The Journal of Laryngology & Otology (2011), ***125***, *869–872*.
© JLO (1984) Limited, 2011
doi:10.1017/S0022215111001290

CLINICAL RECORD

Leiomyosarcoma of the tonsil

B F JONES[1], V SRINIVASAN[1], K GUMPARTHY[2], D HUGHES[3]

Departments of [1]ENT and Head and Neck Surgery [2]Histopathology, and [3]Radiology, Arrowe Park Hospital, Wirral, UK

Abstract

Objective: We report a case of leiomyosarcoma arising in the tonsil in a 73-year-old man.

Method: Case report and review of the English language literature (using Pubmed, Ovid and Proquest databases).

Results: To our knowledge, this is the first case of tonsillar leiomyosarcoma to be reported in the English language literature. Our patient presented with haemoptysis, unilateral odynophagia and an ulcerated, exophytic lesion of the tonsil. Histological examination confirmed the diagnosis of leiomyosarcoma, and the patient was treated with radical radiotherapy.

Conclusion: Leiomyosarcomas are extremely rare in the head and neck; the common sites of origin are the skin and sinonasal tract. The overall prognosis is poor.

Key words: Tonsil; Leiomyosarcoma

Introduction

Leiomyosarcomas are rare, accounting for only 4 per cent of head and neck sarcomas.[1,2] They are malignant mesenchymal tumours and are most commonly seen within the gastrointestinal tract.[3] In the head and neck, leiomyosarcomas have been reported within the larynx, cervical oesophagus, skin, sinonasal tract and oral cavity, with most cases arising from the latter two sites.[4,5] Risk factors include previous irradiation and immunosuppression.[1,4,6]

We report a case of leiomyosarcoma arising from the tonsil in a 73-year-old man. Our patient presented with a short history of haemoptysis and unilateral odynophagia, with no identifiable risk factors. Clinical examination revealed an ulcerated lesion in the tonsil, and histological examination confirmed the diagnosis of leiomyosarcoma. To our knowledge, this is the first reported case in the English language literature of leiomyosarcoma arising from the tonsil.

Case report

A 73-year-old, male, ex-pipe smoker presented acutely with a two week history of left-sided odynophagia associated with haemoptysis. His past medical history was insignificant.

On examination, the patient had an enlarged left tonsil with an ulcerated, exophytic lesion. There was no palpable neck lymphadenopathy, and all inflammatory markers were within the normal range.

He was commenced on antibiotics; however, malignancy was suspected.

The patient underwent left tonsillectomy. Intraoperatively, a firm mass was found on the left tonsil, with no clinical invasion of immediate structures. The left tonsil was resected and sent for histological analysis. Endoscopy of the pharynx and larynx was normal.

A subsequent magnetic resonance imaging scan of the neck did not show any associated lymphadenopathy or extension of the tumour outside the tonsil (Figures 1 and 2), and a computed tomography (CT) scan of the mediastinum, abdomen and pelvis was normal.

Macroscopically, the resected tonsil measured 30 × 25 × 20 mm with focal mucosal ulceration. On sectioning, the tonsillar parenchyma was found to be almost completely replaced by a firm, pale tumour measuring 25 mm in maximum dimension. The entire tonsil was sectioned and processed.

Microscopic examination showed the specimen to consist mainly of interwoven bundles of spindle cells (Figure 3) with eosinophilic cytoplasm, nuclear pleomorphism and a high mitotic rate, including atypical mitoses (Figure 4). The more superficial tissue was formed by polygonal cells with pleomorphic nuclei and abundant eosinophilic cytoplasm. The overlying squamous epithelium was extensively ulcerated.

Extensive immunocytochemical analysis was performed, with strong positivity for smooth muscle actin, desmin and vimentin, focal expression of epithelial membrane antigen, and focal nuclear expression of myogenin. There was no expression of cytokeratins (i.e. MNF116, CAM 5.2, CK7, AE1 and AE3), p63, E-cadherin, BCl2, ALK-1, melanocytic markers (i.e.

Accepted for publication 6 November 2010 First published online 8 June 2011

https://doi.org/10.1017/S0022215111001290 Published online by Cambridge University Press

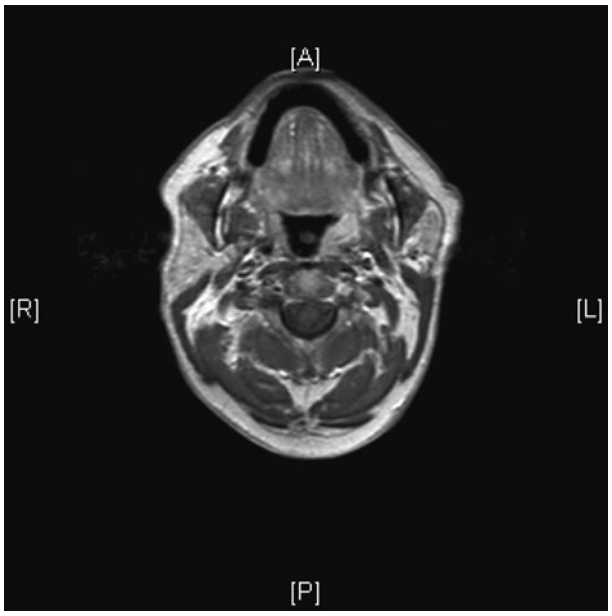

FIG. 1

Axial, T1-weighted, post-gadolinium magnetic resonance imaging scan taken after tonsillectomy. There is swelling and enhancement of the left tonsillar bed and lateral oropharyngeal wall, extending into the parapharyngeal space and medial pterygoid muscle. A = anterior; R = right; L = left; P = posterior

HMB45 and S-100), endothelial markers (i.e. cluster of differentiation 31 and 34 glycoproteins), factor 13a, CD117 or PGP 9.5. The surface squamous epithelium did not show any dysplasia, and the neoplasm did not arise from the squamous epithelium at any point. No conventional squamous cell carcinoma (SCC) was identified in any of the tissue planes examined. Since all cytokeratin markers were consistently negative, epithelial membrane antigen showed very focal

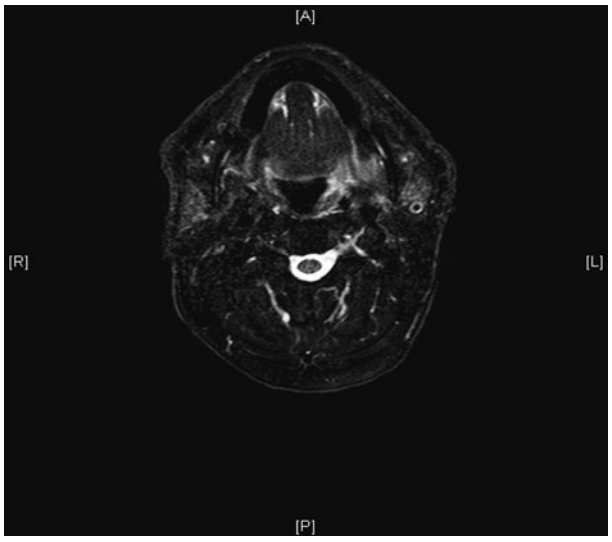

FIG. 2

Axial short T1 inversion recovery (STIR) magnetic resonance imaging scan taken after tonsillectomy. There is swelling and oedema of the left tonsillar bed and lateral oropharyngeal wall, extending into the parapharyngeal space and medial pterygoid muscle. A = anterior; R = right; L = left; P = posterior

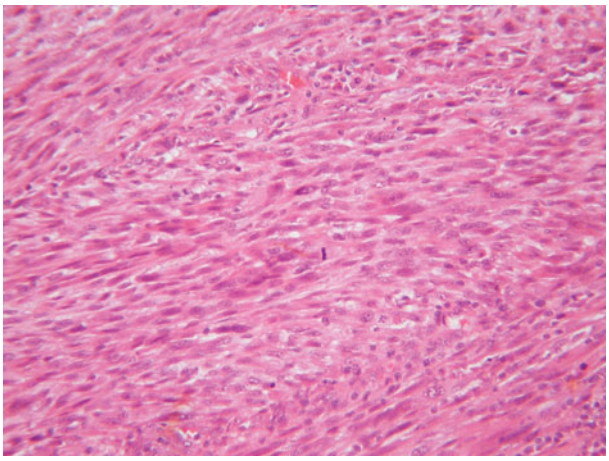

FIG. 3

Low power photomicrograph showing interlacing bundles of spindle cells with mitotic figures. (H&E; ×10)

immunoreactivity, myogenin showed focal nuclear expression, and smooth muscle actin, desmin and vimentin showed marked, extensive positivity, the appearances were those of a high grade spindle cell sarcoma, with an immunocytochemical pattern favouring a diagnosis of high grade leiomyosarcoma.

Post-operatively, the patient was treated with a course of radical radiotherapy.

At 18 months, although he was clinically well, a subsequent CT scan of the chest revealed pulmonary metastases. The patient was offered chemotherapy but refused further treatment.

Discussion

Leiomyosarcomas are malignant tumours originating from smooth muscle cells and myoepithelial cells of salivary glands.[3] Within the head and neck, these cells are mainly found in the walls of blood vessels and in the erector pili of the skin.[7] The first reported case in the English language literature was in a

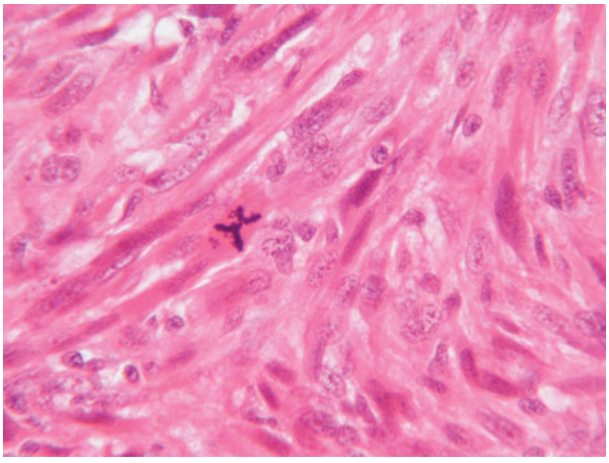

FIG. 4

High power photomicrograph showing an atypical mitotic figure within a spindle cell. (H&E; ×40)

https://doi.org/10.1017/S0022215111001290 Published online by Cambridge University Press

34-year-old patient who presented with a retromolar leiomyosarcoma.[8]

Such tumours account for approximately 7 per cent of all soft tissue sarcomas and are most commonly seen in the gastrointestinal tract, uterus and retroperitoneum. They are extremely rare within the head and neck, where they usually arise in the skin, larynx, cervical oesophagus and sinonasal tract.[3,4]

Factors associated with an increased risk of leiomyosarcoma include previous irradiation and immunosuppression (particularly due to human immunodeficiency virus).[9,10] Associations have also been reported with Epstein–Barr virus infection and with a number of syndromes, including retinoblastoma, Gardner's syndrome and Recklinghausen's neurofibromatosis.[5]

The clinical features of leiomyosarcoma are dependent on the site. Very few cases have been described in the pharynx, although hypopharyngeal lesions have been reported.[7,11,12] One patient presented with pyrexia of unknown origin, another with dysphagia and hoarseness.[12] Another patient with tumour arising from the posterior pharyngeal wall presented with symptoms of dysphagia and hypernasal speech.[13] Two further cases of pharyngeal leiomyosarcoma have been described, but their exact site was unknown.[14,15]

The differential diagnosis of leiomyosarcoma includes leiomyoma, malignant melanoma, spindle cell SCC and sarcoma. Diagnosing leiomyosarcoma based on microscopy alone can be difficult, and immunohistochemical and ultrastructural investigation may be required.[5,14]

In particular, spindle cell SCC can masquerade as leiomyosarcoma, as the conventional element of the SCC can be elusive and the spindle cell element can show striking morphological and immunohistochemical differentiation into sarcoma. Spindle cell SCC is usually a polypoid, mucosa-based tumour, and extensive sampling may be required to demonstrate an in situ or obvious epithelial infiltrating element. The epithelial component may show only focal cytokeratin positivity, although no immunoreactivity with desmin is noted.

The cell morphology of melanomas ranges from spindled to epithelioid, with prominent nucleoli. Pigment may or may not be present in the cytoplasm. However, these cells will be immunoreactive with at least one melanocytic marker, either S100 or HMB45.

Sarcomas (other than leiomyosarcoma) include myofibrosarcoma, fibrosarcoma, synovial sarcoma and malignant nerve sheath tumours. Myofibrosarcomas have tapered (rather than blunt-ended) nuclei and indistinct cell margins, and are immunoreactive for smooth muscle actin and (rather less frequently) for desmin. Fibrosarcomas have a sweeping fascicular arrangement with or without a herringbone pattern, they lack more than very focal muscle marker positivity, and they do not express S100 or epithelial markers. Synovial sarcomas have ovoid, smaller, rather uniform nuclei and scattered positivity for epithelial markers and BC12. Malignant peripheral nerve sheath tumours have serpentine nuclei and focal S100 positivity.

Our patient's tumour had an immunohistochemical profile characteristic of smooth muscle differentiation, with expression of smooth muscle actin and desmin. Immunoreactivity for cytokeratins was not detected, despite using a large panel of epithelial markers, except for very focal epithelial membrane antigen positivity, which can occur in leiomyosarcoma. No immunoreactivity was noted with S100, HMB45, BCL2 or ALK-1, thereby excluding melanoma, synovial sarcoma and myofibroblastic sarcoma. The surgical specimen was processed in its entirety, and there was no epithelial differentiation or dysplasia of the overlying squamous epithelium, which was ulcerated. Despite the various differential diagnoses to be considered in the case of a malignant spindle cell lesion, the findings for this tonsillar neoplasm were morphologically those of leiomyosarcoma. Moreover, extensive immunohistochemical staining was performed to establish that the neoplasm was a leiomyosarcoma; the findings of this staining excluded the presence of a carcinoma, melanoma or other type of sarcoma.

Contrary to previously reported pharyngeal leiomyosarcoma cases, which presented with chronic symptoms, our patient presented acutely with a two week history of odynophagia and haemoptysis. He underwent surgical resection followed by adjuvant radiotherapy; based on the current, limited literature, this would seem to be the treatment of choice. Certainly, for low grade tumours, complete surgical resection is considered to be the most effective treatment, although all treatment modalities have shown poor results when the lesion is high grade.[4] Radiotherapy is frequently used as an adjuvant, as chemotherapy is considered to be less effective.[3,16]

- **Leiomyosarcomas are rare within the head and neck**
- **This is the first reported case in the English language literature of leiomyosarcoma of the tonsil**
- **Leiomyosarcomas of the head and neck have a poor prognosis**

The behaviour of smooth muscle tumours is dependent of their site of origin, and can be unpredictable.[14,17] In the head and neck, leiomyosarcomas have been found to be aggressive tumours with a poor prognosis, with a five-year survival rate of 20 per cent.[3] Metastatic spread is common.[3,14] Leiomyosarcomas presenting within the oral cavity typically show a more aggressive pattern, with higher rates of local recurrence and metastasis.[17] In a series of 13 patients with leiomyosarcoma in the head and neck, 27 per cent suffered recurrence within six to 24 months, and 55 per cent eventually

https://doi.org/10.1017/S0022215111001290 Published online by Cambridge University Press

developed regional or distant metastases.[14] Indeed, 34 per cent of leiomyosarcoma cases have evidence of distant metastasis at the time of presentation.[1]

To our knowledge, our patient represents the first case of leiomyosarcoma of the tonsil to be reported in the English language literature. The patient presented acutely, and was treated with complete surgical excision and adjuvant radiotherapy. This case highlights the natural progression and prognosis of such tumours, and the treatment modalities available.

References

1 Pfeiffer J, Boedeker C, Ridder G, Maier W, Kayser G. Radiation-induced leiomyosarcoma of the oropharynx. *Diagn Pathol* 2006;**1**:1–6
2 Takahashi T, Higo R, Nakata H, Sugasawa M. Leiomyosarcoma of the hypopharynx: a case report. *Auris Nasus Larynx* 2008;**35**: 304–7
3 Ulrich C, Feiz-Erfan I, Spetzler R, Isaacs J, Hott J, Nakaji P *et al*. Sinonasal leiomyosarcoma: review of literature and case report. *Laryngoscope* 2005;**115**:2242–8
4 Ramkrishnan V, Said S, Kingdom T. Primary leiomyosarcoma of the sphenoid sinus. *Arch Otolaryngol Head Neck Surg* 2009;**135**:946–52
5 Maronio G, Bertino G, Mariuzzi L, Berhamin-Bracale A, Lombardo M, Beltrami C. Laryngeal leiomyosarcoma. *J Laryngol Otol* 2000;**114**:398–401
6 Grabowska S, Zimnoch L, Hubert E, Duraj E, Wincewicz-Pietrzykowska A. Radiation induced leiomyosarcoma. Review of literature and case report. *Otolaryngol Pol* 2007;**61**:744–9
7 Tommerup B, Mogensen C, Nielsen A. A leiomyosarcoma of the hypopharynx. *J Laryngol Otol* 1985;**99**:605–8
8 Geever EF, Ackerman LV, Dublin WB, Stout AP. Leiomyosarcoma of the buccal region. *Cancer Semin* 1949;**6**: 15–17
9 Sedghizadeh PP, Angiero F, Allen CM, Kalmar JR, Rawal Y, Albright EA. Post-irradiation leiomyosarcoma of the maxilla: report of a case in a patient with prior radiation treatment for retinoblastoma. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2004;**97**:726–31
10 Mbulaiteye SM, Prakin DM, Rabkin CS. Epidemiology of AIDS-related malignancies: an international perspective. *Hematol Oncol Clin North Am* 2003;**17**:673–96
11 Fuller AM, Van Vliet PD, Lillie JC, Devine KD. Pharyngeal leiomyosarcoma with fever of unknown etiology. *Arch Otolaryngol* 1966;**84**:96–8
12 Mankodi RC, Shah SS, Kanvinde MS, Joshi JS. Pharyngeal leiomyosarcoma. *J Laryngol Otol* 1970;**84**:327–30
13 Glover GW, Park WW. Pharyngeal leiomyosarcoma. *J Laryngol Otol* 1971;**85**:1031–8
14 Montgomery E, Golbulum J, Fisher C. Leiomyosarcoma of the head and neck: a clinicopathological study. *Histopathology* 2002;**40**:518–25
15 Lajer CB, Daugaard S, Hansen HS, Kirkegaard J, Holmgaard S, Christensen ME. Soft tissue sarcomas of the head and neck: a single-centre experience. *Clin Otolaryngol* 2005;**30**:176–82
16 Goda JS, Saravanan K, Vashistha RV, Kumar V, Gupta A. Leiomyosarcoma of the larynx: case report and review of the literature. *Ear Nose Throat J* 2008;**87**:283–7
17 Dry S, Jorgensen J, Fletcher C. Leiomyosarcomas of the oral cavity: an unusual topographic subset easily mistaken for non-mesenchymal tumours. *Histopathology* 2000;**36**:210–20

Address for correspondence:
Mr Venkat Srinivasan,
Department of ENT and Head and Neck Surgery,
Arrowe Park Hospital,
Upton CH49 5PE, UK

Fax: +44 (0)151 6047103
E-mail: cheenu51@hotmail.com

Mr V Srinivasan takes responsibility for the integrity of the content of the paper
Competing interests: None declared

https://doi.org/10.1017/S0022215111001290 Published online by Cambridge University Press