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CLINICAL RECORD

Management of an incidental malignant peripheral nerve sheath tumour in the parapharyngeal space

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Abstract

Background: Parapharyngeal space tumours are uncommon and represent 0.5–1 per cent of all head and neck neoplasms; 20–30 per cent of these are malignant. Malignant peripheral nerve sheath tumours are rare and mostly encountered in patients with neurofibromatosis type 1. Only four cases of parapharyngeal space tumours have been reported in the English language in patients without neurofibromatosis type 1.

Case report: We report the case of a 64-year-old man with no stigmata of neurofibromatosis type 1, in whom a mass in the left pre-styloid region of the parapharyngeal space was an incidental finding following magnetic resonance imaging for investigation of cervical spine problems. The mass was consequently removed using a transcervical approach. A histological review revealed a low-grade malignant peripheral nerve sheath tumour.

Discussion: We consider the pathophysiology of this highly malignant tumour as well as the challenging anatomy of the parapharyngeal space and the surgical and other therapeutic modalities utilised to treat this condition.

Key words: Parapharyngeal Space; Peripheral Nerve Sheath Tumors; Surgical Exposure; Histology; Neurofibromatosis

Introduction

Parapharyngeal space tumours are uncommon and represent 0.5–1 per cent of all head and neck neoplasms.[1] Most of these tumours originate in the salivary glands or are neurogenic in nature; 20–30 per cent of the tumours are malignant.[2] Malignant peripheral nerve sheath tumours are rare and are mostly encountered in patients with neurofibromatosis type 1. Only four cases of parapharyngeal space tumour have been reported in the English literature in patients without neurofibromatosis type 1.[3,4] This case report describes an unusual presentation of parapharyngeal space tumour in a patient with no stigmata of neurofibromatosis, and reviews the literature on the clinical management of this rare condition.

Case report

We report the case of a 64-year-old man in whom a 6-cm mass in the left pre-styloid region of the parapharyngeal space was an incidental finding following magnetic resonance imaging (MRI) for investigation of cervical spine problems (Figure 1). The mass was asymptomatic.

A neck examination revealed a level II mass. Intra-oral inspection demonstrated anteromedial displacement of the tonsil and soft palate (Figure 2). There was no dysfunction in the patient's lower cranial nerves, vocal fold movement was normal and there were no stigmata of neurofibromatosis type 1.

A review of the patient's history revealed that the mass was visible on a previous MRI scan performed two years earlier for similar cervical problems. Comparisons with the subsequent MRI indicated that the mass had remained unchanged.

The differential diagnosis included pleomorphic adenoma, neurogenic tumour and lymph node pathology. Transcervical fine needle aspiration was performed under ultrasound guidance but it was inadequate for diagnosis.

Following a comprehensive review in the head and neck multidisciplinary team meeting, and after discussion with the patient, transcervical surgical excision was planned. The mass was removed with no complications. The patient made a full recovery and was discharged after 3 days.

A histological review revealed a low-grade malignant peripheral nerve sheath tumour. A subsequent computed tomography scan of the chest and abdomen showed no evidence of distant metastatic disease. The patient underwent adjuvant radiotherapy to the tumour bed with tight margins. At seven months post-operation, the patient remained disease-free.

Discussion

The peripheral nervous system consists of the cranial and spinal nerves, the sympathetic and parasympathetic divisions of the autonomic nervous system, and the peripheral ganglia. Peripheral nervous system tumours arise from neural crest derivatives, such as Schwann cells, perineurial cells and fibroblasts, and include neurofibroma, schwannoma and malignant peripheral nerve sheath tumours.[5] The latter are rare, highly aggressive tumours capable of arising *de novo*

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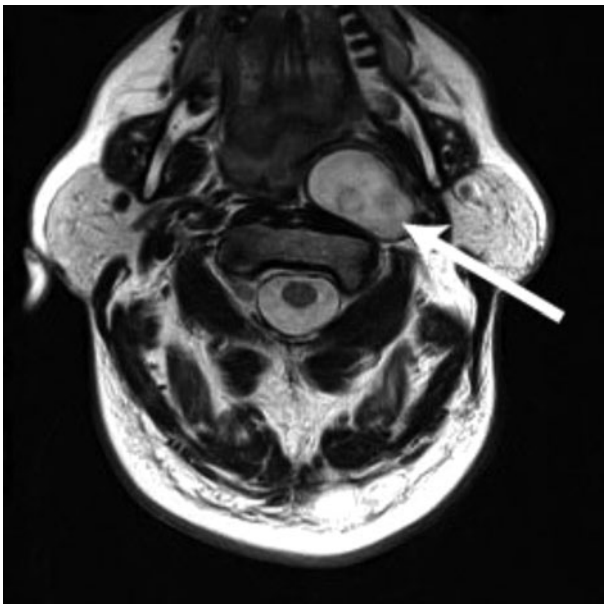

FIG. 1
Axial T2-weighted magnetic resonance image showing a mass (arrow) in the pre-styloid region of the left parapharyngeal space.

or from pre-existing benign neurofibromas or schwannomas. Up to 70 per cent of malignant peripheral nerve sheath tumours are associated with neurofibromatosis, and 9–14 per cent occur in the head and neck. There is an equal incidence amongst men and women and they commonly affect adults between 20 and 50 years of age.[3,6,7]

The parapharyngeal space is a complex anatomical area that is difficult to examine. It is often described as being in the shape of an inverted pyramid with the floor at the skull base and its tip at the greater cornu of the hyoid bone. The importance of the parapharyngeal space lies partly in its relationship with the other spaces of the neck. The masticator and parotid spaces are located laterally, the pharyngeal mucosal space is located medially and the retropharyngeal space is located posteromedially. The contents of the pre-styloid compartment include the minor or ectopic salivary glands, branches of the mandibular division of the trigeminal nerve, the internal maxillary artery, ascending pharyngeal artery and pharyngeal venous plexus. The contents of the post-styloid compartment include the internal carotid artery, internal jugular vein, IX–XIIth cranial nerves, the cervical sympathetic chain and glomus bodies. It is essential that the surgeon has a comprehensive understanding of the treatment of tumours in this space in order to reduce patient morbidity, and to provide adequate pre-operative counselling.[8]

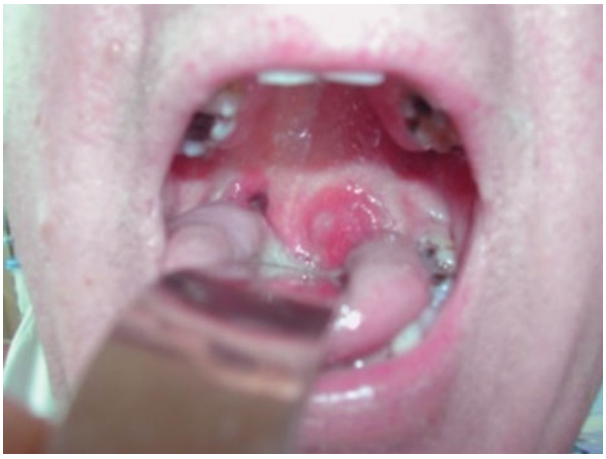

FIG. 2
Transoral view of the tumour extending from the parapharyngeal space.

The signs and symptoms of parapharyngeal space neoplasms can be subtle and clinical evaluation of this space is difficult. Lesions most often present as an oropharyngeal bulge or as a neck mass. Because of their location deep in the neck, tumours must reach a size of 2.5–3 cm before becoming palpable as a mass. They can therefore reach a large size prior to patient presentation, a fact which is demonstrated by the case reported here. Other signs and symptoms include: dysphagia, dyspnoea, unilateral middle-ear effusion, pulsatile tinnitus, bruit, thrill, otalgia, airway obstruction, hoarseness, foreign body sensation, true vocal fold palsy, Horner's syndrome, dysarthria, and symptoms of catecholamine excess such as hypertension and flushing.[2,9]

The differential diagnosis of a neoplastic mass in the parapharyngeal space commonly includes salivary gland tumour, neurogenic tumour, skull base and vertebral tumour such as meningioma or chordoma, rhabdomyosarcoma, and more rarely, Castleman's disease, which arises from adjacent nodes and extends into the space. Lesions of a non-neoplastic nature include branchial cleft cysts, lymphangiomas, and arteriovenous malformations and infective extensions arising from odontogenic, submandibular gland and pharyngeal tonsil sources.[7,10]

Magnetic resonance imaging is the imaging modality of choice for a suspected malignant peripheral nerve sheath tumour. The scan can highlight certain features that are more suggestive of this type of malignancy, such as the size of the mass (more than 5 cm), invasion of fat planes, heterogeneity, ill-defined margins and oedema surrounding the lesion. These tumours metastasise mainly to the lungs, followed by the bones. A computed tomography scan of the chest is therefore recommended once a histological diagnosis has been made.[11]

The mainstay of treatment for a malignant peripheral nerve sheath tumour of the parapharyngeal space is surgery. Surgical resection techniques described in the literature are classified as transoral, transcervical, transparotid–transcervical, transcervical–transmandibular or infratemporal. The appropriate choice of surgery depends upon accurate information with regards to mass size and location, the relationship between the tumour and the surrounding vessels and nerves, and its nature. The success of surgery depends on two conditions: correct identification and exposure of the lesion, allowing for complete removal; and minimum functional and aesthetic morbidity as a consequence of the surgery.[12]

A study by Kar *et al.* showed that routine nodal dissection is not indicated in patients with malignant peripheral nerve sheath tumours. However, the authors recommended that when a major nerve is identified, the cut end should be sent for frozen section to assess the tumour-free margin of the resection.[13] Malignant peripheral nerve sheath tumours are generally considered to be resistant to chemotherapy and radiotherapy. Nevertheless, post-operative radiotherapy is recommended by the Oncology Consensus Group as part

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of a uniform treatment policy for these tumours, much like other high-grade soft tissue sarcomas that have clear surgical margins.[14] Basso-Ricci demonstrated 56 per cent disease-free survival using combined surgery and radiation therapy.[15] This type of tumour has the highest recurrence rate of any sarcoma, and adequate initial treatment enables the best chance of survival.

Due to the relative rarity of malignant peripheral nerve sheath tumours, there have been few large studies investigating survival and recurrence rate. The local recurrence rate has been reported to range from 40 to 65 per cent. Similarly, the distant recurrence rate has been described as ranging from 40 to 68 per cent. Five-year survival has been reported to range from 16 to 52 per cent. Long-term survival has been correlated with complete surgical excision, small tumour size (less than 5 cm) and the presence of a low-grade component.[16–18] One recent study investigating patients treated at a sarcoma centre showed an overall survival rate of 84 per cent.[19] This has largely been attributed to improvements in imaging, which has led to early diagnosis and aggressive treatment, and the employment of adjuvant and neoadjuvant treatment modalities such as chemotherapy and radiation.

Future gains are likely to stem from a better understanding of the genetics and molecular biology of these tumours as well as improved imaging modalities, which will help to delineate the anatomy and facilitate complete surgical excision.

- **Malignant peripheral nerve sheath tumours are rare**
- **These tumours are even rarer in patients with no stigmata of neurofibromatosis**
- **We report a case in which the tumour was situated in the parapharyngeal space**
- **Histological diagnosis is important to determine the correct treatment modality**

Although these tumours are rare, this report highlights the importance of considering the diagnosis of a malignant peripheral nerve sheath tumour for neoplasms that arise in the parapharyngeal space.

References

1 Allison RS, Van der Waal I, Snow GB. Parapharyngeal tumours: a review of 23 cases. *Clin Otolaryngol* 1989;**14**:199–203
2 Carrau RL, Myers EN, Johnson JT. Management of tumours arising in the parapharyngeal space. *Laryngoscope* 1990;**100**: 583–9
3 al-Otieschan AA, Saleem M, Manohar MB, Larson S, Atallah A. Malignant schwannoma of the parapharyngeal space. *J Laryngol Otol* 1998;**112**:883–7
4 Lee JH, Lee HK, Choi CG, Suh DC, Lee KS, Khang SK. Malignant peripheral nerve sheath tumor in the parapharyngeal space: tumor spread through the eustachian tube. *AJNR Am J Neuroradiol* 2001;**22**:748–50
5 Weber AL, Montandon C, Robson CD. Neurogenic tumors of the neck. *Rad Clin North Am* 2000;**38**:1077–90
6 Bailet JW, Abemayor E, Andrews JC, Rowland JP, Fu YS, Dawson DE. Malignant nerve sheath tumors of the head and neck: a combined experience from two university hospitals. *Laryngoscope* 1991;**101**:1044–9
7 Woodruff JM, Selig AM, Crowley K, Allen PW. Schwannoma (neurilemmoma) with malignant transformation. A rare, distinctive peripheral nerve tumor. *Am J Surg Pathol* 1994;**18**:882–9
8 Shin JH, Lee HK, Kim SY, Choi CG, Suh DC. Imaging of parapharyngeal space lesions: focus on the prestyloid compartment. *AJR Am J Roentgenol* 2001;**177**:1465–70
9 Eisele DE, Netterville JL, Hoffman HT, Gantz BJ. Parapharyngeal space masses. *Head Neck* 1999;**21**:154–9
10 Shin JH, Lee HK, Kim SY, Khang SK, Park SH, Choi CG *et al*. Castleman's disease in the retropharyngeal space: CT and MR imaging findings. *AJNR Am J Neuroradiol* 2000;**21**:1337–9
11 Pilvaki M, Chourmouzi D, Kiziridou A, Skordalaki A, Zarampoukas T, Drevelengas A. Imaging of peripheral nerve sheath tumors with pathologic correlation: pictorial review. *Eur J Radiol* 2004;**52**:229–39
12 Fernández Ferro M, Fernández Sanroman J, Costas López A, Sandoval Gutiérrez JS, López de Sanchez AL. Surgical treatment of benign parapharyngeal space tumours. Presentation of two clinical cases and revision of the literature. *Med Oral Patol Oral Cir Bucal* 2008;**13**:61–4
13 Kar M, Deo SV, Shukla NK, Malik A, DattaGupta S, Mohanti BK *et al*. Malignant peripheral nerve sheath tumors (MPNST) – clinicopathological study and treatment outcome of twenty-four cases. *World J Surg Oncol* 2006;**4**:55
14 Ferner RE, Gutmann DH. International consensus statement on malignant peripheral nerve sheath tumors in neurofibromatosis. *Cancer Res* 2002;**62**:1573–7
15 Basso-Ricci S. Therapy of malignant schwannomas: usefulness of an integrated radiologic. Surgical therapy. *J Neurosurg Sci* 1989;**33**:253–7
16 Hruban RH, Shiu MH, Senie RT, Woodruff JM. Malignant peripheral nerve sheath tumors of the buttock and lower extremity. A study of 43 cases. *Cancer* 1990;**66**:1253–65
17 Kourea HP, Bilsky MH, Leung DH, Lewis JJ, Woodruff JM. Subdiaphragmatic and intrathoracic paraspinal malignant peripheral nerve sheath tumors: a clinicopathologic study of 25 patients and 26 tumors. *Cancer* 1998;**82**:2191–203
18 Wong WW, Hirose T, Scheithauer BW, Schild SE, Gunderson LL. Malignant peripheral nerve sheath tumor: analysis of treatment outcome. *Int J Radiat Oncol Biol Phys* 1998;**42**:351–60
19 Carli M, Ferrari A, Mattke A, Zanetti I, Casanova M, Bisogno G *et al*. Pediatric malignant peripheral nerve sheath tumor: the Italian and German soft tissue sarcoma cooperative group. *J Clin Oncol* 2005;**23**:8422–30

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Miss E Mushi takes responsibility for the integrity of the content of the paper
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