The Journal of Laryngology and Otology
January 1995, Vol. **109**, pp. 77–79

Pathology in Focus

Smooth muscle tumours of the larynx

D. C. McKiernan, M.B., B.S., G. W. R. Watters, F.R.C.S.

Abstract

Two cases of laryngeal smooth muscle tumours are reported: one a benign leiomyoma, the other a malignant leiomyosarcoma. These tumours may present diagnostic difficulties and immunocytochemistry is helpful in distinguishing smooth muscle tumours from other connective tissue neoplasms and spindle cell squamous carcinoma. Primary treatment in both cases should be surgical resection with small (T_1, T_2) malignant tumours suitable for partial laryngectomy. Post-operative radiotherapy may have a role in allowing a more limited surgical resection.

Key words: Leiomyoma; Leiomyosarcoma; Larynx

Introduction

Smooth muscle tumours are very rare in both the upper and lower respiratory tracts (Naresh *et al.*, 1993). In their review of the literature, Hellquist *et al.* (1994) found only 33 cases of leiomyoma of the larynx to add to their case. Leiomyosarcoma is even more unusual and to our knowledge there have been only 12 previously reported laryngeal cases (Chen *et al.*, 1991; Tewary and Pahor, 1991; Rowe-Jones *et al.*, 1994). We report two cases which presented to our department in the same month. Following histological examination one case was diagnosed as a benign leiomyoma, the other a malignant leiomyosarcoma. In view of the rarity of smooth muscle neoplasms in the larynx, their behaviour and response to treatment can only be inferred from what is known of these tumours at other sites.

Case reports

Case 1

A 28-year-old woman presented with a six-month history of a hoarse voice. She had no other symptoms and smoked 15 cigarettes a day. On examination, she had no cervical lymphadenopathy. Indirect laryngoscopy revealed swelling of the middle third of the right vocal fold, which was fully mobile.

Microlaryngoscopy was carried out under general anaesthetic. A firm, exophytic lesion was found arising from the middle third of the right vocal fold, extending into the laryngeal ventricle. The lesion stripped easily from the vocal fold. Post-operatively her voice was much improved and on review two weeks later the right fold looked well healed. Six months after surgery there was no sign of recurrent tumour.

The specimen contained fibrous tissue covered partially by benign ciliated respiratory epithelium and also benign squamous epithelium. In the stroma were two nodules composed entirely of spindle-shaped cells which showed no evidence of nuclear pleomorphism or atypical mitotic figures (Figure 1). Immunocytochemical staining was performed and the spindle cells were positive for vimentin and desmin, features which support a smooth muscle origin. Staining for cytokeratin was negative. On the basis of these findings the overall appearance is consistent with the diagnosis of a benign leiomyoma of the vocal fold.

Case 2

A 43-year-old man presented with a three-month history of a hoarse voice. He was a nonsmoker and had no other symptoms. Indirect laryngoscopy revealed a lesion anteriorly on the left vestibular fold, with both vocal folds fully mobile. Examination was otherwise normal and in particular he had no cervical lymphadenopathy. A chest X-ray was also normal.

Microlaryngoscopy was performed under general anaesthetic. The mass extended from the left vestibular fold across to the right vestibular fold above the level of the anterior commissure. Biopsies were taken and the mass debulked using a carbon dioxide laser. Histological examination (with the aid of immunohistology) showed the tumour to be a leiomyosarcoma. In view of this finding a repeat microlaryngoscopy was carried out eight weeks later. There was obvious residual tumour with the same distribution as noted before and repeat biopsies confirmed the diagnosis of leiomyosarcoma.

A CT scan of the larynx confirmed the tumour was limited to the supraglottis with no extralaryngeal extension. Therefore it was decided to carry out a supraglottic partial laryngectomy with a covering tracheostomy. Macroscopic clearance was achieved although the tumour was very close to the superior surface of the left vocal fold (Figure 2).

The patient made a good recovery and was successfully decannulated two weeks after surgery. He has a satisfactory voice although had some difficulty with swallowing. A temporary percutaneous gastrostomy has been fashioned for feeding. Histological examination confirmed that complete excision had been achieved although the inferior resection margin on the left side was within 2 mm of the tumour. In view of this he underwent post-operative external beam radiotherapy. He remains well with no signs of recurrent disease six months after surgery.

The tumour was partly lobulated and stretching the overlying respiratory-type epithelium. It consisted of spindle cells showing moderate pleomorphism, together with numerous mitoses,

From the Department of Otolaryngology, Radcliffe Infirmary, Woodstock Road, Oxford OX2 6HE.
Accepted for publication: 8 October 1994.

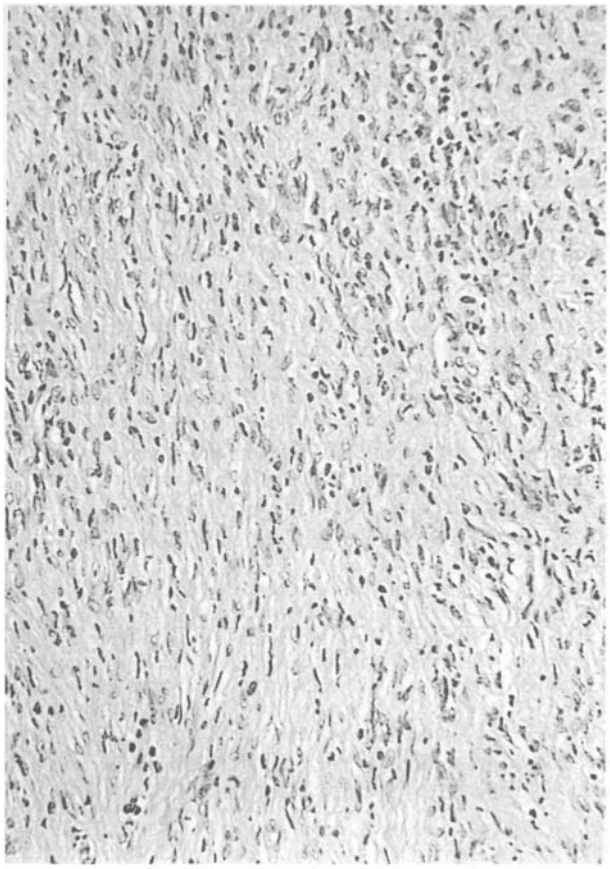

FIG. 1

High power of leiomyoma. Packets of fairly uniform spindle cells with long cigar-shaped nuclei are evident. A scattering of mononuclear cells lies between these cells. Mitotic figures amongst the spindle cells are not seen. (H&E).

many also bizarre (Figure 3). Immunocytochemistry showed positive staining for actin and desmin, supporting a smooth muscle basis for the tumour. Staining for cytokeratin was negative. These features supported the diagnosis of leiomyosarcoma.

Discussion

Smooth muscle tumours are rare in the head and neck as there is little smooth muscle compared with other sites in the body. It is presumed that head and neck tumours arise from smooth muscle in the walls of blood vessels or in the erector pili muscles or hair follicles. Kaya *et al.* (1990) speculate that another possible source may be aberrant, undifferentiated mesenchyme.

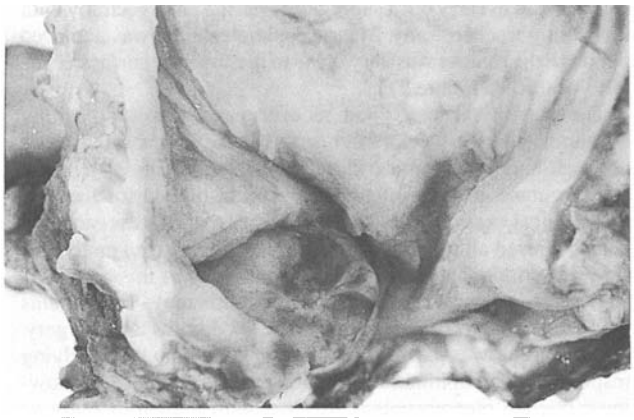

FIG. 2

Supraglottic laryngectomy specimen showing a leiomyosarcoma mainly involving the left vestibular fold and extending close to the interior resection margin.

Leiomyosarcoma is a very rare malignant neoplasm. As a proportion of malignant soft tissue tumours the percentage varies from 2.3 to 5.3 per cent (Fields and Helwig, 1981), and the percentage of leiomyosarcomas occuring in the neck is only three per cent (Goldberg *et al.*, 1988). The first case of laryngeal leiomyosarcoma was reported by Frank (1941). However definitive diagnosis in this case, and most of the subsequence cases, is debatable as immunohistology was not available. It is also possible, in the absence of immunohistology, that some cases of leiomyosarcoma have been misdiagnosed in the past as spindle cell squamous carcinomas.

Three different types of leiomyoma are recognized: these are the 'common' leiomyoma, the vascular leiomyoma (angiomyoma) and the epithelioid leiomyoma (leiomyoblastoma). All three have now been found in the larynx (Hellquist *et al.*, 1994). Benign leiomyoma is more common in males (ratio 2:1) with a mean age at presentation of 43 years (Hellquist *et al.*, 1994). Leiomyoma of the larynx has been reported in children and may present with respiratory obstruction owing to the smaller airway (Kaya *et al.*, 1990).

In the absence of metastatic spread it may be difficult to distinguish benign leiomyoma from leiomyosarcoma. The diagnosis of leiomyosarcoma relies on the size of the presenting tumour, the presence of nuclear pleomorphism and increased mitotic activity, with the number of mitotic figures per high power field being the most important criterion of malignancy (Mindell *et al.*, 1975). Smooth muscle neoplasms also need to be distinguished from other mesodermal tumours such as fibroma or fibrosarcoma (Kleinsasser and Glanz, 1979), and also spindle cell squamous carcinoma. This relies heavily on immunohistology in particular positive staining for desmin, which is an intermediate filament

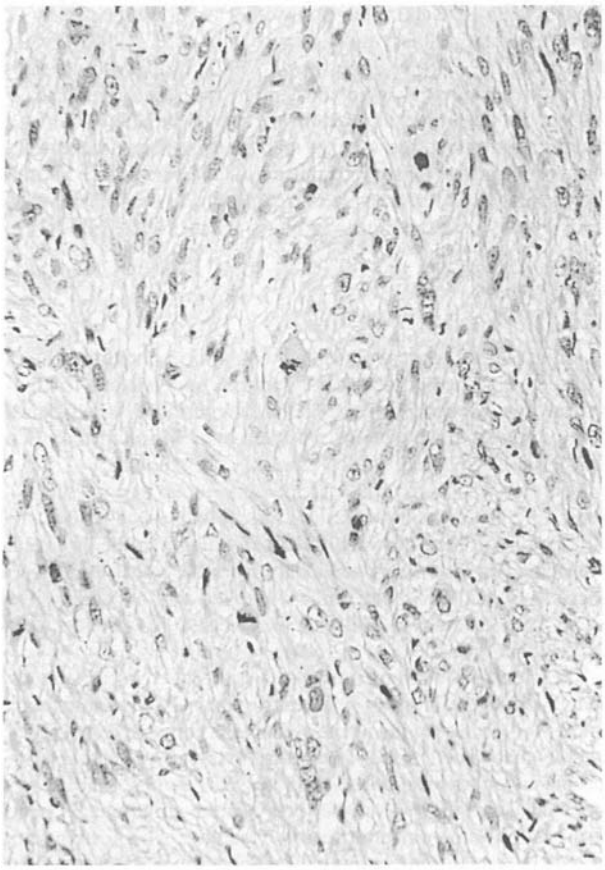

FIG. 3

High power of leiomyosarcoma. The cells are lying in groups with a basket weave pattern and include marked pleomorphism. Mitotic figures are numerous. (H&E).

https://doi.org/10.1017/S0022215100129317 Published online by Cambridge University Press

found in smooth muscle cells. Both of our cases gave a positive result with desmin markers. In our case of leiomyosarcoma the tumour would have been diagnosed as a spindle cell carcinoma were it not for immunocytochemistry (Michaels, personal communication). Other differential diagnoses may include schwannoma and malignant melanoma (Chen *et al.*, 1991).

Ultrastructural examination using an electron microscope can also be of help in identifying smooth muscle tumours (Kahn and Korol, 1989; Hellquist *et al.*, 1994), though is less readily available than immunohistology. Ultrastructural characteristics include pinocytotic vesicles, desmosomes and myofibrils. Fusiform dense bodies are present among parallel actin filaments (Michaels, 1987).

As with other malignant connective tissue neoplasms, metastatic spread of leiomyosarcoma is usually via the bloodstream to lungs and liver and may occur in 30–50 per cent of cases (Fields and Helwig, 1981). Early lymph node metastasis in head and neck leiomyosarcomas is rare (Mindell *et al.*, 1975). Leiomyosarcoma is not very sensitive to radiotherapy (Fu and Perzin, 1975; Kleinsasser and Glanz, 1979; Kuruvilla *et al.*, 1990) though post-operative radiotherapy may allow a more conservative surgical resection (Wile *et al.*, 1981). In the presence of metastatic disease chemotherapy may have a role in palliation (Josephson *et al.*, 1985), although much of the evidence in support of this is based on responses seen in abdominal leiomyosarcoma. The value of chemotherapy in head and neck leiomyosarcoma is questionable, particularly in view of the toxicity of chemotherapy regimes used for tumours at other sites. In the absence of wide spread metastatic disease treatment of laryngeal leiomyosarcoma should be primarily surgical resection and small tumours would seem to be ideally suited to partial laryngectomy, as in our *Case 2*. Chen *et al.* (1991) also treated a laryngeal leiomyosarcoma using a conservative surgical approach (vertical hemilaryngectomy) and post-operative radiotherapy, with a satisfactory outcome. Radical neck dissection is indicated for cervical nodal disease (Mindell *et al.*, 1975). Benign leiomyoma can be excised by local surgical resection. Usually this can be achieved using microlaryngoscopy techniques and use of the carbon dioxide laser may be helpful. However, for large subglottic tumours thyrotomy may be the best surgical approach (Karma *et al.*, 1978). Resection of vascular leiomyomas can be complicated by haemorrhage, but this stops completely after removal (Kleinsasser and Glanz, 1979).

The outcome in cases of benign leiomyoma should be excellent, with all cases cured by adequate surgical resection. However, prognosis of leiomyosarcoma is unpredictable. Rate of growth is related to the degree of differentiation but even in the better differentiated cases metastasis may appear several years after surgery (Michaels, 1987). Local recurrence is also seen a number of years after initial therapy, so patients with leiomyosarcoma require long-term follow-up (Mindell *et al.*, 1975).

Acknowledgements

We would like to thank Mr C. A. Milford, F.R.C.S., for allowing us to report these cases, and Dr P. R. Millard, M.D., F.R.C. Path., for his help and advice.

References

Chen, J. M., Novick, W. H., Logan, C.A. (1991) Leiomyosarcoma of the larynx. *Journal of Otolaryngology* **20:** 345–348.

Fields, J. P., Helwig, E. B. (1981) Leiomyosarcoma of the skin and subcutaneous tissue. *Cancer* **47:** 156–169.

Frank, D. I. (1941) Leiomyosarcoma of the larynx. *Archives of Otolaryngology* **34:** 493–500.

Fu, Y. S., Perzin, K. H. (1975) Nonepithelial tumours of the nasal cavity, paranasal sinuses, and nasopharynx: a clinicopathologic study. IV: Smooth muscle tumours (leiomyoma, leiomyosarcoma). *Cancer* **35:** 1300–1308.

Goldberg, S. H., Hanft, K., Ossakow, S. J. (1988) Residents page. Pathologic quiz: Case 1. *Archives of Otolaryngology, Head and Neck Surgery* **114:** 1330–1332.

Hellquist, H. B., Hellqvist, H., Vejlens, L., Lindholm, C. E. (1994) Epithelioid leiomyoma of the larynx. *Histopathology* **24:** 155–159.

Josephson, R. L., Blair, R. L., Bedard, Y. C. (1985) Leiomyosarcoma of the nose and paranasal sinuses. *Otolaryngology–Head and Neck Surgery* **93:** 270–274.

Kahn, J., Korol, H. (1989) Residents page. Pathologic quiz: Case 2. *Archives of Otolaryngology, Head and Neck Surgery* **115:** 114–117.

Karma, P., Hyrynkangas, K., Rasenan, O. (1978) Laryngeal leiomyoma. *Journal of Laryngology and Otology* **92:** 411–415.

Kaya, S., Levent, S., Sevket, R. (1990) Laryngeal leiomyoma. *International Journal of Pediatric Otorhinolaryngology* **19:** 285–288.

Kleinsasser, O., Glanz, H. (1979) Myogenic tumours of the larynx. *Archives of Otorhinolaryngology* **225:** 107–119.

Kuruvilla, A., Wenig, B. M., Humphrey, D. M., Heffner, D. K. (1990) Leiomyosarcoma of the sinonasal tract. *Archives of Otolaryngology, Head and Neck Surgery* **116:** 1278–1286.

Michaels, L. (1987) *Ear, Nose and Throat Histopathology.* Springer-Verlag, Berlin, p 443.

Mindell, R. S., Calcaterra, T. C., Ward, P. H. (1975) Leiomyosarcoma of the head and neck. *Laryngoscope* **85:** 904–910.

Naresh, K. N., Pai, S. A., Vyas, J. J., Soman, C. S. (1993) Leiomyoma of the bronchus: a case report. *Histopathology* **22:** 288–289.

Rowe-Jones, J. M., Solomons, N. B., Ratcliffe, N. A. (1994) Leiomyosarcoma of the larynx. *Journal of Laryngology and Otology* **108:** 359–362.

Tewary, A. K., Pahor, A. L. (1991) Leiomyosarcoma of the larynx: emergency laryngectomy. *Journal of Laryngology and Otology* **105:** 134–136.

Wile, A. G., Evans, H. L., Romsdal, M. M. (1981) Leiomyosarcoma of soft tissue: a clinicopathologic study. *Cancer* **48:** 1022–1032.

Address for correspondence:
Mr G. W. R. Watters, F.R.C.S.,
Department of Otolaryngology,
Radcliffe Infirmary,
Woodstock Road,
Oxford OX2 6HE.