Benign Extracranial Nerve Sheath Tumors of the Skull Base: Postoperative Morbidity and Management

Deb Biswas, M.S. (ORL), M.R.C.S., D.O.H.N.S.,¹ Conor Marnane, F.R.C.S. (ORL),¹ Ranjit Mal, F.R.C.S.,¹ and David Baldwin, F.R.C.S.¹

ABSTRACT

The purposes of this retrospective case series study were to examine the outcome of the operative treatment of extracranial nerve sheath tumors (NSTs) of the skull base and to learn the optimal management. The study was conducted at a university teaching hospital and a regional referral center. A total of 19 cases of benign extracranial NSTs of the skull base who presented to the otolaryngology department over a period of 10 years were studied regarding the clinical, radiological, and pathological features and the operative and postoperative management. In the majority, these tumors originated from cranial nerves; postoperative complications were frequent and depended on the nerve of origin. Postoperative nerve deficit was apparent in 10 cases, and a second operation was necessary in 8 cases. The greatest postoperative morbidity was associated with the parapharyngeal NSTs (i.e., dysphagia in 30%, dysphonia in 30%, and Horner's syndrome in 20% of cases). The conclusion from this study is that high postoperative morbidity resulting from surgery on skull base NSTs demands an integrated approach between the otolaryngologist, plastic surgeon, neurosurgeon, speech therapist, physiotherapist, dietician, and occupational therapist and a clear strategy of long-term follow-up.

KEYWORDS: Benign, neurofibroma, schwannoma, extracranial, skull base

¹Department of Otolaryngology–Head and Neck Surgery, Southmead Hospital, North Bristol NHS Trust, Westbury on Trym, Bristol, United Kingdom.

Address for correspondence and reprint requests: Deb Biswas, M.S. (ORL), M.R.C.S., D.O.H.N.S., Department of Otolaryngology–Head and Neck Surgery, Southmead Hospital, North Bristol NHS Trust, Westbury on Trym, Bristol BS10 5NB,

United Kingdom (e-mail: drdbiswas@hotmail.com).

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Nerve sheath tumors (NSTs) are rare, although they are relatively common in the head and neck region. They may grow from any myelinated nerve, but in the skull base region they have an affinity for cranial nerves. Because of the high postoperative complication rate of NSTs, it is important to make a correct preoperative diagnosis regarding the histology and nerve of origin to enable advance planning in, for example, nerve grafting. A multidisciplinary team discussion between surgeons, radiologists, and histopathologists is essential to plan surgery and for an informed consent.

MATERIAL AND METHODS

The clinical, radiological, and pathological records of all patients with extracranial NSTs of the skull base treated in our institution between January 1993 and January 2004 were retrospectively reviewed. These patients were managed under joint care of the head, neck and skull base surgeons; neurosurgeons; and plastic surgeons. The minimum follow-up period was 2 years. Intracranial NSTs are not included in this study. Particular attention was paid to the postoperative complications and management.

RESULTS

Clinical Features

Nineteen cases of NSTs were treated in our institution between 1993 and 2004. There were 14 cases of schwannoma and 5 cases of neurofibroma, the ratio being 3:1. Three out of 5 patients with skull base neurofibroma were previously known to have type 1 neurofibromatosis. The male-to-female ratio was 7:12. The mean age was 43 years (range, 28 to 74 years).

We classified these tumors into five anatomical subgroups (Table 1). The highest number of

cases was found in the parapharyngeal space (53%), followed by the mastoid and middle ear (21%), and the frontonasal ethmoidal (16%). The presentation of an NST was dependent on its location in the skull base and the nerve of origin.

A soft tissue lump was the most common presentation (57.8%). In most of the cases, the initial symptoms and signs were related to the pressure caused by the tumor mass. Among the middle ear NSTs, facial nerve palsy was the most common presentation followed by conductive deafness and postaural mass. A preoperative tissue diagnosis was possible in two cases from biopsy of aural polyps. Nasal polyp and nasal obstruction were the presentation in anterior skull base NSTs. A skull vault plexiform neurofibroma presented with a very large soft tissue mass in the temporo-cervical region, associated with osteomatous changes of the underlying skull bone.

Parapharyngeal space NSTs involving the skull base had the most diverse presentation, with neck lumps being the most common (60%). Three out of six patients with neck lumps had the following associated symptoms: dysphagia in three cases, palatal palsy in two cases, and dysphonia in one case. Of the patients presenting without neck lumps, one had otalgia and the other had facial weakness. Headache, vertigo, and blurring of vision were the presenting symptoms of the schwannoma of the second division of the trigeminal nerve (infratemporal/pterygopalatine fossa).

Nerve of Origin

Clinical features indicating the nerve of origin are shown in Table 1. Although identification of the nerve of origin is important for informed patient consent, it was possible in only 15.8% of cases (three cases) before operation. The final nerve identification was made in 57.9% (11 cases). The nerve involved can be ascertained during the operation or from presenting features like facial palsy and from postoperative complications like Horner's syndrome.

Table 1 Description of Cases of Extracranial Skull Base Nerve Sheath Tumors (n = 19)

Presentation	Operation	Diagnosis	Morbidity
Middle Ear and Mastoid (n = 4)			
1 Conductive deafness	1st: tympanotomy and excision of tumor from facial nerve and end-to-end anastomosis 2nd: residual—translabyrinthine with sural nerve graft	Facial nerve schwannoma	Residual facial weakness (grade III) and tinnitus
2 Facial palsy, aural polyp, postaural fullness	1st: transmastoid approach 2nd: residual—petrosectomy with sural nerve graft	Facial nerve schwannoma	Persistent facial palsy
3 Facial palsy, aural polyp	Modified radical mastoidectomy and sural nerve grafting	Facial nerve schwannoma	Persistent facial palsy
4 Postaural lump	Transmastoid approach	Neurofibroma (nerve unknown)	None
Skull Vault (n = 1)			
5 NF1, temporo-cervical soft tissue tumor (osteomatous changes of underlying skull on CT scanning)	1st: excision of soft tissue and reduction of osteoma 2nd: meatoplasty	Plexiform neurofibroma	Meningoencephalocele; No symptoms, no neurological deficit
Frontonasal ethmoidal (n = 3)			
6 Unilateral nasal obstruction, epiphora	1st: endoscopic removal 2nd: residual—bifrontal approach	Ethmoidal schwannoma	None
7 Nasal polyp	Transfrontal transbasal coronal flap	Frontonasal schwannoma	None
8 Nasal obstruction	Endoscopic removal	Neurofibroma	None
Infratemporal/Pterygopalatine Fossa (n = 1)			
9 Headache, vertigo, blurred vision (MRI—SOL in pterygopalatine fossa to middle cranial fossa)	1st: transantral approach 2nd: residual—bicoronal right temporozygomatic arch approach	Schwannoma of 2nd division of trigeminal nerve	None
Parapharyngeal (n = 10)			
10 Neck lump	Excision upper transcervical approach and phrenic nerve graft to posterior cricoarytenoid	Vagal schwannoma	Initial hoarseness of voice but acceptable voice after 2 y of voice therapy
11 Otalgia (MRI—parapharyngeal tumor pushing tonsil and soft palate forward)	Mandibulotomy	Neurofibroma (nerve unknown)	Scar keloid
12 Neck lump	Transcervical excision	Schwannoma (nerve unknown)	None
13 Neck lump	Cervical approach—transverse incision	Sympathetic schwannoma	Horner's syndrome
14 NF1, dysphagia, reduced palatal mobility, neck lump	1st: transcervical excision 2nd: PEG insertion 3rd: VC medialization	Vagal Schwannoma	VC palsy, dysphagia, full voice recovery 2 y after VC medialization

Table 1 (Continued)

	Presentation	Operation	Diagnosis	Morbidity
15	Neck lump	Upper cervical approach	Hypoglossal schwannoma	Horner's syndrome; facial weakness recovery
16	Hoarseness of voice, dysphagia, palatal palsy; MRI—parapharyngeal mass deep to styloid process	1st: parotid-like incision with cervical extension 2nd: PEG insertion	Vagal schwannoma	Facial palsy and myotonia, dysphagia, weakness of voice
17	NF1, neck lump; MRI—right upper pararetropharyngeal mass	1st: right pharyngotomy approach 2nd: PEG insertion	Glossopharyngeal neurofibroma	Dysphagia
18	Neck lump, dysphagia, swelling occupying two thirds of oropharynx	Cervical approach	Glossopharyngeal schwannoma	Mild hypoglossal palsy
19	Multiple neck swelling; FNAC of neck mass—squamous cell carcinoma; MRI of neck—prevertebral lesion with parapharyngeal extension	Biopsy—CT-guided prevertebral lesion; not operated for NST	Schwannoma (nerve unknown)	Not operated on

NF1, neurofibroma type 1; CT, computed tomography; MRI, magnetic resonance imaging; SOL, space occupying lesion; PEG, percutaneous endoscopic gastrostomy; VC, vocal cord; FNAC, fine-needle aspiration cytology; NST, nerve sheath tumor.

Figure 1 T2-weighted magnetic resonance imaging (MRI) scan (postcontrast) at the level of the temporal bone showing a nerve sheath tumor (NST) (schwannoma) arising from the facial nerve in the left middle ear (arrow).

Radiological Investigation

All patients underwent MRI scanning with contrast before and after the operation. Only in one instance were the features strongly suggestive of a schwannoma; otherwise NST was one of the possibilities in the differential diagnosis. We found that a STIR (short tau inversion recovery) sequence in MRI scanning was extremely helpful for the localization of the tumor. Preoperative computed tomography (CT) scanning was also done as a routine in seven cases, including four NSTs of the middle ear and mastoid. MRIs of NSTs of the middle ear, parapharyngeal space, and anterior skull base are shown in Figs. 1, 2 and 3, respectively.

Summary of Operative Management

Eighteen patients were operated on (94.7%), a second operation was necessary in eight cases (42.1%), and a third operation only in one case (case 14). The indications for second intervention was removal of residual tumor, functional correction

(e.g., vocal cord medialization, meatoplasty), and supportive procedure (e.g., percutaneous endoscopic gastrostomy [PEG]). The tumor excision was incomplete in three cases (15.8%), requiring a second operation. In one case of a facial nerve schwannoma (case 1), the residual tumor was defined a month after initial tympanotomy on repeat MRI scanning and was subsequently treated with translabyrinthine approach surgery. Radical excision of the petrous temporal bone was performed for the residual part of a facial schwannoma after an initial transmastoid excision (case 2). In the case of the nasoethmoidal tumor where a complete excision could not be achieved by endoscopic surgery, a second operation was undertaken through a bifrontal craniotomy (case 6). The second operation for the residual tumor was successful in all three cases. There was no recurrence or malignant transformation of benign NSTs, nor was there any mortality. The low incidence of recurrence and malignant transformation of the benign NST encourages us to consider functional surgery such as phrenic nerve rerooting to the cricoarytenoid muscles and vocal cord medialization.

Figure 2 T2-weighted magnetic resonance imaging (MRI) scan of skull base in short tau inversion recovery (STIR) sequence showing a nerve sheath tumor (NST) (neurofibroma) in the left jugular foramen extending down into the left parapharyngeal space (arrow).

Figure 3 T2-weighted magnetic resonance imaging (MRI) scan of anterior skull base showing a giant nerve sheath tumor (NST) (schwannoma) of unknown nerve origin with intracranial extension (arrow).

Nerve Damage and Morbidity

Nerve damage can be due to division of the nerve of origin at operation or due to an injury to a nerve close to the operating field such as facial nerve palsy occurring after surgery for vagal schwannoma. The resulting morbidity is dependent on the functional outcome of nerve grafting. The overall complication rate was 66.7% (12 cases). A postoperative nerve

deficit was apparent in 10 cases (55.5%). At the end of a 2-year follow-up period, functional improvement of neurological disability had occurred in 5 out of 10 cases. In the other five cases, there were three cases of persistent facial weakness in the cases of facial nerve schwannomas. There was one case of persistent Horner's syndrome that was asymptomatic and one case of dysphagia after excision of a vagal schwannoma that required a permanent PEG.

The greatest postoperative morbidity was associated with parapharyngeal NSTs (Table 2). Dysphagia was the most common complication, and all cases required PEG feeding for a variable period of time (4 and 10 months in two subjects and permanently in one). One of the two cases of Horner's syndrome recovered after 9 months but the other did not. Horner's syndrome followed resection of a sympathetic chain schwannoma excision and in another case it developed after an inadvertent injury during a hypoglossal nerve schwannoma excision. Only one out of three vagal schwannomas presented with a hoarse voice, but postoperatively all the three cases developed vocal cord palsy. In one case of the above three vagal schwannomas, a split phrenic nerve graft to the posterior cricoarytenoid muscle resulted in a reasonable voice after 2 years of voice therapy. Vocal cord medialization was highly successful in another case (5 months after the first^t operation). One patient had permanent glottic weakness but did not wish to have further surgery; however, this

Table 2 Postoperative Complications and Morbidity (n = 19)

Site	Complications	Follow-up
Mastoid—middle ear (4)	Residual facial weakness (3) Tinnitus (1)	Persistent in two and one improved to grade III after 1 y
Frontonasal ethmoidal (3)	None	Complete recovery
Skull vault (1)	Meningoencephalocele	No neurological deficit, no symptoms
Infratemporal fossa (1)	None	
Parapharyngeal (10)	Dysphagia (3) VC palsy (3) Horner's syndrome (2) Hypoglossal palsy (1) Facial palsy (1) Facial myotonia (1) Scar/keloid (1)	PEG feeding (two temporary and one permanent) Improvement of voice Improvement in one, persistent but asymptomatic in the other Compensated Recovery after 1 y Persisted for 9 mo Unchanged

PEG, percutaneous endoscopic gastrostomy; VC, vocal cord.

patient's voice improved after 2 years of voice therapy. The speech and language therapists were involved in all cases of parapharyngeal NST. Weakness of the tongue following excision of a glossopharyngeal schwannoma was well compensated after 10 months.

Facial nerve defects were grafted using sural nerve interposition grafts in all cases; the functional outcome was poor in two cases and one regained House-Brackmann grade III function. Conductive deafness in the case of plexiform neurofibroma of skull vault (case 5) was corrected by meatoplasty after primary excision. Four years after the first operation, she developed meningoencephalocele through the defect in her skull from the excision of the skull osteoma. No surgical intervention was necessary as she had no neurological deficit and had no other symptoms.

DISCUSSION

A detailed preoperative evaluation of these tumors is essential before embarking on any surgical approach, keeping in mind all possible postoperative complications. MRI is the best diagnostic aid for these tumors. On T1-weighted images, tumors are isointense or slightly hyperintense to muscles. On T2-weighted images, benign tumors may show a target pattern with peripheral hyperintense rim and central low intensity or may be variably inhomogeneous. MRI cannot distinguish schwannomas from neurofibromas, and even benign tumors may mimic malignant NSTs when cystic, hemorrhagic, and necrotic degeneration is present.¹ In the STIR imaging, fat is suppressed leading to increased conspicuity of the lesion. MRI is better than CT for assessment of intracranial tumor spread of NSTs, particularly postcontrast T1-weighted images with frequency selective fat suppression.²

It is nearly impossible to differentiate between a neurofibroma and a schwannoma clinically; sometimes it is even quite challenging distinguishing schwannomas from neurofibromas histologi-

cally. Verocay bodies (nuclear palisades separated by anuclear areas) and hyalinized blood vessels are two distinguishing features of a schwannoma. Both are S100 protein positive but can be differentiated by employing a stain to identify axons. The neurofilament antibody labels the axons brown separated by neoplastic cells within the neurofibroma. In schwannomas, neurofilament antibody labels axons in the nerve adjacent to the tumor.

NSTs of the skull base may reach a considerable size before the diagnosis is made because of a natural slow growth rate and absence of symptoms. In a series of 13 patients with lower cranial NSTs of the infratemporal fossa over a 10-year period, Leonetti et al found that the most common presentations were pressure symptoms and voice change; where submucosal palatal fullness and vocal cord paralysis were the most common visible signs. The most common postoperative complication was dysphonia.³

In a series of 33 cases of extracranial solitary neurogenic tumors reported by Katz and McAlpin, four patients with vagus nerve tumor had permanent ipsilateral cord paralysis despite careful dissection. Our experience is very similar.⁴ In another retrospective analysis of 25 neurogenic tumors of the head-neck area by Kempf et al, 23 patients underwent operations, and 18 tumors arose from cranial nerves. One of two great auricular nerve autografts in cases of middle ear facial nerve neuroma resulted in an adequate facial muscle tone. There was no recurrence in benign tumors.⁵ We had similar results of facial nerve grafting.

In a series of 26 cases of nonvestibular head and neck schwannomas over a period of 20 years, Malone et al found that the parapharyngeal space was the most common site of tumor origin, occurring in 31% of cases.⁶ Symptoms secondary to a mass effect or a nerve deficit occurred in 69%, and the nerve of origin was identified in 62%. Twenty-three patients (88%) had complete surgical excision, and three patients (12%) had subtotal resection. Postoperative nerve deficit was apparent in 16 patients (62%), with resolution in 7 patients. Malone et al opined that a complete tumor removal was

often achieved, but subtotal or near-total resection might have been indicated for patients with extensive skull base, middle ear, or facial nerve involvement.⁶ They were more successful than us in identifying the nerve of origin. The postoperative complication was as high as ours.

Toriumi et al suggested that the vast majority of head and neck nerve tumors are benign.⁷ Correct management requires accurate preoperative evaluation and anticipation of complications. Complete resection is the ideal treatment but debulking procedures have a definite role. Micro-neural dissection and reconstruction should be attempted in all cases.⁷

CONCLUSIONS

Surgeons should be aware that a solitary mass in the region of the cranial nerves, brachial plexus, cervical sympathetic chain, or a major peripheral nerve could be an NST. If they are resected without adequate consideration, major and permanent nerve deficits can occur. Every effort should be made to establish a preoperative histological diagnosis of extracranial skull base lesions, although it is difficult to achieve. Establishing the nerve of origin of NSTs is important for the planning of reconstructive surgery and for patient consent.

The surgical complication rate is high, and combining the skills of various surgical disciplines during operation is essential. The authors emphasize the multidisciplinary approach for the management of postoperative morbidity including speech

and language therapists, physiotherapists, and occupational therapists. Nerve grafting and other reconstructive surgery should be used in all cases where indicated to restore function. A long-term follow-up plan is helpful in detecting residual tumors, identifying delayed complications, monitoring rehabilitation, and planning surgery to improve quality of life.

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