The Journal of Laryngology & Otology
December 2002, Vol. **116,** pp. 1050–1052

A solitary neurofibroma of the palatine tonsil

C.J. Surwald, M.D., D.D.S., M. A. Salam, M.S., M.Sc., M.D., F.R.C.S., F.R.C.S (Orl)*,
R. C. G. Rowe, M.B., B.S., F.D.S., F.R.C.Path.*

Abstract
A rare case of a tonsillar neurofibroma in a 28-year-old Caucasian male is presented. Benign tumours of the tonsils are rare and of those reported, only a few are benign peripheral nerve sheath tumours (BPNSTs). This is the first report of a solitary neurofibroma of the palatine tonsil in the English literature.

Key words: Neurofibroma; Tonsil; Nerve Sheath Tumours; Head and Neck Neoplasms

Introduction

Benign tumours of the palatine tonsil other than squamous cell papillomas are rare,[1] whilst benign peripheral nerve sheath tumours (BPNSTs) are extremely rare. These neoplasms include mainly schwannomas (neurilemmomas) and neurofibromas. In the head and neck, BPNSTs are usually seen in the soft tissues of the lateral cervical region and in the pharynx.[2] Schwannomas of the palatine tonsil have been reported sporadically with one recent case,[3] whereas to date only one case of a tonsillar neurofibroma has been documented in a 61-year-old Russian female.[4] This neurofibroma was found in association with two similar pedunculated lesions next to the tonsil. Unfortunately, no description of the histopathological findings was given. A unique case of a *solitary* neurofibroma of the palatine tonsil in a 28-year-old Caucasian male is presented here, the histopathological findings discussed and the pertinent literature reviewed.

Case report

Clinical and radiological findings

A 28-year-old Caucasian male presented with a year's history of a slowly progressing gagging sensation and painless fullness in his throat. There was an entirely negative family history of pigmented skin abnormalities or neural tumours. Clinical examination revealed a grossly enlarged right tonsil indenting the base of the tongue (Figure 1), a normal post-nasal space seen by fibre-optic laryngoscopy and no palpable cervical lymph nodes or stigmata of neurofibromatosis. Subsequent magnetic resonance (MR) imaging of the neck demonstrated a well-defined solid mass in the right palatine tonsil with increased signal on the T2-weighted and short tau inversion recovery (STIR) sequences respectively and of intermediate signal intensity on the T1-weighted sequences without invasion through the prevertebral fascia (Figure 2). No lymphadenopathy or other abnormalities were seen. The tumour was excised with complete resection of the right palatine tonsil. The patient recovered unremarkably, became asymptomatic and subsequent follow-up has not shown any evidence of recurrence.

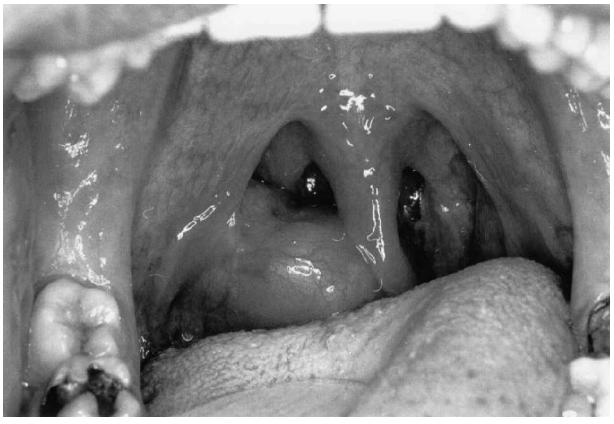

Fig. 1
Right tonsillar swelling indents the base of the tongue and displaces part of the uvula.

Pathological findings

Macroscopically, the specimen measured 53 × 20 × 25 mm and consisted of a discrete soft pale grey mass within tonsillar tissue. Microscopically, there was a discrete loose spindle-cell tumour within normal tonsillar tissue covered by intact mucosa. The uniform wavy tumour cells showed elongated nuclei with much cytoplasm and were set within plentiful loose extracellular tissue (Figure 3). There was neither evidence of Antoni A or B type patterns nor the presence of an associated peripheral nerve. Mitotic figures were absent. Immunohistochemical staining showed the spindle cells to be positive for S-100 protein (Figure 4). Staining for anti-CD34 was positive but negative for epithelial membrane antigen EMA). The results indicate Schwann cell and endoneurial differentiation. Furthermore, immunostaining with antibodies against neurofilament identified small nerve fibres. These features are consistent with a neurofibroma of the palatine tonsil and exclude a schwannoma. Surgical margins adjacent to the mass were free of tumour.

From the Departments of Otolaryngology/Head and Neck Surgery and Pathology*, Ipswich Hospital, Ipswich, UK.
Accepted for publication: 11 July 2002.

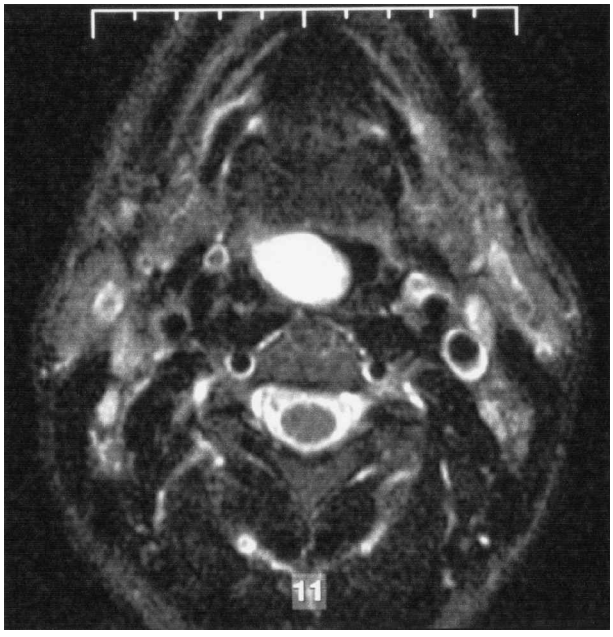

FIG. 2

Axial contrast MR scan (STIR) shows an enhancing mass within the right palatine tonsil without breaching the prevertebral facia.

A Medline and Embase search for reports of benign tumours, nerve sheath tumours, neurofibromas or head and neck neoplasms of the tonsil was carried out. A recent enumeration of benign tumours of the palatine tonsil[5] was also reviewed. In the papers retrieved and on analysis of their references cited, only one Russian report dating back to 1975 with similar characteristics to the present lesion was found.

Discussion

Among BPNSTs, schwannomas and neurofibromas are regarded as distinct entities.[6] Whilst schwannomas consist of Schwann cells only, neurofibromas consist of Schwann, perineurial and endoneurial cells and fibroblasts. Owing to the presence of distinct cell types, which are derived from both neural crest and mesenchymal precursors, a mixed or hamartomatous origin for neurofibromas has been suggested.[7] When compared with schwannomas, neurofibromas are usually less cellular, looser in texture, and without the characteristic pattern of alternating Antoni type patterns. Despite these histopathological differences, various electron microscopic, immunohistochemical and tissue culture studies support an alternative hypothesis, that neurofibromas may result from divergent differentiation of a single cell type.[8] This is supported by the documentation of somatic loss of heterozygosity and mutation of the neurofibromatosis 1 (NF1) gene in NF1-associated neurofibromas.[9] The existence of BPNSTs with both neurofibroma and schwannoma features also suggests that these subtypes are closely related neoplasms with varying degrees of schwannian differentiation.[10,11]

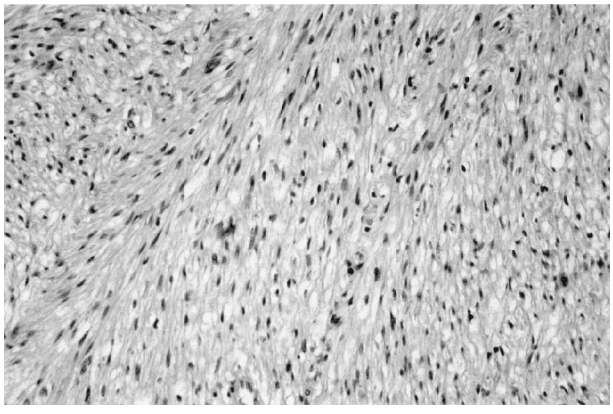

FIG. 3

The tumour is composed of loosely dispersed spindle cells in fibromyxoid stroma showing uniform elongated nuclei and pale cytoplasm (H & E; ×25).

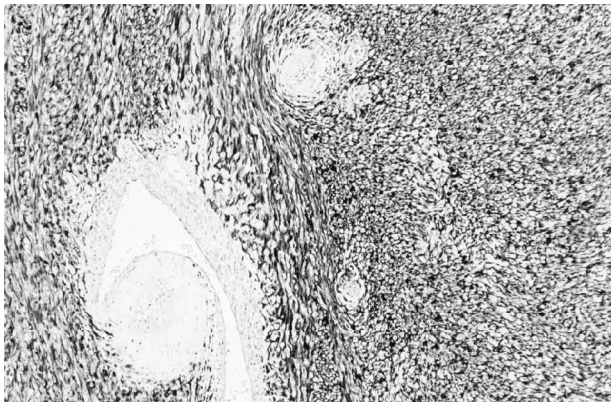

FIG. 4

The cytoplasm of the tumour cells diffusely stains positive for S100 protein (×10).

Neurofibromas are usually solitary tumours, however, up to a tenth of patients have multiple lesions. They may undergo malignant degeneration, especially when associated with von Recklinghausen's disease, whilst schwannomas very rarely do so.[12] In the head and neck, malignant transformation ranges between five and 12 per cent[13] which therefore requires thorough attention and knowledge of the manifestation of neurogenic tumours.

Macroscopically, neurofibromas appear as firm, circumscribed, mucosally covered red or grey masses. In the head and neck they are usually painless but may compress surrounding vital structures and so interfere with phonation, deglutition or respiration therefore necessitating early surgical intervention. Although history and examination are helpful, biopsy establishes a definitive diagnosis. Magnetic resonance (MR) imaging can assess tumour extent, invasiveness and cervical lymph nodes, but differentiation between different types of BPNSTs is not possible using MR imaging alone.[14] Based on the findings of clinical, histopathological and radiological investigations, complete surgical excision is the treatment of choice and is curative.

Acknowlegement

The authors would like to thank T. Nesterova, Ph.D., M.R.C. Clinical Sciences Centre, for her translation from Russian of an original article, which has been cited in this paper.

References

1 Heffner DK. Pathology of the tonsils and adenoids. *Otolaryngol Clin North Am* 1987;**20**:279–86
2 Hamza A, Fagan JJ, Weissman JL, Myers EN. Neurilemmomas of the parapharyngeal space. *Arch Otolaryngol Head Neck Surg* 1997;**123**:622–6

3 Lall GS, Walsh RM, Rowlands DC, Donaldson I. Schwannoma (neurilemmoma) of the tonsil. *J Laryngol Otol* 1999;**113**:585–6
4 Pen'kovskii GM. Neurofibroma of the palatine tonsil. *Vestn Otorinolaringol* 1975;**2**:91
5 Lupovitch A, Salama D, Batmanghelichi O. Benign hamartomatous polyp of the palatine tonsil. *J Laryngol Otol* 1993;**107**:1073–5
6 Woodruff JM. Tumors and tumorlike conditions of the peripheral nerve. In: Chung EB, Cavazzana AO, Ninfo V, eds. *Tumors and Tumorlike Lesions of Soft Tissue: Contemporary Issues in Surgical Pathology*. New York, London: Churchill Livingstone, 1991;**18**:205–28
7 Erlandson RA, Woodruff JM. Peripheral nerve sheath tumors: an electron microscopic study of 43 cases. *Cancer* 1982;**49**:273–87
8 Kharbanda K, Dinda AK, Sarkar C, Karak AK, Mathur M, Roy S. Cell culture studies on human nerve sheath tumors. *Pathology* 1994;**26**:29–32
9 Serra E, Rosenbaum T, Winner U, Aledo R, Ars E, Estivill X, *et al.* Schwann cells harbor the somatic NF1 mutation in neurofibromas: evidence of two different Schwann cell subpopulations. *Hum Mol Genet* 2000;**9**:3055–64
10 Kayem MJ, Dufour JJ, Robert F. Development of a schwannoma within a facial nerve neurofibroma: a case report and literature review. *Otolaryngol Head Neck Surg* 1995;**112**:483–7
11 Feany MB, Anthony DC, Fletcher CD. Nerve sheath tumours with hybrid features of neurofibroma and schwannoma: a conceptual challenge. *Histopathology*, 1998;**32**:405–10
12 Loree TR, North JH, Jr., Werness BA, Nangia R, Mullins AP, Hicks WL, Jr. Malignant periperhal nerve sheath tumors of the head and neck: analysis of prognostic factors. *Otolaryngol Head Neck Surg* 2000;**122**:667–72
13 Rapado F, Simo R, Small M. Neurofibromatosis type 1 of the head and neck: dilemmas in management. *J Laryngol Otol* 2001;**115**:151–4
14 Som PM, Curtin HD. Lesions of the parapharyngeal space. Role of MR imaging. *Otolaryngol Clin North Am* 1995;**28**:515–42

Address for correspondence:
M. A. Salam, F.R.C.S. (Orl),
Department of Otolaryngology/HNS,
The Ipswich Hospital,
Heath Road,
Ipswich IP4 5PD, UK.

Dr C. Surwald takes responsibility for the integrity of the content of the paper.
Competing interests: None declared
