempts.

Radiographically, bony erosion of the skull base was present on computed tomography scans in all cases. Two of the tumors were exclusively extracranial (cases 3 and 4). Of the remaining 6, 4 had a small extradural extension to the Meckel's cave, and 2 showed cavernous sinus infiltration—one above and the other below the level of the posterior clinoid process. The extent of tumor involvement as depicted with computed tomography and magnetic resonance imaging is shown in Table 1.

Preoperative angiographic embolization was carried out in two subjects (cases 2 and 4) due to hypervascularity. Case 2 demonstrated tumor encasement of the petrous carotid artery and was subjected to a permanent internal carotid balloon occlusion preoperatively, to permit a complete resection. This was done without central nervous system complication.

Schwannomas in this series were well encapsulated and dissected from adjacent soft tissues without much

Figure 1. Basal skull view depicting the surgical exposures afforded by infratemporal fossa approaches types B, C, and D. Type D approach is achieved with a preauricular incision, preserving the middle ear function.

difficulty. Neurofibromas, on the other hand, were quite hard and fibrous, and often had significant dural adhesions. In patients with evidence of eustachian tube dysfunction the tube was discovered to be either invaded or tightly adherent to the tumor. Total extirpation was possible only when it was included in the resection. A type D

infratemporal approach was used for lesions in the pterygo-palatine fossa and anterior ITF (cases 3 and 5). This is a modification of the type C approach previously described by Close et al.¹⁸ Its use is limited to tumors that are situated anteriorly in the lateral skull base without involving the petrous carotid artery. By avoiding a subtotal petrosectomy the middle ear system and the eustachian tube are spared.

The uppermost limit of the ITF approaches appeared to be the posterior clinoid process. Total tumor extirpation was achieved in lesions with an intracranial component below this level (7/8 cases). Tumor extension above this level (case 2) was incompletely excised, which incidentally also failed two previous transcranial surgeries.

Operative time of this series ranged from 4 to 6½ hours (average 5½ hours). The average blood loss was 1800 cc, and the length of hospital stay was 14.3 days in average (ranging from 10 to 25 days).

Postoperative complications apart from the expected trigeminal deficits included 2 transient facial pareses and 1 facial paralysis. The transient facial paresis was the result of excessive intraoperative traction, and full recovery was realized in both within 1 month. One patient (case 6) had extensive neurofibromatosis also involving the facial nerve and the parotid gland. A total parotidectomy with facial nerve resection was carried out. A sural nerve interposition graft resulted in grade III (House/Brackmann) recovery at 3 years.

Conductive hearing loss as a result of obliteration of the middle ear and mastoid cavity is the expected sequela associated with type B and C approaches. Hearing was preserved in patients treated with the Type D approach. There were no central nervous system complications such as cerebrospinal fluid leak, meningitis, subdural or epidural hematomas, temporal epilepsy, and aphasia.

The average follow-up was 3 years in this study (ranging from 1 month to 6 years), with no radiological evidence of recurrence. In the case of malignant schwannoma, the patient remains disease-free following postoperative irradiation.

CASE PRESENTATION

Three representative cases of trigeminal nerve tumors are hereby detailed to better illustrate their management.

Patient M.G. (Case 2)

This 42-year-old woman was referred in 1985 with a trigeminal schwannoma, first diagnosed in 1974 following a 3-year history of right-sided headache, proptosis, and facial paresthesia. She underwent two unsuccessful neurosurgical interventions in 1976 and 1980. She suffered multiple cranial nerve (III/IV/V) palsies and temporal lobe epilepsy requiring anticonvulsive therapy. At referral the

Table 2. Initial Presentation (N = 8)

Trigeminal dysesthesia	4
Facial pain	3
Frontotemporal headaches	3
Facial mass or asymmetry	3
Pharyngeal bulge	6
Conductive hearing loss	7
Sixth nerve palsy	1
Horner's syndrome	1
Proptosis	2
Hemianopsia	1

patient demonstrated the above cranial nerve deficits. She also had a right tympanic perforation and bulging of the right nasopharynx. Computed tomography revealed a large ITF tumor with nasopharyngeal extension. An intracranial component involving the parasellar region above the posterior clinoid process was also noted (Fig. 2A,B). Angiography demonstrated significant tumor vascularity and encasement of the petrous carotid artery. An embolization and permanent balloon occlusion were carried out in preparation for surgery. A type C ITF approach was performed. Tumor extirpation involved the resection of the carotid artery.

Despite meticulous tumor removal, a small remnant above the posterior clinoid process could not be accessed. Postoperatively, the patient developed a transient facial palsy, which recovered fully within 1 month. There was no

appreciable growth of the residual tumor after 5 years of radiographic follow-up (Fig. 2C).

Patient F.C. (Case 4)

This 38-year-old physician presented in February 1988 complaining of a 1-year history of intermittent right-sided temporal headaches. He had also been treated for a right-sided serous otitis media with a series of ventilation tubes for the previous 8 years. On examination there was a visible swelling of the right canine fossa, with medialization of the oropharyngeal wall. A right atelectatic tympanic membrane was present with a 50- to 60-dB hearing loss.

Computed tomography and magnetic resonance imag-

Figure 2. A: Coronal computed tomography scan with contrast. Large infratemporal fossa tumor (*) with an extensive middle fossa component (arrows). Significant bony erosion of the lateral skull base is evident. B: Axial computed tomography view demonstrating the medial and lateral extensions. Note the cavernous sinus involvement (arrow). (Figure continued on the next page.)

Figure 2. (Continued). C: Post-operative axial computed tomography image displaying the residual tumor (arrow), showing no appreciable growth after 5 years.

ing (Fig. 3) revealed an ITF and parapharyngeal mass. There was evidence of bony erosion of the skull base with expansion of the foramen ovale. Angiographic examination demonstrated hypervascularity, and the tumor was embolized prior to surgery.

A type C ITF approach was carried out. The lesion was completely excised with the eustachian tube, which was encased by tumor. The histological diagnosis was a neurofibroma.

Incidentally, a separate tumor was discovered to involve the tympanic segment of the facial nerve. This

lesion was left untreated because the patient demonstrated no evidence of facial paresis preoperatively. This was thought to be a facial neuroma, which remained clinically asymptomatic after 3 years of follow-up.

Patient R.B. (Case 5)

A 13-year-old girl presented in June 1988 with a 6-month history of malocclusion associated with swelling of the right upper jaw and canine fossa. A tumor in the

Figure 3. A: T1-weighted magnetic resonance image. Sagittal view without gadolinium enhancement showing a well-circumscribed lesion (*) in the infratemporal fossa. B: Coronal magnetic resonance image displaying a dumbbell-shaped lesion (*) with expansion of the foramen ovale, and extension into the parapharyngeal space. No evidence of middle fossa involvement.

Figure 4. A: Coronal computed tomography scan without contrast enhancement. A large right-sided infratemporal fossa and parapharyngeal mass (*) with a small intracranial-extradural component (arrows). B: Axial computed tomography scan demonstrating tumor extension anteriorly with bony erosion of the maxilla and the pterygoid plates (arrows).

parapharyngeal space and the anterior ITF was seen on computed tomography scan (Fig. 4). It had eroded the lateral skull base, the posterior maxillary wall, and the hard palate. Superior extension into Meckel's cave was also present. A biopsy of this lesion via a Caldwell-Luc approach at another institution was suggestive of a neurofibroma.

Using a coronal, preauricular incision, a type D ITF approach was carried out. The tumor was removed completely, with the resection of parts of the posterior maxilla, alveolus, and hard palate. There was no involvement of the

eustachian tube, carotid artery, or the cavernous sinus. Routine histological studies with immunohistochemistry revealed a malignant schwannoma. A full course of radiotherapy was instituted postoperatively, and the patient has been without recurrence for 3 years.

DISCUSSION

Our understanding of the clinical behaviour of trigeminal nerve tumors is derived mainly from neurosurgi-

cal sources, involving lesions of the middle and posterior cranial fossae. Jefferson¹⁹ classified trigeminal neuromas into three types: (1) predominantly from the middle cranial fossa, (2) predominantly from the posterior cranial fossa, and (3) from both fossae. This classification failed to account for tumors of the lateral skull base, which clearly have a different mode of presentation. Two other categories must therefore be added—namely, (4) predominantly in the lateral skull base with a small middle cranial fossa involvement and (5) exclusively extracranial.

As expected, the most consistent presentations were associated with the mass effect of the tumors. Eustachian tube obstruction resulting in serous otitis, tympanic atelectasis, and perforation generally leads to a conductive deafness. This type of hearing loss has been mistakenly attributed to tensor tympani denervation in the past.²⁰ Facial asymmetry, pharyngeal swelling, and proptosis became evident with progressive tumor enlargement. Interestingly, in spite of trigeminal dysfunctions facial pain was not a consistent or significant feature in our patients. This is in contrast to the frequent manifestation of typical or atypical trigeminal neuralgia,^{21–24} characteristic of tumors arising from the proximal nerve root or Meckel's cave. Also of interest, severe facial pain was associated with a malignant schwannoma (case 5).

A surgical approach must be tailored to the site and the extent of the lesion. A class 1, 2, or 3 lesion (Jefferson's classification) requires primarily a neurosurgical access, while lateral skull base lesions with or without middle fossa involvement (class 4 and 5) are best managed by some form of lateral infratemporal technique.

Traditionally, head and neck surgeons have ap-

proached tumors in this region via a Weber-Ferguson incision, some incorporating variations of Crockett's technique,²⁵ with resection of the anterolateral and posterolateral maxillary walls. Arena and Hilal⁶ described a lateral preauricular approach, mobilizing the parotid gland and masseter muscle, in conjunction with a mandibular ramus resection, putting facial nerves in considerable traction. Nager⁸ reported a combined external-transpalatal-transmaxillary procedure to access the same area. Most recently, Arriaga et al²⁶ reported a facial translocation technique as another variation. These techniques generally suffer from limited exposure of the skull base, poor cosmetic result, or both. When a neurosurgical subtemporal approach is used, the lateral skull base component is often difficult to resect due to its limited inferior access. In spite of the benign nature of these tumors in the majority of cases, the high rate of recidivism calls for the complete extirpation of tumor.

In our series the ITF approaches were used exclusively. They provide well-documented access to the lateral skull base while ensuring control of the petrous carotid artery, cavernous sinus, and cranial nerves. Most importantly, this improved surgical access can be accomplished with minimal functional and cosmetic deficits.

The issue of conductive hearing loss associated with ITF approaches as a result of subtotal petrosectomy should be addressed. It must be emphasized that securing the carotid artery in its vertical and horizontal petrous segments is the single most important step when encountering lesions encroaching upon the petrous carotid artery, cavernous sinus, and parasellar region (Fig. 5). Apart from safety, a subtotal petrosectomy dramatically im-

Figure 5. Schematic representation of a typical trigeminal neuroma (TT) involving the lateral skull base. Tumor below the posterior clinoid process (PCP) is accessible via the infratemporal fossa approaches. ICE = intracranial extension, G = gasserian ganglion, PP = pterygoid plate, ICA = internal carotid artery, SS = sigmoid sinus.

proves the exposure and illumination of the skull base. If this step is circumvented to favor a preauricular approach, as a matter of convenience, the compromised access will undoubtedly result in incomplete tumor removal.

Furthermore, a poorly ventilated middle ear, if not obliterated, will serve as a continued source of infection. In selected cases a preauricular technique is preferred when tumor is clearly separate from the carotid artery.

Recently, Sekhar et al²⁷ and Mickey et al²⁸ described modifications of the ITF approaches with a sizable fronto-temporal craniotomy. This may be indicated in the presence of massive tumor involvement of the middle cranial fossa or significant dural invasion. Most tumors of the lateral skull base can be safely resected with a limited basal craniectomy, thus obviating the need for significant temporal lobe retraction and its sequelae. However, according to our experience the posterior clinoid process serves as the superior limit of surgical exposure with the ITF techniques.

Carotid artery occlusion has become a well-established procedure to ensure radical tumor excision. Yet, even with this radical approach total extirpation may not be achieved. The risk of neurological impairment associated with this form of management remains a contentious issue.

With regard to wound closure, a rotational temporalis muscle flap is generally adequate to reconstruct surgical defects of the skull base following ITF resections; if necessary, a superiorly based sternocleidomastoid muscle flap can be used jointly. We have not found the need to use free vascularized tissues for reconstruction in this series.

CONCLUSION

1. Trigeminal nerve tumors of the lateral skull base are ideally approached with types B and C ITF techniques. The initial subtotal petrosectomy offers the best exposure and security that are vital in complete tumor extirpation with minimal risk.
2. The use of the type D ITF approach should be reserved for lesions in the anterior ITF and the pterygopalatine fossa, when the petrous carotid artery is clearly separate from the tumor.
3. The posterior clinoid process serves as the upper limit of exposure in ITF approaches.

REFERENCES

1. Marttuzza RL, Ojemann RG: Bilateral acoustic neuromas—clinical aspects, pathogenesis, and treatment. *Neurosurgery* 10:1–12, 1982
2. Conley JJ, Janecka IP: Neurilemmomas of the head and neck. *Trans Am Acad Ophthalmol Otolaryngol* 80:459–464, 1975
3. Toriumi DM, Atiyah RA, Murad T, et al: Extracranial neurogenic tumors of the head and neck. *Otolaryngol Clin North Am* 19: 609–617, 1986
4. Kragh LV, Soule EH, Masson JK: Benign and malignant neurilemmomas of the head and neck. *Surg Gynecol Obstet* 111:211–218, 1960
5. McGuirt WF, Browne JD: An anterolateral approach to the anterior skull base—report of a malignant schwannoma of the pterygomaxillary space. *Otolaryngol Head Neck Surg* 98:323–327, 1988
6. Arena S, Hilal EY: Neurilemmoma of the infratemporal space. *Arch Otolaryngol Head Neck Surg* 102:180–184, 1976
7. Robitaille Y, Seemayer TA, El Deiry A: peripheral nerve tumors involving paranasal sinuses: A case report and review of the literature. *Cancer* 35:1254–1258, 1975
8. Nager GT: Neurinomas of the trigeminal nerve. *Am J Otolaryngol* 5:301–333, 1984
9. Puterman M, Tovi F, Sidi J: Neurilemmoma of the pterygomaxillary fossa. *J Laryngol Otol* 95:421–424, 1981
10. Jones HS, Clywd H: Benign schwannoma involving the infratemporal fossa and orbit. *Laryngoscope* 93:200–201, 1983
11. Gooder P, Farrington T: Extracranial neurilemmomata of the head and neck. *J Laryngol Otol* 94:243–249, 1980
12. Iwai Y, Hakuba A, Noguchi K: A gigantic neurilemmoma originating in the pterygopalatine fossa—case report. *Surg Neurol* 30: 452–456, 1988
13. Fisch U, Pillsbury HC: Infratemporal fossa approach to lesions in the temporal bone and base of the skull. *Arch Otolaryngol* 105: 99–107, 1979
14. Fisch U, Mattox D: *Microsurgery of the skull base*. New York: Thieme Medical Publishers, 1988
15. Fisch U: The infratemporal fossa approach for nasopharyngeal tumors. *Laryngoscope* 93:36–44, 1991
16. Shotton JC, Kuhwoede R, Fisch U: Mesenchymal tumors of the skull base with particular reference to surgical management and outcome. *Skull Base Surg* 2:112–117, 1992
17. May JS, Wiestler OD, Fisch U: Leiomyosarcoma of the skull base—a diagnostic challenge. *Skull Base Surg* 2:103–111, 1992
18. Close LG, Mickey BE, Samson DS, Anderson RG, Schaefer SD: Resection of upper aerodigestive tract tumors involving the middle cranial fossa. *Laryngoscope* 95:908–914, 1985
19. Jefferson G: Trigeminal neuromas with some remarks on malignant invasion of the gasserian ganglion. *Proc Cong Neurol Surg* 1:11–54, 1955
20. Devois FJ: Contribution à l'étude de neurinomes trigéminaux (a propos de 9 observations). Paris: 1970. Thesis
21. McCormick PC, Bello JA, Post KD: Trigeminal schwannoma—surgical series of 14 cases with review of the literature. *J Neurosurg* 69:850–860, 1988
22. Schisano G, Olivecrona H: Neurinomas of the gasserian ganglion and trigeminal root. *J Neurosurg* 17:306–322, 1960
23. Bullitt E, Tew JM, Boyd J: Intracranial tumours in a patient with facial pain. *J Neurosurg* 64:865–871, 1986
24. Yonas H, Jannetta PJ: Neurinoma of the trigeminal root and atypical trigeminal neuralgia—their commonality. *Neurosurgery* 6: 273–277, 1980
25. Crockett DJ: Surgical approach to the back of the maxilla. *Br J Surg* 50:819–821, 1963
26. Arriaga MA, Janecka IP: Facial translocation approach to the cranial base—the anatomic base. *Skull Base Surg* 1:26–33, 1991
27. Sekhar LN, Schramm VL, Jones NF: Subtemporal preauricular infratemporal fossa approach to large lateral and posterior cranial base neoplasms. *J Neurosurg* 67:488–499, 1987
28. Mickey B, Close L, Chaefer S, et al: A combined frontotemporal and lateral infratemporal fossa approach to the skull base. *J Neurosurg* 68:678–683, 1988

Dr. Chen's research was supported by the Samuel McLaughlin Fellowship Foundation, Canada.