The Journal of Laryngology and Otology
March 1992, Vol. **106**, pp. 243–246

Parapharyngeal neurofibromas

D. A. TANDON, S. BAHADUR, N. K. MISRA, R. C. DEKA, K. KAPILA (New Delhi, India)

Abstract

This paper is a retrospective study of 15 patients with parapharyngeal neurofibromas operated over a six year period. No patient presented with a neurological deficit. CT scans revealed a well encapsulated moderately enhancing mass in ten cases. The tumour was removed by a transcervical approach in ten cases while in five a cervical-transpharyngeal route with mandibulotomy was used; in three of the latter group the tumour was retropharyngeal. In two cases a recurrence was successfully excised. It was realized that a mandibulotomy was required when the internal carotid was displaced medially, or if the tumour extended to the base of skull or when its vertical diameter exceeded 8 cm.

Introduction

Parapharyngeal lesions are uncommon. Neoplasia commonly encountered in this region are of salivary or neural origin. Benign nerve sheath tumours have varied from 7 per cent (Maran *et al.*, 1984) to 21 per cent (Shoss *et al.*, 1985) of all tumours, with deep lobe parotid and glomus tumours forming the majority. Most series deal comprehensively with the management of the latter two varieties of tumours. The present paper is devoted exclusively to considerations involved in management of parapharyngeal neurofibromas.

The parapharyngeal space is an inverted pyramid with its base being the base of skull and the apex the greater horn of hyoid bone. The superior constrictor bounds it medially, while the lateral surface is formed before backwards by the medial pterygoid, mandibular ramus, deep lobe of the parotid and the posterior belly of the digastric muscle. The vertebral column and the paravertebral muscles form the posterior boundary. The fascia enveloping the styloid apparatus extends anterolaterally to merge with the fascia covering the tensor veli palatini muscle. This divides the space into an anterior (prestyloid) and a posterior (retrostyloid) space. The latter contains the internal carotid artery, internal jugular vein, sympathetic trunk and the last four cranial nerves. A posterolateral extension to the prevertebral fascia from the carotid sheath separates the medial portion of the retrostyloid compartment from the retropharyngeal space.

This paper describes our experience with 15 cases of parapharyngeal neurofibromas and attempts to establish specific indications for a mandibulotomy.

Materials and methods

From 1984 to 1989, 15 patients with parapharyngeal neurofibromas were treated at the All India Institute of Medical Sciences, New Delhi. This is a retrospective study of these patients. Records of all patients and the respective histological and radiological material were reviewed.

Besides a complete physical examination, each patient underwent a CT scan (axial and coronal sections) with intravenous contrast and a fine needle aspiration cytology (FNAC) to establish the diagnosis and determine the surgical approach. The aspiration was done from the most prominent part of the swelling, whether external or oropharyngeal. An angiogram was not considered necessary for any patient.

All patients were operated on under hypotensive general anaesthesia. All patients were available for follow-up for six months to five years.

Observations

There were nine males and six females with an average age of 29.9 years (Table I). All but one had a virgin tumour (case 1). Case 1 presented with a second recurrence having been operated on twice earlier elsewhere. No patient had a neurological deficit or respiratory distress.

Eight patients presented with a neck mass with only a slight medial displacement of the lateral pharyngeal wall. In four cases the tumour was equally evident in the neck and on oropharyngeal examination. Three patients had a retropharyngeal tumour with a normal neck.

In nine cases FNAC yielded adequate material. The tumour comprised of spindle-shaped cells with thin wavy cytoplasm and cigar shaped nuclei in a fibrillary background. At times the cells were arranged in a palisading pattern. In three of these cases moderate pleomorphism was observed. In five cases a few drops of fluid were obtained which on microscopic examination was suggestive of a cystic change. In one case with a recurrent tumour the yield was inadequate for a morphological diagnosis.

Coronal CT sections were taken where tumour was in close proximity to the skull base to rule out bony erosion

Accepted for publication: 19 November 1991.

TABLE I
SHOWING THE PATIENTS' PROFILE

S no	Age	Sex	Clinical features	Operative approach	Morbidity	Follow-up
1.	26	M	Third recurrence of oroph. and neck mass.	Cervical transpharyngeal with mandibulotomy	Horner's syndrome	Recurrence at 2 years
2.	25	M	Oropharyngeal and neck swelling	Transcervical		NED—3 years
3.	35	M	Oropharyngeal swelling	Cervical transpharyngeal with mandibulotomy		NED—3 years
4.	18	F	Oropharyngeal and neck mass	Transcervical		NED—8 months
5.	40	M	Oropharyngeal mass	Cervical transpharyngeal with mandibulotomy		NED—2 years
6.	23	M	Neck mass	Transcervical		NED—1 year
7.	21	F	Neck mass	Transcervical	X nerve palsy	NED—5 years
8.	38	M	Neck mass	Transcervical	Nil	Recurrent at 2 years
			Recurrence of mass	Repeat transcervical	X nerve palsy	NED 1.5 years
9.	31	F	Neck mass	Cervical transpharyngeal with mandibulotomy		NED—1 year
10.	35	F	Neck mass	Transcervical		NED—3 years
11.	38	M	Neck mass	Transcervical		NED—1.5 years
12.	30	F	Neck mass	Cervical transpharyngeal with mandibulotomy	X and XII nerve	NED—6 months
13.	28	M	Neck mass	Transcervical		NED—2.5 years
14.	25	M	Neck mass	Transcervical	Horner's syndrome	NED—5 years
15.	35	M	Neck mass	Transcervical		Recurred at 6 months
			Recurrence of mass	Repeat transcervical		NED—4 years

*NED—No evidence of disease.

and an intracranial extension (Fig. 1). Erosion of the transverse process was seen at C3 and C4 in the case with recurrent tumour. This patient had undergone excision twice earlier by transcervical route, and presented with a tumour of limited mobility. In eight cases the internal jugular vein was seen on CT on the lateral surface of the tumour. It could not be identified in five cases with large tumours due to compression over the surface of the mass. In no case was the jugular vein displaced over the medial surface of the tumour. Central necrosis of the tumour (Fig. 1) and moderate enhancement was seen in ten cases. The parapharyngeal fat plane was identifiable on CT in nine cases (Fig. 2); it could not be identified in three patients with very large tumours probably due to severe compression of the connective tissues in the parapharyngeal space.

Ten patients underwent a transcervical excision of the tumour (Table I). The nerve of origin was identified in only one case in which the tumour involved the vagus nerve which had to be sacrified (case 7). Another patient developed Horner's syndrome post-operatively. The tumour recurred in two cases; in both a transcervical excision was performed again. Inadvertent injury to the vagus in one of the latter resulted in vocal cord paralysis. Both cases are tumour-free, four and one and a half years later respectively.

In five patients a cervical-transpharyngeal approach with mandibulotomy was utilized. In cases 1, 3 and 5 the

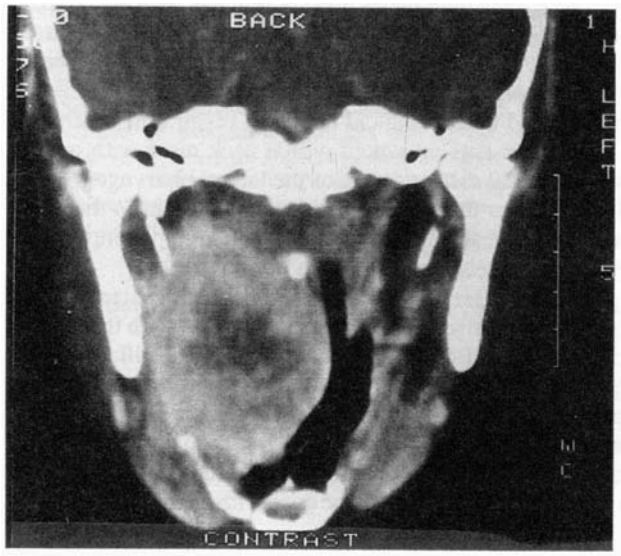

FIG. 1
Coronal CT section showing a large well defined parapharyngeal mass with moderate enhancement and a central necrotic area. Note the mass extending from hyoid to the base of skull (case 12).

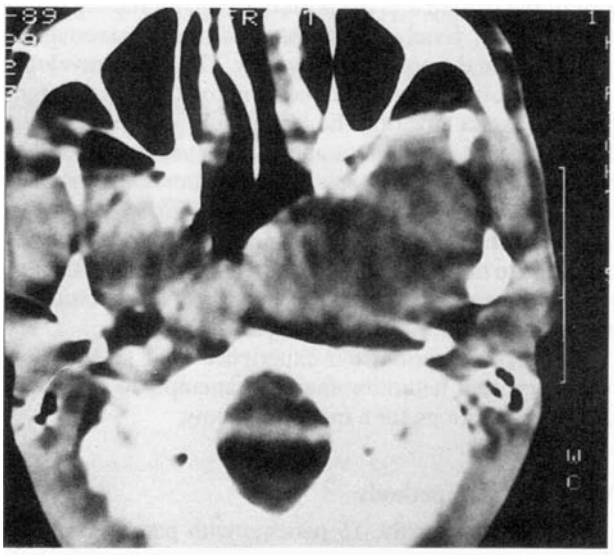

FIG. 2
Axial CT cut at the level of the maxillary sinuses showing a right sided parapharyngeal tumour, on the lateral surface of which the fat plane is identified (case 11).

https://doi.org/10.1017/S0022215100119164 Published online by Cambridge University Press

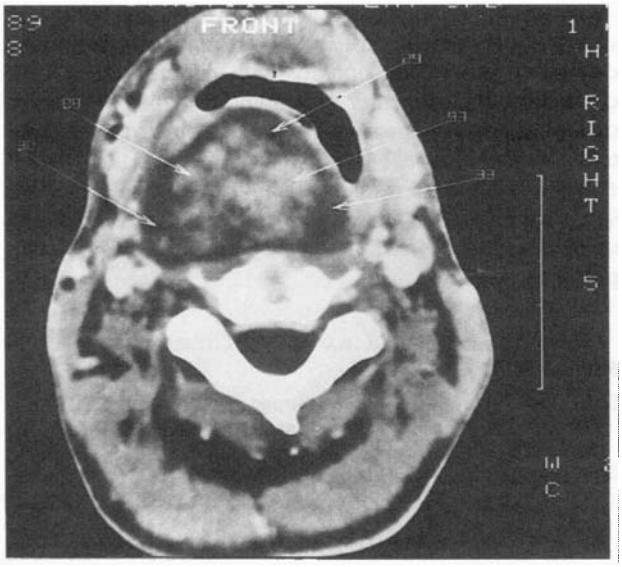

FIG. 3

A large lesion in the retropharyngeal compartment which required a mandibulotomy for removal. Note the irregular attenuation (case 5).

bulk of the tumour was in the retropharyngeal compartment (Fig. 3). In two others the tumour extended to the base of skull necessitating a mandibulotomy for tumour mobilization. Post-operatively Horner's syndrome developed in one patient (case 1) from inadvertent injury to the sympathetic trunk due to dense peri-tumoural fibrosis causing adhesions with the vertebral column and base of skull and necessitating ligation of the internal jugular vein and external carotid artery. Vocal cord paralysis occurred in another patient in whom the tumour originated from the vagus. In this patient a hypoglossal palsy also resulted from damage during dissection. The tumour recurred in case 1 two years later, but was declared inoperable. There was no evidence of malignant transformation on histological study of the specimen. The remaining four cases have been tumour-free for six months to three years.

Discussion

An expanding mass originating in this area will present with medial displacement of the lateral pharyngeal wall and/or as a mass emerging from beneath the ramus of mandible. Symptoms are usually related to tumour bulk and encroachment on nearby structures since these tumours may grow to a large size. Hearing loss due to Eustachian tube dysfunction, though rare (Maran *et al.*, 1984), may be the presenting feature. Work and Hybels (1974) have described hypoglossal palsy resulting from stretching of the nerve over a large vagal schwannoma. Horner' s syndrome from sympathetic involvement must not be overlooked. Benign tumours rarely cause a neurological deficit, which if present suggests a malignancy or jugular foramen involvement. Swallowing problems are usually the result of displacement of the palate or pharyngeal wall by the tumour. The pattern of pharyngeal displacement is variable: parotid tumours displace the whole tonsil and fauces, whereas neurogenic lesions displace the posterior part of the pharynx and posterior pillar.

Schwannoma was the third most common of 101 parapharyngeal space tumours seen at the Mayo Clinic prior to 1958 (Das Gupta *et al.*, 1969). Of the 40 cases reported by Work and Hybels in 1974, schwannomas were the second most common (22.5 per cent). Fifty per cent of all schwannomas arise from the vagus (Maniglia *et al.*, 1979). Though neurofibromas are rarer, their precise incidence is unclear. The glossopharyngeal nerve is rarely involved (Reddick and Myers, 1972).

Cystic degeneration and haemorrhage are characteristic and may alter the consistency and the colour of the tumour may vary from yellow to red. Ten of the 15 cases in the present series showed central cystic degeneration. The tumour was typically firm and encapsulated.

Neurofibromas may engulf the nerve of origin (Heeneman, 1988) in contrast to schwannomas, making preservation of the nerve difficult. The nerve of origin may not always be evident. In this study also, the nerve could be identified in only two cases (vagus in both). When tumour arises from the vagus, vocal cord paralysis is inevitable, though some rehabilitation is possible by Teflon paste injection.

Benign mixed tumours of the parapharyngeal space will displace the internal carotid artery in a posteromedial direction, whereas neurofibromas may reposition the internal carotid in any direction due to the multiplicity of the nerves from which these lesions may arise.

Of prime importance on CT scanning is identification of the fat plane, (Som *et al.*, 1984) the presence of which suggests a tumour of extraparotid origin (Fig. 2). It was identifiable in nine patients as very large tumours may obliterate the plane by compression of parapharyngeal connective tissue. Coronal sections are helpful in assessing the superior extent of tumour (Fig. 3), base of skull erosion and intracranial extension. Neurofibromas show variable contrast enhancement. Highly vascular lesions may simulate chemodectomas on contrast enhanced CT. Nevertheless, presence of central necrotic/non-enhancing foci (Fig. 1) suggest a neurofibroma.

A per-oral biopsy in such lesions is contraindicated because of the possibility of both implantation and uncontrollable haemorrhage. Fine needle aspiration cytology from a prominent site, whether intra- or extra-oral, was the most useful method of diagnosis in this study. Characteristic tumour morphology on aspirates, a long history, absence of neurological deficit and preservation of soft tissue planes was taken as strongly suggestive of a benign nerve sheath tumour.

Anterior dislocation of the mandible doubles the access

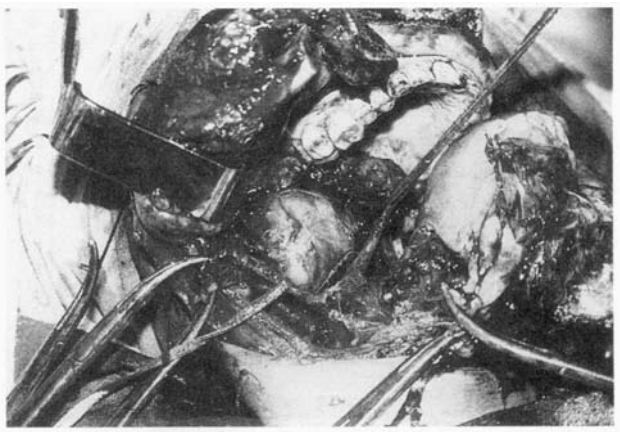

FIG. 4

A paramedian mandibulotomy and mandibular swing exposing widely the parapharyngeal space up to the base of skull (case 12).

https://doi.org/10.1017/S0022215100119164 Published online by Cambridge University Press

available to the parapharyngeal space (Maran *et al.*, 1984). However, the widest possible exposure is obtained only after a mandibulotomy. The cervical-transpharyngeal approach with mandibulotomy is reserved primarily for large extraparotid tumours (Som *et al.*, 1981). This facilitates protection of the major blood vessels, the lower four cranial nerves and also tumour mobilization.

The internal carotid artery comes to lie in a deeper plane as it ascends, particularly above the posterior belly of digastric. Also, close to where the internal carotid enters the carotid canal it is intimately related to the last four cranial nerves. This portion of the internal carotid is concealed from the surgeons view by the styloid apparatus, superficial to which lies the delicately walled internal jugular vein. Iatrogenic damage to the latter can be difficult to manage without wide access. This region is best exposed by a mandibulotomy and lateral mandibular swing (Fig. 4). Fortunately the large amount of loose areolar tissue in the parapharyngeal space allows easy isolation of the usually well encapsulated neurofibromas. Division of the sternocleidomastoid or a pre-emptive ligation of the external artery was not found to be necessary (Lau *et al.*, 1986) for augmenting exposure.

A paramedian mandibulotomy was done where the tumour extended into the retropharyngeal compartment (Fig. 3). Large tumours (8 cm or more in diameter) also required a mandibulotomy where blind dissection was considered unsafe because of adhesions to the base of the skull or when separation of a medially displaced internal carotid artery was hampered by the rigid lateral wall of the parapharyngeal space. A transcervical approach was adequate for tumours which though reaching a diameter of 8 cm did not impinge on the base of skull.

We prefer a stepped paramedian mandibulotomy as it yields an exposure as good as a mid-line bone cut without being prone to a delayed or a fibrous union (Tandon and Bahadur). A paramedian cut also facilitates a more secure stabilization of the mandibular segments. In addition, the incisor containing arch of the mandible is preserved as is the delicate geniohyoid-genioglossus muscle complex.

Conclusion

Parapharyngeal neurofibromas typically present with a long standing history of a throat/neck mass and without a neurological deficit. CT scans reveal a moderately enhancing mass, occasionally with central necrosis or cavitation. This, together with spindle shaped cells seen on aspiration cytology strongly suggest a neurofibroma. A transcervical approach usually suffices unless the tumour extends to the base of skull, or into the retropharyngeal space, or exceeds 8 cm in diameter, when a paramedian mandibulotomy is required. If completely excised, the prognosis is good.

Key words: Neurofibroma; Neck

References

Das Gupta, T. K., Brasfield, R. D., Strong, E. W., Hajdu, S. I. (1969) Benign solitary schwannomas (neurilemommas). *Cancer*. **24:** 355–366.

Heeneman, H. (1988) In Scott-Brown's *Otolaryngology*, fifth edn., Vol. 5, (Stell, P. M. and Kerr, A. G., eds.) Butterworths: London. p. 380–391.

Lau, W. F., Lam, K. H., Wei, W. (1986) Parapharyngeal space tumours. *Australian and New Zealand Journal of Surgery*. **56:** 835–842.

Maniglia, A. J., Chandler, J. R., Goodwin, W. J., Parker, J. C. (1979) Schwannomas of the parapharyngeal space and jugular foramen. *Laryngoscope*. **89:** 1405–1414.

Maran, A. G. D., MacKenzie, I. J., Murray, J. A. M. (1984) The parapharyngeal space. *Journal of Laryngology and Otology*. **98:** 371–380.

Reddick, L. P., Myers, R. T. (1972) Neurilemmomas of cervical portion of vagus nerve. *American Journal of Surgery*. **125:** 744–747.

Shoss, S. M., Donovan, D. T., Alford, B. R. (1985) Tumours of the parapharyngeal space. *Archives of Otolaryngology*. **111:** 753–757.

Som, P. M., Biller, H. F., Lawson, W. (1981) Tumours of the parapharyngeal space. *Annals of Otology, Rhinology and Laryngology*. **90 (Suppl 8):** 3–15.

Som, P. M., Biller, H. F., Lawson, W., Sacher, M., Lanzieri, C. F. (1984) Parapharyngeal space masses: a updated protocol based on 104 cases. *Radiology*. **153:** 149–156.

Tandon, D. A., Bahadur, S. (in press).

Work, P. W., Hybels, R. L. (1974) A study of tumours of parapharyngeal space. *Laryngoscope*. **84:** 1748–1755.

Address for correspondence:
Dr D. A. Tandon,
Assistant Professor,
Department of Otorhinolaryngology,
All India Institute of Medical Sciences,
New Delhi 110029,
India.