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Main Article

Surgical management of trigeminal neuromas: a report of eight cases

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Abstract

Objective: Trigeminal neuromas are rare tumours that may involve any part of the Vth nerve complex, including extracranial peripheral divisions of the nerve. A series of eight patients with intracranial trigeminal neuromas who underwent surgical management are presented.

Methods: The hospital records and radiological images were reviewed with regard to clinical presentation, surgical approach, operative findings, peri-operative morbidity and neurological outcome, and percentage of tumour recurrence.

Results: The principal presenting symptoms were those of involvement of the trigeminal nerve with sensory impairment in one or more of the three divisions. Tumour location was the prime determinant of surgical approach. Lateral skull base approaches were used as they are considered to be superior for identifying tumour origin, extension, and relationship to surrounding structures. Total excision of the tumour was carried out in three of the eight patients. In the remaining five patients some tumour was left purposely in order to minimize neurological deficit and optimize post-operative quality of life. There was no peri-operative mortality or major morbidity in our series. Five patients experienced symptomatic tumour recurrence and revision surgery was performed.

Conclusion: Management of trigeminal neuromas is complex and requires a multidisciplinary approach. Pre-operative surgical planning allows tumour removal with preservation of important neural structures in the majority of cases. For large tumours occupying both the middle and posterior cranial fossae, the retrosigmoid/retrolabyrinthine/middle cranial fossa approach provides good exposure and results in minimal brain retraction. A Fisch type C approach is necessary for the largest tumours. Long-term follow up with interval imaging is mandatory to exclude long-term tumour recurrence.

Key words: Schwannoma; Trigeminal Nerve; Otologic Surgical Procedures; Radiosurgery

Introduction

Trigeminal neuroma is a benign schwannoma of the Vth cranial nerve first described by Dixon in 1846.[1] Vestibular schwannoma is the most common tumour to present in the cerebellopontine angle (CPA) (81 per cent).[2] The remaining 19 per cent comprise a fascinating group of CPA tumours, of which meningioma (6.5 per cent), and CPA cholesteatoma (4.6 per cent) are the most common.[2] Trigeminal neuromas are very rare intracranial tumours, accounting for 0.07 to 0.33 per cent of all intracranial tumours and 0.8 per cent to 8 per cent of intracranial schwannomas,[3,4] and comprise 1 per cent of CPA tumours.[2]

Because of the different neural root origins and direction of growth of these tumours along the trigeminal nerve, several classification systems have been suggested that have implications for the clinical findings, surgical approach, and outcomes of surgery in patients with these tumours. Jefferson[5] classified these tumours into three types: Type A, tumours located in the middle fossa that arise from the gasserian ganglion; Type B, tumours located predominantly in the posterior fossa that arise from the root of the trigeminal nerve; and Type C, tumours with significant components in both middle and posterior fossae (dumbbell shaped tumours).

Trigeminal neuromas often present incidentally and are challenging to manage. They have characteristic clinical and anatomical features.[6] They can originate from any section of the Vth cranial nerve and the corresponding symptoms and signs are largely determined by the tumour location. The majority of patients present with numbness involving one or all branches of the trigeminal cranial nerve, whereas motor involvement is an unusual feature. Facial pain is not a consistent symptom in trigeminal

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neuromas and changes in the corneal reflex and decreased facial sensation are more common than pain.[5,7,9,10]

The diagnosis and management of trigeminal neuromas presents the skull base surgeon with a formidable challenge because of their rarity, and in particular their anatomical position. Presentation is often late and the surgeon may be faced with a large tumour in a young person. Early diagnosis depends on a high index of suspicion and good imaging.

We present the senior authors' (DAM and DGH) surgical experience of these difficult tumours. The modes of presentation, investigation, management strategies, surgical approaches, complications and outcome are analysed. A review of the literature and the controversies and dilemmas surrounding these tumours is discussed.

Patients and methods

Eight patients with trigeminal neuromas were treated at the Department of Neuro-otology and Skull Base Surgery, Addenbrookes Hospital, Cambridge, between 1991 and 2002. The hospital records and radiological images of these patients were reviewed. The imaging modalities employed consisted of computed tomography (CT) scanning including bone settings and magnetic resonance imaging (MRI) T1- and T2-weighted images with gadolinium DTPA enhancement. The site of the primary and recurrent tumour, the size, the surgical approach used, operative findings, rate of tumour recurrence, peri-operative morbidity and neurological outcome were all recorded.

In this series, there were five female and three male patients (See Table I). The mean age of the patients was 28 years (range 22–41). Trigeminal neuromas tend to become manifest at a relatively young age. The duration of symptoms before referral ranged from two months to two years with a mean of 10 months.

Results

Presenting clinical features

The principal presenting symptoms resulted from involvement of the trigeminal nerve, either as sensory impairment in one or more of the three divisions, or motor deficits of the muscles of mastication. All eight patients had facial numbness; four had facial pain and two patients had imbalance. Four patients complained of double vision and one patient, who had previous surgery elsewhere, already complained of facial weakness. On examination, six patients had involvement of V1, seven had V2 nerve involvement and five had V3 involvement. One patient had involvement of the motor branch of the trigeminal nerve. One patient who had previous surgery elsewhere had a facial nerve paralysis House-Brackman (HB) grade 3, one a III nerve paralysis, one a IV and two had a VI nerve involvement. Abnormal gait and cerebellar signs were present in four patients and one patient had weakness of the masseter muscle. Five patients had normal hearing at the time of presentation. Hearing loss was seen in three patients – one had a mild conductive hearing loss and two had a profound sensorineural hearing loss.

TABLE I

DEMOGRAPHICS OF PATIENTS WITH TRIGEMINAL NEUROMA

Demographics	No. of patients
Age	
20–30	6
31–40	1
41–50	1
Sex	
Male	3
Female	5

Tumour location

Pre-operative MRI scans were obtained routinely in all patients to identify the lesion, its extent, morphology and anatomical site (Figures 1 and 2). Table II summarizes the sites of the tumours and classifies them according to Jefferson.

Surgical approach

The surgical approach in each case was determined by the anatomical location and extent of the tumour. A schematic presentation of the skin incision and lateral skull base approaches used are illustrated in Figures 3 and 4 respectively. In most instances lateral skull base approaches were adopted (see Table III). The combined retrosigmoid/retrolabyrinthine/middle fossa approach was used in five patients. One patient had

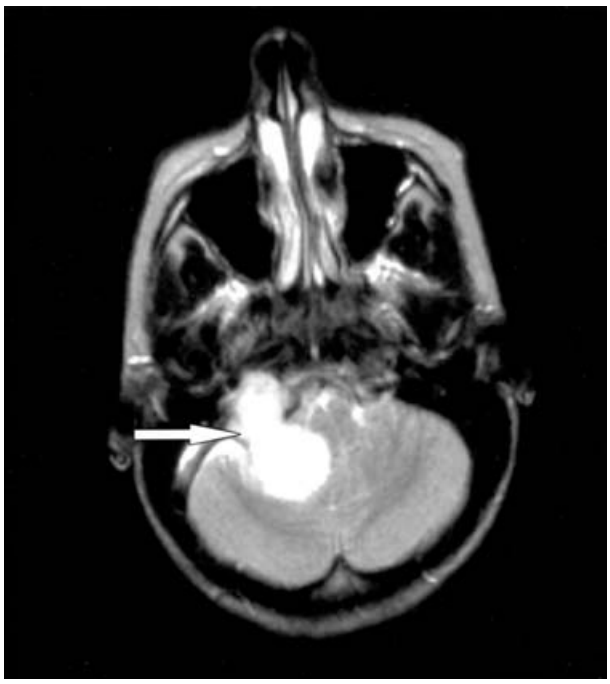

FIG. 1

Axial T2-weighted MRI scan with gadolinium DTPA contrast demonstrating a typical enhancing dumbbell tumour occupying both middle and posterior fossae. There is significant brainstem compression.

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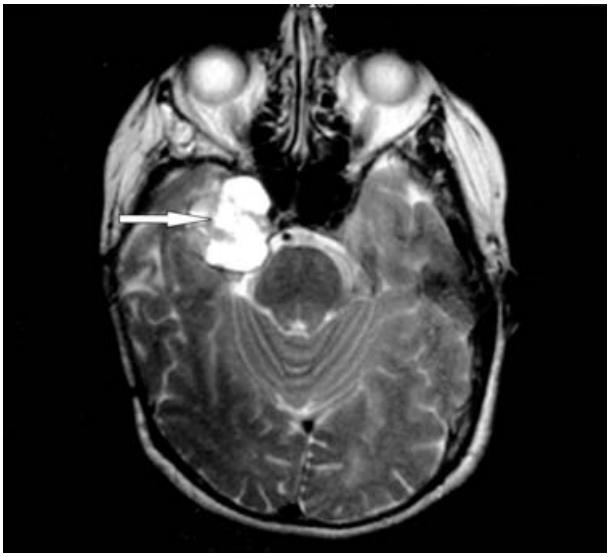

FIG. 2
Axial T2-weighted MRI scan with gadolinium DTPA contrast showing a trigeminal neuroma located in the middle cranial fossa.

retrosigmoid/transtemporal/middle fossa approach and in another patient middle fossa approach was considered appropriate. One tumour was excised via Fisch type C approach.[11]

These lateral skull base approaches allow good exposure of the tumour with minimal brain retraction. Total excision was possible in three of the eight patients and in five a balanced clinical judgement was used by the surgical team to determine the extent of the resection. In view of the indolent nature of these tumours, preservation of cranial nerve function was considered more important than total extirpation of every morsel of the tumour, particularly when the tumour capsule was inextricably involving cranial nerves or the cavernous sinus.

In those patients with recurrence of their tumour, the approaches were dictated by the site and extent of the tumour. One patient had retrosigmoid/transtemporal/middle fossa approach, whereas another patient had retrosigmoid/middle fossa approach. The Fisch type B[11] approach was considered appropriate in one case and a pterional approach was necessary in another case in order to provide better access to the tumour. The different approaches adopted for removal of these recurrences is also shown in Table III.

Surgical outcome

The surgical outcome and a comparison of the neurological status pre- and post-operatively can be seen in

TABLE II
TUMOUR LOCATION

Tumour location (Jefferson classification)	No. of patients
Type A – Middle fossa alone	4
Type B – Posterior fossa alone	1
Type C – Middle and posterior fossae	3

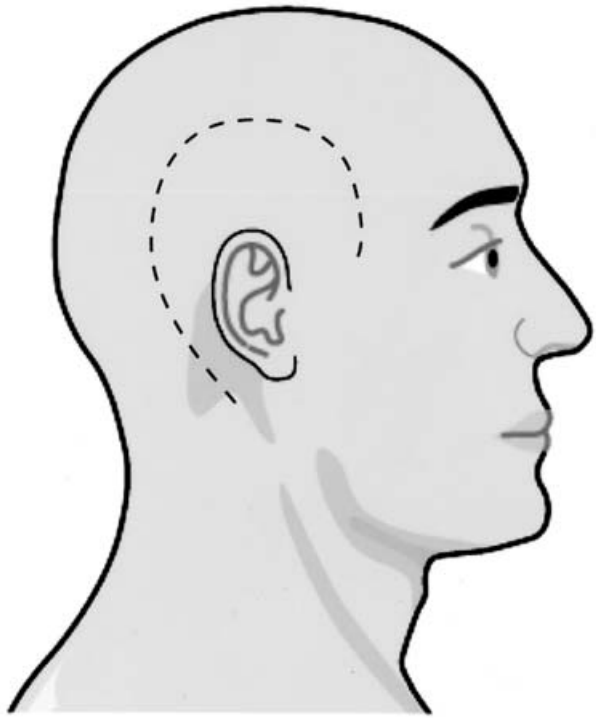

FIG. 3
The skin incision used in transtemporal and middle fossa approaches to provide surgical access to the middle and posterior cranial fossae.

Table IV. Facial numbness improved in two patients post-operatively, whereas symptoms like headache and facial pain settled completely. Diplopia remained a symptom in four patients. Of the five patients with

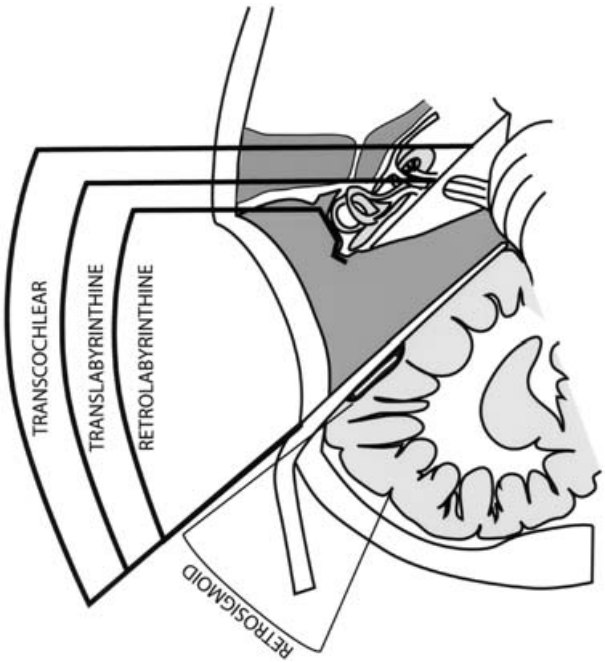

FIG. 4
Schematic presentation of the lateral skull base approaches used for access to the middle and posterior cranial fossae. The retrolabyrinthine approach is a presigmoid posterior fossa craniotomy preserving the inner-ear structures.

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TABLE III

SURGICAL APPROACHES IN PRIMARY AND REVISION SURGERY

Surgical approach	Number of patients	
	Primary procedure	Revision procedure
RS/RL/MF	5	0
RS/TT/MF	1	1
RS/MF	0	1
MF	1	0
Fisch B	0	1
Fisch C	1	0
Pterional	0	1

RS = Retrosigmoid, RL = Retrolabyrinthine, MF = Middle Fossa, TT = Transtemporal (Translabyrinthine / Transcochlear)

pre-operative normal hearing, one developed mild sensorineural hearing loss, whereas in one patient with conductive deafness, the hearing returned to normal. Gait disturbance was present in three out of the four patients who presented with this sign pre-operatively. Improvement in the paralysis of the trigeminal nerve branches after the removal of the tumour was noticed in five patients. Post-operatively, one patient had paralysis of the trochlear and abducent nerves. There was one patient with oculomotor nerve, two with abducent nerve and three with grade 3 facial nerve paralyses. Other complications included one patient with CSF meningitis which settled with antibiotics, one with post-operative seizures who needed long-term anti-epileptics and one developed mild hemiplegia which required rehabilitation. (see Table IV).

Recurrences

Five patients experienced symptomatic tumour recurrence (Figure 5). The interval between the primary surgery and revision surgery ranged from one to five years, with a mean of 2.9 years. In one patient a third operation was required for a further recurrence at an interval of 13 years from the first operation. Following revision surgery there was no mortality and the number of post-operative cranial neuropathies was largely unaltered except for one patient who had V3 deficit. The pure tone averages and speech discrimination scores remained at the pre-operative levels apart from one patient who had a mild hearing loss. Other post-operative complications from surgery for recurrent tumours included one patient with meningitis and two patients with seizures post-operatively where the middle fossa (MF) route was used intra-operatively. One patient had deterioration in facial nerve function (from House–Brackmann grade 3 to grade 4). The outcomes of surgery for recurrent tumours are summarized in Table V.

TABLE IV

CLINICAL FEATURES AND CRANIAL NERVE INVOLVEMENT PRE AND POST SURGERY

Clinical features	No. of patients $n = 8$	
Presenting symptoms	Pre-operative	Post-operative
Facial numbness, paraesthesia	8	6
Facial pain	4	0
Headache	4	0
Diplopia	4	4
Seizures	0	1
Hearing – Normal	5	4
Conductive loss	1	0
Sensorineural loss	2	3
Gait disturbance	4	3
Cranial nerve involvement		
V1	6	5
V2	7	6
V3	5	3
Motor	1	0
VII	1	3
VIII	3	3
III	1	1
IV	1	1
VI	2	2
Cerebellar signs	4	3
CSF leak	0	1
Mild hemiplegia	0	1

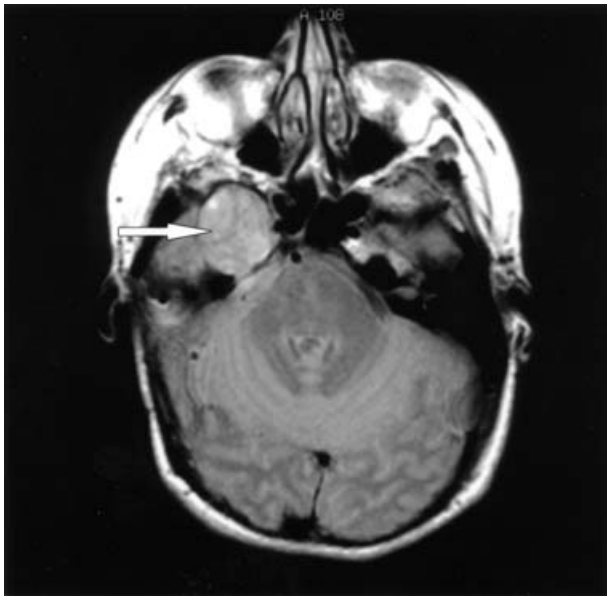

FIG. 5

Axial T1-weighted un-enhanced MRI scan showing a recurrent tumour in middle cranial fossa. There is no evidence of mass defect, midline shift or hydrocephalus.

Discussion

The presenting features in this series of patients were not significantly different from those observed in earlier reviews.[3,6,7] The majority of patients present with numbness involving one or all branches of the trigeminal cranial nerve, indicating involvement of the sensory branch, whereas motor involvement with weakness of the muscles of mastication is unusual and occurred in only one patient. The relationship between facial pain and tumours involving the trigeminal nerve has been a clinical curiosity. Several authors believed that facial pain is not a consistent feature in trigeminal neuromas and that changes in the corneal reflex and decreased facial sensation are more common than pain.[5,7,9,10] This observation is supported by this series where only

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TABLE V

PRESENTATION AND OUTCOME FOLLOWING REVISION SURGERY FOR RECURRENT TUMOUR

Clinical features	No. of patients $n = 4$	
Presenting symptoms	Pre-operative	Post-operative
Facial numbness, paraesthesia	4	4
Diplopia	2	2
Headache	1	0
Hearing loss deterioration	1	2
Gait disturbance	1	1
Cranial nerve involvement		
V1	4	3
V2	4	4
V3	3	4
Motor	–	–
VII	1	1
VIII	1	2
VI	1	1
IV	1	1
Lower cranial nerves (IX, X, XI)	1	1
Cerebellar signs	1	1
Meningitis	0	1
Seizures	0	2

(One patient with recurrence was not re-operated in our institution.)

50 per cent of patients had facial pain. Bullitt *et al.*[12] suggested that pain in the form of trigeminal neuralgia is most commonly caused by tumours involving the trigeminal root. Interestingly, the quality of pain in the majority of patients with trigeminal neuroma differs from that in patients suffering from idiopathic trigeminal neuralgia.[7,13] Most patients with pain from neuromas have paroxysmal lancinating facial pain; however the episodes tend to last longer and do not always have specific triggering mechanisms. This pain does not respond to the medication commonly used to treat trigeminal neuralgia.[14]

McCormick *et al.*[7] found that 75 per cent of patients had other cranial nerve abnormalities at the time of diagnosis. The associated cranial neuropathies were largely determined by the location of the tumour and its relationship to adjacent neural and vascular structures. Tumours involving the posterior cranial fossa predominantly affected the VIIth and VIIIth cranial nerves whereas larger tumours involving the posterior cranial fossa were also associated with cerebellar and long-tract signs.[7,15] Motor deficit of the Vth or VIIth nerve was found to be significantly less frequent than generally reported, 12 per cent in this study, compared with 30 per cent reported by several authors.[8,10] Patients who presented with diplopia suffered mostly from deficits of the abducens nerve, supposedly secondary to compression by the tumour.[13] It is important to stress that diminished corneal reflex together with facial paraesthesia or facial pain, are not diagnostic of trigeminal neuroma, a similar symptom complex being shared by meningioma of Meckel's cave and cholesteatoma of the CPA. VIIIth nerve involvement was found in almost half of the patients in this series, concurring with previous studies.[8] Hearing may also be affected by impaired tensor tympani function due to loss of its trigeminal innervation. Also, hearing loss and facial nerve dysfunction have been reported in patients whose tumours have significantly eroded the petrous bone with damage to the inner and/or middle ear.[8]

The diagnosis of trigeminal neuroma is a difficult one to make on clinical grounds alone. Imaging is particularly important to establish the precise size, morphology and anatomical location of the tumour, and its relationship to neighbouring structures. This will determine the optimal surgical approach adopted for any one particular tumour. The primary radiological investigation for the diagnosis of these lesions should be high resolution CT scanning using bony windows, to determine the extend of bony erosion and MRI for soft tissue delineation. MRI has an added advantage in the pre-operative imaging of these tumours because of its multiplanar capability. Trigeminal neuromas appear slightly hyperintense on the T1-weighted images with significant enhancement after gadolinium DTPA injection. In view of this, a high resolution CT scan with bony windows and MR imaging with gadolinium DTPA enhancement are complementary in providing complete pre-operative information. Rarely, angiography is useful in pre-operative assessment as these tumours are not highly vascular and embolization is not necessary. In very large tumours, however, cerebral angiography may be valuable in order to identify the degree of displacement of intracranial vessels prior to undertaking surgery.[3] In anteriorly situated tumours, cerebral angiography shows the displacement of the intracavernous and petrosal segment of the intrapetrous carotid artery. When a significant posterior fossa component is present, dorsocranial displacement of the posterior cerebral and superior cerebellar arteries with downward displacement of the anterior inferior cerebellar artery has been reported.[17,18,19]

Trigeminal neuromas tend to displace surrounding neurovascular structures rather than engulf them.[20] Surgery for these lesions may be very challenging, particularly in large tumours. The potential of these tumours to extend into two intracranial compartments will influence the choice of surgical approach. The principles of microsurgery include careful dissection of the tumour capsule from the surrounding structures following adequate debulking of the tumour. The aim of total tumour removal[21,22] which offers the best chance of cure, and dramatically reduces the tumour recurrence rates,[6] must be balanced by the need to preserve the neural structures and, consequently, quality of life. Sindou and Pelissou[23] found that total removal of the tumour was achieved in only 50 per cent of cases because of the close relationship of the lesion to the CPA, petrous apex, cavernous sinus and multiple cranial nerves. Despite the emphasis on total removal in later series,[7] this has not always been possible because of risks to vital structures.[6,7] In our opinion, it is important to be cognisant of post-operative quality of life and some compromise of

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the surgical ideal of total tumour removal may be of overriding importance to the patient since these indolent tumours are benign and slow growing. Our aim is preservation of cranial nerve function and to minimize post-operative complications. If the tumour is found to be firmly adherent to the cavernous sinus, it may be left in order to reduce subsequent morbidity. Although this may result in a higher recurrence rate, recurrences can be observed with high resolution imaging and treated conservatively or with further surgery if necessary. Lateral skull base approaches allow good exposure of these tumours,[24] with minimal brain retraction obviating the need for two stage tumour resection.

Significant advances in neuroimaging, electrophysiological monitoring, microsurgical techniques, and newer skull base approaches have made a significant contribution towards achieving the surgical goal of complete tumour removal with minimal morbidity. There were no deaths or major morbidity in this series, which corresponds with other series reported in the literature.[7,13,21] Post-operative facial numbness improved in five cases, and facial pain was alleviated in all six cases where it was present pre-operatively. Hearing deteriorated in only one patient who had no previous hearing problems.

Reported recurrence rates are high. Taha *et al.*[4] reported a 60 per cent recurrence rate in his series and in this series it was similar. Injury or permanent damage to the trigeminal nerve branches has been inevitable in many cases.[7,9,22] Good quality of life with only trigeminal nerve symptoms is considered to be an excellent outcome.

Stereotactic radiotherapy in the form of either single dose radiosurgery (gamma knife) or fractionated from a linear accelerator source (LINAC) is reported as an effective treatment for small and medium size trigeminal schwannomas.[25] Some larger tumours, up to 3 cms in maximum diameter, may also be suitable for stereotactic radiotherapy if there is no significant brainstem compression. The upper limit of size for radiotherapy treatment is governed by the fact that tumours will swell in the immediate post treatment phase and this may increase brainstem compression and precipitate decompensation. Stereotactic radiotherapy has also been proposed as an alternative and minimally invasive primary treatment option for patients with newly diagnosed or residual trigeminal schwannoma, especially if the tumour size is small.[26,27] Tumour shrinkage was achieved in 565 of treated patients and tumour growth arrested in 44 per cent of a series reported by Huang *et al.* (1999).[27]

In this series, many cases were treated prior to the prevalent use of radiosurgery in this country and many of these tumours were very large and presented with varying degrees of brainstem compression and would not have been suitable for this form of treatment. For small tumours, expectant management with interval scanning was preferred, since the evidence base for treating these very rare tumours with radiosurgery had not yet been established. Recurrent tumours were treated by further surgical resection in concurrence with current opinion at that time.[21,22,24]

Stereotactic radiotherapy will certainly be considered as an alternative treatment option for any patient with a small tumour which has demonstrated significant growth on interval imaging and also for known residual or emergent recurrent tumour as this may obviate the need for revision surgery.

Conclusion

Trigeminal neuromas are rare benign slow-growing tumours most commonly occurring in young adults and their management is complex and requires a multidisciplinary approach. Total excision reduces the risk of recurrence but may be associated with significant morbidity and deterioration in quality of life. Pre-operative surgical planning based on high resolution imaging allows tumour removal with preservation of important neural structures in the majority of cases. Large tumours occupying both the middle and posterior cranial fossae, are best excised by a lateral skull base approach where good exposure is obtained at the expense of bone removal rather than brain retraction. Long-term follow up with interval imaging is mandatory to exclude tumour recurrence, which if it occurs may require stereotactic radiotherapy or further surgery.

- **Trigeminal neuromas are rare benign slow-growing tumours most frequently seen in young adults. Their management is complex, and requires a multidisciplinary approach**
- **Total excision reduces the risk of recurrence but may be associated with significant morbidity**
- **A lateral skull base approach is needed for large tumours with both middle and posterior cranial fossa involvement**
- **Stereotactic radiotherapy may offer an alternative to surgery in some cases**

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