The Journal of Laryngology and Otology
December 1994, Vol. **108**, pp. 1102–1104

Fibromatosis of the parapharyngeal space

M. J. PORTER, M.A., F.R.C.S.*, W. M. SUEN, M.R.C.PATH.†, D. G. JOHN, F.R.C.S.*, C. A. VAN HASSELT, M.MED., F.C.S. (SA)*

Abstract
Tumours of the parapharyngeal space are not common. The majority arise from the deep lobe of the parotid gland or neurovascular structures. We describe a case of fibromatosis, which has not previously been reported at this site.

Key words: Fibromatosis; parapharyngeal space; Head and neck neoplasms

Case report

A 25-year-old male, Vietnamese boat person was referred from a refugee detention centre because of a swelling in the right side of his neck. It had first been noticed seven years earlier and had slowly enlarged since then. There was no history of trauma. A previous fine needle aspiration (FNA) and open biopsy, at another hospital, was reported as showing 'spindle cells of myofibroblastic origin', but no specific diagnosis had been reached.

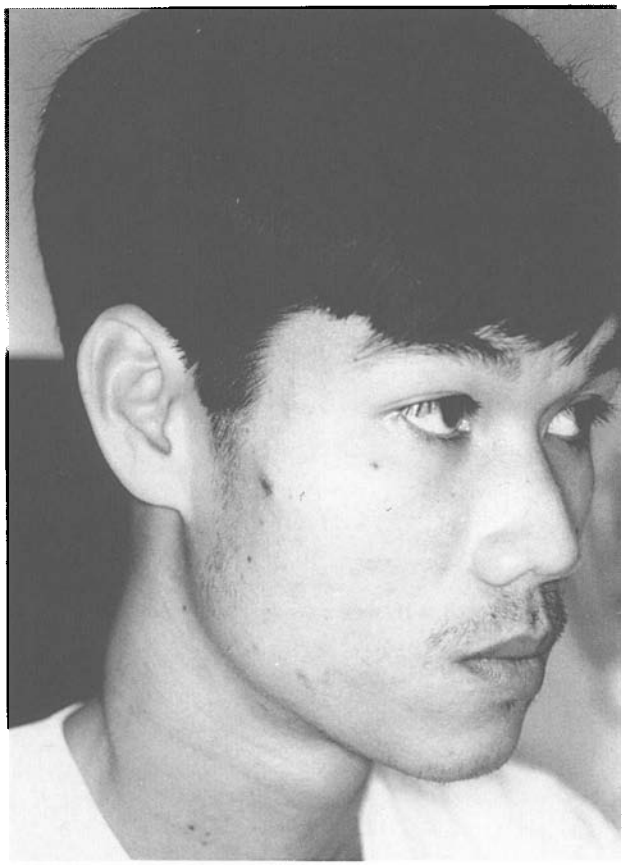

FIG. 1
Right side neck swelling.

He had no other complaints and on examination was a well man with a firm, non-pulsatile swelling which was smooth and fixed to the deep structures of his neck (Figure 1). He showed no bruit, no cranial nerve palsies and on oral examination the mass was seen to push the tonsil antero-medially. Ultrasound scans showed the mass to be 4 × 4 × 6 cm in size and solid. A CT scan showed a mass which extended from the level of the oropharynx down to the subglottic level (Figure 2).

At operation, through an upper cervical skin crease incision, the mass was seen to be lobulated with a smooth surface and could be separated from the overlying structures by blunt finger dissection. On its deep surface the mass was infiltrating the prevertebral muscles which had to be resected in order to obtain complete removal of the tumour. From the previous open biopsy it was difficult to distinguish whether the accessory nerve was entrapped by the tumour or by scar tissue and therefore it had to be sacrificed.

After the operation the patient had Horner's syndrome in addition to the accessory nerve palsy, but made an otherwise uneventful recovery. Horner's syndrome disappeared two years after the resection and there was no evidence of tumour recurrence.

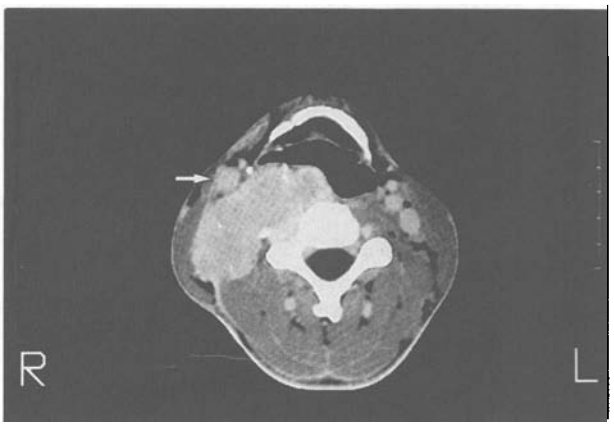

FIG. 2
CT scan at level of the hyoid bone showing anterior displacement of carotid sheath (arrowed).

From the Department of Surgery, Division of Otolaryngology* and the Department of Cellular and Anatomical Pathology†, Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, N.T., Hong Kong.
Accepted for publication: 22 July 1994.

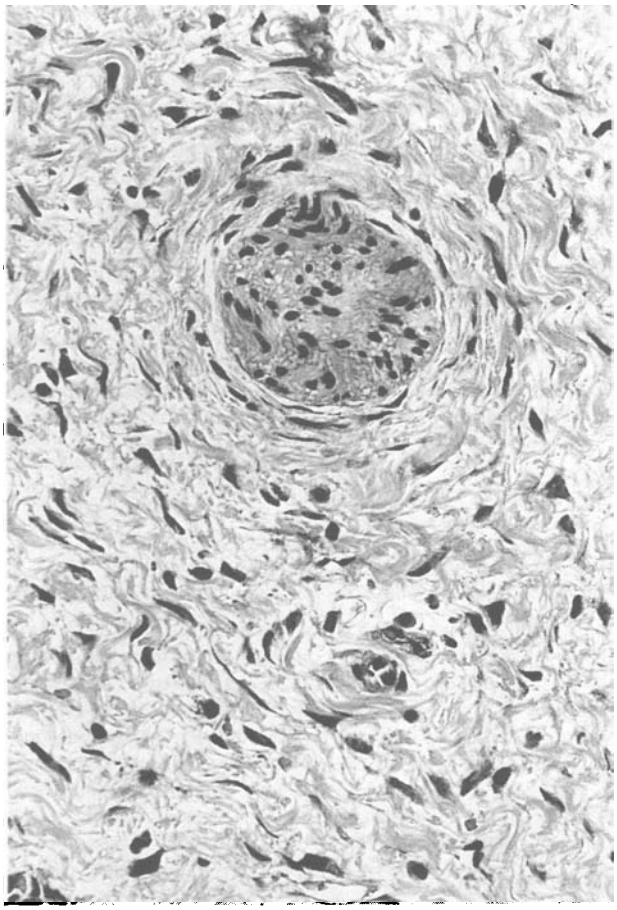

FIG. 3

Section of the tumour showing entrapment of a nerve by a fibromatosis (H&E; ×100).

Pathology

Microscopic examination of the tumour showed infiltrative fibroblastic proliferation with elongated spindle-shaped cells scattered within a collagenous stroma. There was no cellular atypia and mitoses were infrequent (less than one per ten high power fields). The margin was ill-defined. Entrapped nerves and muscles were seen (Figure 3). The spindle cells stained positively for vimentin but did not stain for S100 protein or desmin, thus demonstrating that the tumour was of mesenchymal derivation and not of neurogenic or muscular origin.

Discussion

The parapharyngeal space is a potential space in the neck. It has the shape of an inverted pyramid with its base on the body of the spenoid bone and its apex at the hyoid bone. Its medial boundary is the buccopharyngeal fascia lying over the superior pharyngeal constrictor muscle. The lateral boundary is the deep surface of the parotid gland and the ascending ramus of the mandible. Posteriorly there are prevertebral muscles covered by the prevertebral fascia. Anteriorly lie the pterygoid muscles. The styloid process divides the space into a pre-styloid compartment and a post-styloid compartment. The pre-styloid compartment contains the deep lobe of the parotid gland, the internal maxillary artery and the lingual, inferior alveolar and auriculotemporal nerves. The post-styloid compartment contains the internal carotid artery, the internal jugular vein, cranial nerves IX, X, XI and XII, the cervical sympathetic chain, glomus bodies and lymph nodes (Heenemann, 1987).

The distribution of parapharyngeal tumours as reported by different authors recent large series is shown in Table I. The miscellaneous group contains over 20 different pathologies both benign and malignant.

Enzenger (1988) classified fibromatoses as:

(1) Superficial (fascial) fibromatoses: (i) Palmer (Dupuytren's); (ii) Plantar (Ledderhoses disease); (iii) Penile (Peyronies' disease); (iv) Knuckle pads.

(2) Deep (musculo-aponeurotic) fibromatosis: (i) Extra-abdominal fibromatosis (extra-abdominal desmoid); (ii) Abdominal fibromatosis (abdominal desmoid); (iii) Intra-abdominal lipomatosis (intra-abdominal desmoid).

The peak incidence for deep fibromatosis occurs in the age group 25 to 35 years: 9.5 per cent occur in the head and neck and 22 per cent are found around the shoulder including the supraclavicular fossa.

Presentation is usually a painless, slowly-enlarging mass. Neurovascular structures may be displaced or encased rather than invaded, hence the lesions are relatively asymptomatic. Although preoperative imaging defines the lesion anatomically, an incisional biopsy is required for histological diagnosis. Initial misdiagnosis is not uncommon, partly due to an inadequate biopsy specimen. The definitive treatment is wide local resection in order to prevent local recurrence, although the proximity to vital structures makes this difficult in the head and neck. The morbidity and mortality of this condition is related to local recurrence and the effects of local spread.

Histologically the fibromatoses are seen to be poorly circumscribed with infiltrating boundaries. The cells are elongated spindle cells of uniform appearance with abundant collagen. The differential diagnosis histologically is from a fibrosarcoma to reactive fibrosis. Recurrence rates are 25–68 per cent notably in the pelvis and shoulder girdle and usually occur within one year of the initial treatment.

One large series of fibromatosis included several arising in the head and neck (Masson and Soule, 1966) whereas small series concentrated on fibromatosis in the head and neck alone (Conley *et al.*, 1966; El-Sayed 1992; Varghese *et al.*, 1992). In a review of 284 cases of fibromatosis at the Mayo Clinic, Masson and Soule (1966) reported 34 lesions in the head and neck region most commonly situated in the supraclavicular region or in the lateral neck. Three patients died due to spread into the chest or because of the tumour causing tracheal compression. There was a 75 per

TABLE I

DISTRIBUTION IN RECENT REPORTS OF PARAPHARYNGEAL TUMOURS ACCORDING TO THEIR PATHOLOGY. BECAUSE DIFFERENT AUTHORS USE A VARIETY OF CATEGORIES WITH WHICH TO CLASSIFY THESE TUMOURS, THE NUMBERS DO NOT NECESSARILY CORRESPOND EXACTLY TO THE FIGURES PUBLISHED IN THE ORIGINAL ARTICLES

Author	No.	Salivary	Neurogenic	Paraganglioma	Lymphoma	Carcinoma	Nodes	Miscellaneous
McIlrath *et al.* (1963)	101	43	16	12	25	0	0	5
Lawson *et al.* (1979)	6	1	1	0	0	1	0	3
Som *et al.* (1981)	30	8	6	5	0	2	1	8
Davidge-Pitts *et al.* (1983)	54	12	1	18	0	18	0	5
Maran *et al.* (1984)	66	53	1	4	0	0	5	3
Shoss *et al.* (1985)	42	16	9	6	1	3	1	6
Total	299	135	34	45	26	24	7	30
Percentage	100	45	11.3	15	8.6	8	2.1	10

https://doi.org/10.1017/S0022215100129020 Published online by Cambridge University Press

cent local recurrence rate after surgical excision and some patients required multiple procedures. Radical local surgery offers the only hope of eradicating the disease. The role of radiotherapy has not been firmly established due to the long natural history of the condition and the small number of cases. However, Masson and Soule (1966) favoured adjuvant radiotherapy in an attempt to prevent recurrence.

Fibromatosis is a rare condition in the head and neck. Diagnosis is difficult until surgery has provided the histopathologist with sufficient tissue to demonstrate the infiltrative nature of the disease. The surgeon must be prepared to undertake a radical resection and follow-up the patient for many years in order to detect any local recurrence.

References

Conley, J., Healey, W. V., Stout, A. P. (1966) Fibromatosis of the head and neck. *American Journal of Surgery* **112:** 609–614.

Davidge-Pitts, K. J., van Hasselt, C. A., Modi, P. C. (1983) Parapharyngeal space tumours. *South African Journal of Surgery* **21 (2):** 83–91.

El-Sayed , Y. (1992) Fibromatosis of the head and neck. *Journal of Laryngology and Otology* **106:** 459–462.

Enzenger, F. M. (1988) Fibromatoses. In *Soft Tissue Tumours*. Ch. 6. (Enzenger, F. M., Weiss, S. W., eds.), C. V. Mosby, St Louis, pp 136–163.

Heeneman, H. (1987) Parapharyngeal space tumours. In *Scott-Brown's Otolaryngology*. 5th Edition. vol. 5. (Kerr, A. G., Stell, P. M., ed.), Butterworths, London, pp 380–391.

Lawson, V. G., Le Liever, W. C., Makerewich, L. A., Rabuzzi, D. D., Bell, R. D. (1979) Unusual pharapharyngeal lesions. *Journal of Otolaryngology* **8 (3):** 241–249.

Maran, A. G. D., Mackenzie, I. J., Murray, J. A. M. (1984) The parapharyngeal space. *Journal of Laryngology and Otology* **98:** 367–369.

Masson, J. K., Soule, E. H. (1966) Desmoid tumours of the head and neck. *American Journal of Surgery* **112 (4):** 615–622.

McIlrath, D. C., ReMine, W. H., Devine, K. D., Duckerty, M. D. (1963) Tumours of the parapharyngeal region. *Surgery, Gynaecology and Obstetrics* **116 (1):** 88–94.

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: preoperative evaluation, diagnosis and surgical approaches. *Annals of Otology, Rhinology and Laryngology* **90 (supp. 80):** (1.4).

Varghese, G., Murthy, P. S. N., Puranik, K. R., Kurien, M., Narayan, K. R. (1992) Fibromatosis of the dorsum of the neck. *Journal of Laryngology and Otology* **106:** 821–823.

Address for correspondence:
Professor C. A. van Hasselt, M.Med.F.C.S. (SA),
Department of Surgery,
Prince of Wales Hospital,
Shatin,
New Territories,
Hong Kong.

Fax: 646–6312.