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# Leiomyosarcoma of the larynx: emergency laryngectomy

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**Abstract**
Leiomyosarcoma of the larynx is extremely rare. A patient with respiratory obstruction due to such a tumour is presented. This was originally misdiagnosed as hysteria. The treatment of choice was an emergency laryngectomy.

## Case report

A 56-year-old Caucasian man was sent to the casualty department by his GP in January 1987 who was concerned about his noisy breathing which had been getting worse during the previous week. He had asthma for the past ten years for which he was using a salbutamol inhaler and taking oral prednisolone. In 1983, he had an episode of aphonia for which no cause was found. A dignosis of functional aphonia was made then, and he recovered fully from this.

On examination (January 1987) he was very anxious and had inspiratory stridor. This was noted to be worse when he was being examined and better when he was left alone. On auscultation of his chest there were transmitted sounds from his upper respiratory tract. The rest of the clinical examination was normal. A diagnosis of hysteria was made (influenced by his past history of functional aphonia) and he was discharged back to the care of his GP.

Two days later, he was sent back to the hospital as his symptoms had become much worse. He was referred to the ENT department where he was found to be extremely anxious with both inspiratory and expiratory stridor. A general examination did not reveal anything new. Indirect laryngoscopy with a laryngeal mirror was very difficult due to the extreme apprehension. Examination with a fibreoptic nasendoscope revealed the ventricular bands to be closed during inspiration and expiration allowing no view of the cords. After a few breaths, the ventricular bands would snap open to reveal a granular lesion on the right vocal cord but no time was allowed to examine the glottis in detail. There were no enlarged glands in the neck.

He was taken to the theatre the same afternoon for a direct laryngoscopy under general anaesthesia. Intubation did not present any particular difficulties. He had a large tumour on the right vocal cord. A biopsy of the tumour was taken and it was debulked in an attempt to relieve his immediate distress. During recovery from the anaesthetic he was unable to maintain a satisfactory airway in spite of all efforts and he had to be reintubated. He stayed overnight in the ITU on a ventilator.

The result of the frozen section study, on the same day, was inconclusive. The result of the full histological examination (paraffin H&E) was available the next morning and it was diagnosed as a fibrosarcoma. A chest X-ray did not reveal any metastasis. A total laryngectomy was carried out the same day as an emergency procedure. The tumour was found to be much larger than was assessed during direct laryngoscopy and was found to be invading the subglottis (Fig. 1). There were no particular difficulties during the operation.

He did well post-operatively except for a small fistula which healed by conservative measures. On discharge he had good oesophageal speech and both he and his family were very pleased and appreciative of the outcome. He was last seen in May 1990 when he was doing very well with no signs of any recurrence and excellent oesophageal speech.

## Histopathology

Microscopy showed the tumour composed mainly of spindle-

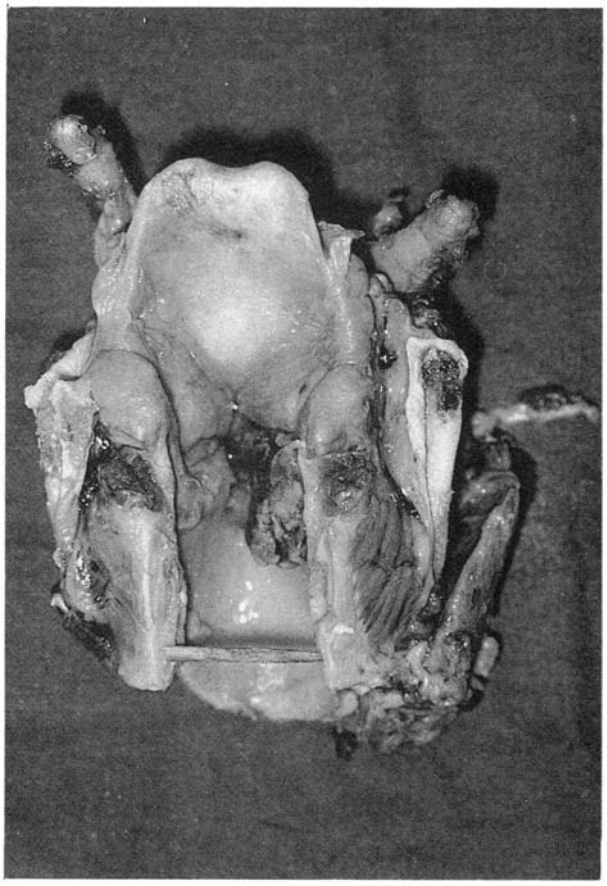

Fig. 1
Operative specimen showing tumour involving right cord and extending into the subglottis.

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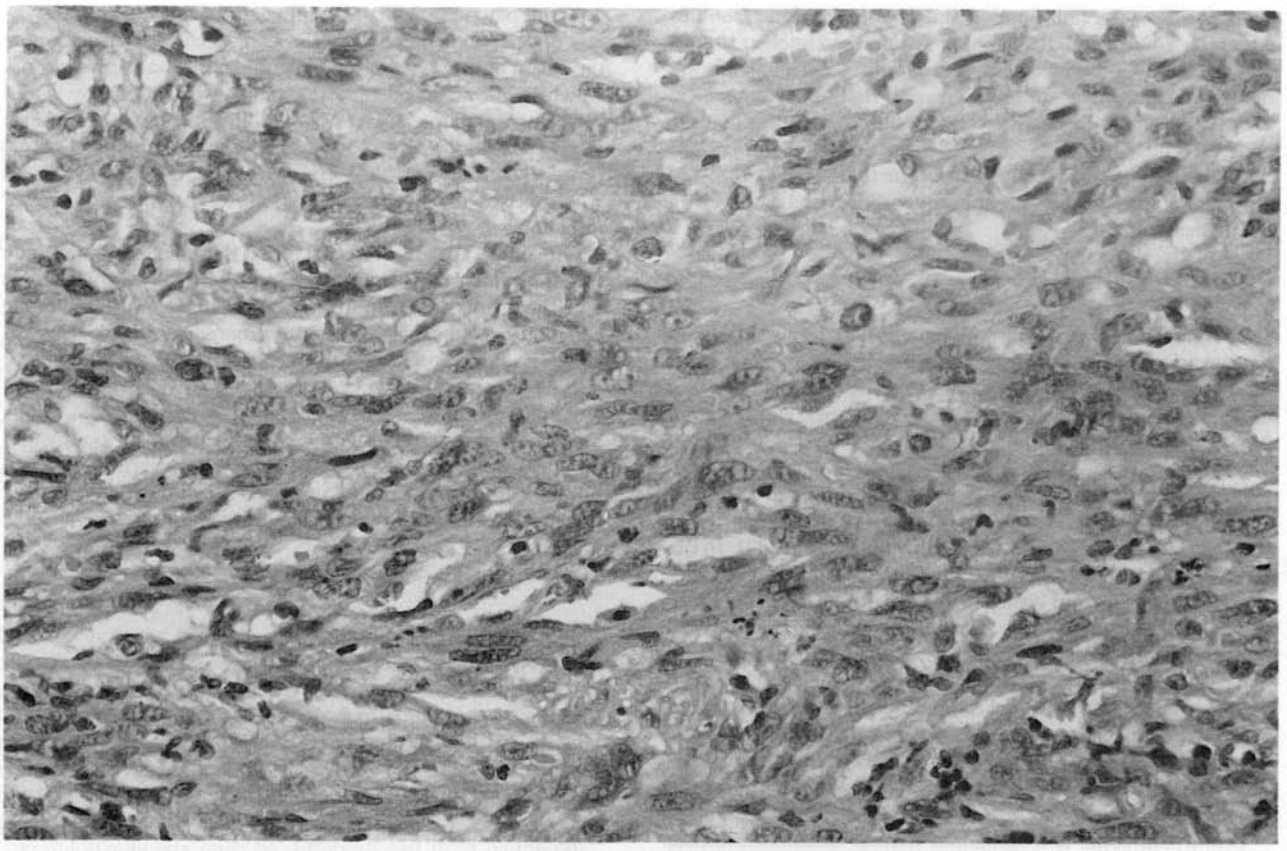

FIG. 2

The tumour is made up of mainly spindle-shaped cells with blunt-ended 'cigar-shaped' nuclei. The cells form thick bundles streaming across the tissue. Haematoxylin-eosin ×400.

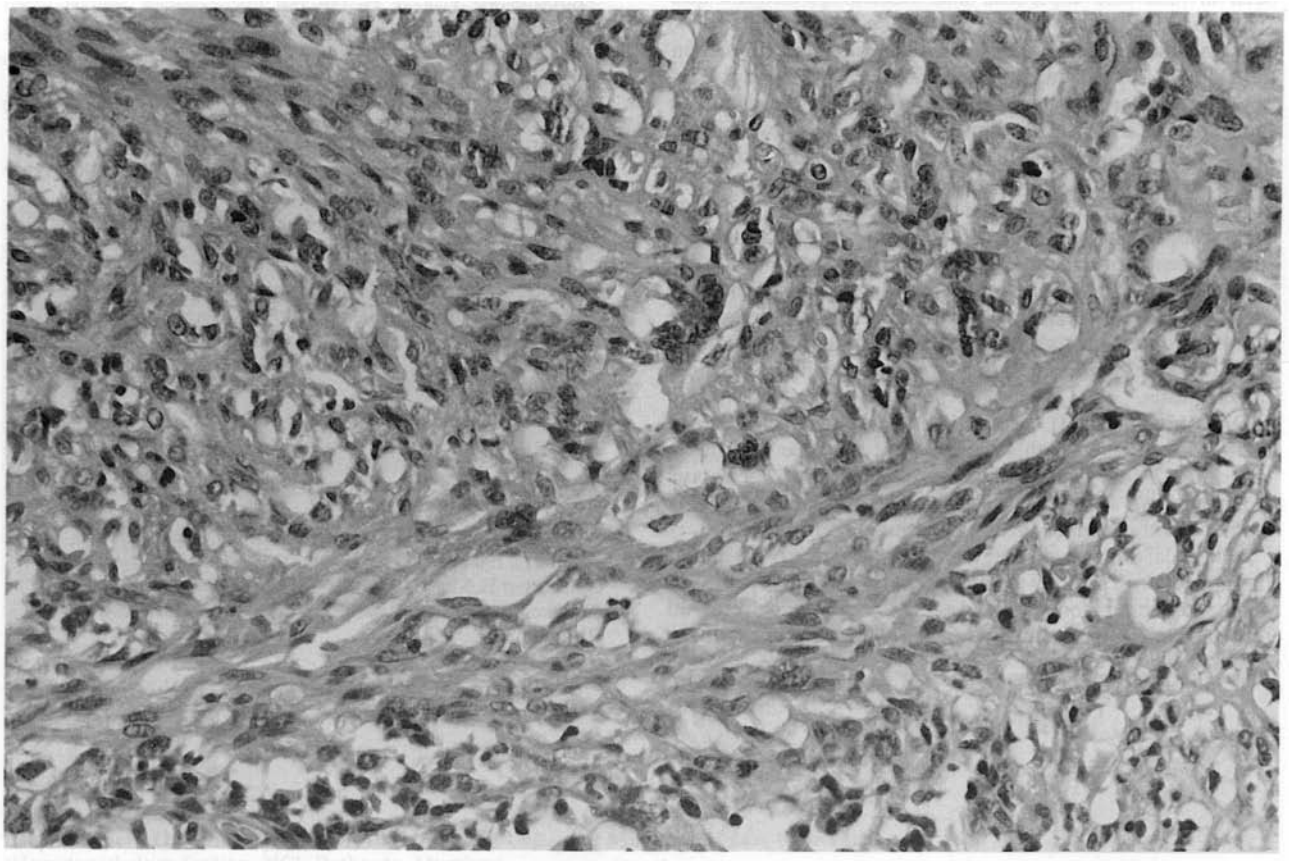

FIG. 3

To show several multinucleated cells within the mainly spindle-shaped cells of a leiomyosarcoma of the larnyx. As Fig. 1. Haematoxylin-eosin ×400.

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shaped cells with blunt-ended nuclei forming long bundles streaming through the field. There was marked nuclear irregularity of size and shape and there were occasional mitotic figures and multinucleated cells present. Immunocytochemical tests with vimentin were negative but many of the cells expressed desmin.

A herring-bone pattern in parts of the tumour has suggested a fibrosarcoma but this was subsequently revised to that of a leiomyosarcoma.

## Discussion

Leiomyosarcomas are rare malignant neoplasms. The incidence varies from 2.3 per cent to 5.3 per cent of malignant soft tissue tumours (Fields and Helwig, 1981). Eighty-five per cent of leiomyosarcomas develop in the extremities and approximately three per cent in the head and neck region (Goldberg *et al.*, 1988).

Leiomyosarcoma was first described in the head and neck region by Dobben in 1958. Twenty cases have been reported in the nose and paranasal sinuses (Kahn and Korol, 1989). In the larynx it is rarer still with a total of nine cases having been described in the world literature, the first by Frank (1941). In the English literature there have been only four reports and one communication by Kay to Wolfowitz (Wolfowitz and Schmaman, 1973).

Mindell *et al.* (1975) reviewed 29 reported cases of leiomyosarcoma in the head and neck and added two cases. They found the commonest site of presentation to be the scalp and superficial soft tissues; the next most were the hypopharynx, nose, tongue and trachea.

Leiomyosarcoma is a malignant tumour of smooth muscle origin. In the head and neck smooth muscle is sparse, being found mainly in the walls of blood vessels and the erector pilli musculature of the skin. Leiomyosarcomas have accordingly been divided into superficial and vascular (Wolfowitz and Schmaman, 1973).

Histological diagnosis of leiomyosarcomas has always been difficult in spite of all the progress made in the differential diagnosis of mesodermal tumours. The differential diagnosis from fibrosarcoma is especially difficult (Kleinsasser and Glanz, 1979). Leiomyosarcomas are characterized by proliferations of spindle cells with elongated cytoplasmic extensions arranged in bundles (Dobben, 1958). Nuclei tend to be rounded at the ends (cigar-shaped), a feature that allows some differentiation from fibroblasts. Ultra-structural examination for myofibrils, pinocytotic vesicles and desmosomes may be necessary for differentiation from spindle cell sarcoma, fibrosarcoma and neurofibrosarcoma (Kahn and Korol, 1989). In this case immunocytochemical tests were positive for desmin which confirmed the diagnosis.

Only three of the 31 cases of leiomyosarcomas of the head and neck presented with a concomitant palpable neck node suggesting a low incidence of early lymph node metastasis (Mindell *et al.*, 1975). The usual route of metastatic spread is via the blood stream to the lungs (Stout and Hill, 1958).

Wide local excision appears to be the most effective means of therapy for both primary and recurrent leiomyosarcoma of the head and neck region, radical neck dissection being reserved for manifest regional node metastasis (Mindell *et al.*, 1975).

**Key words: Laryngeal neoplasms; Leiomyosarcoma**

In this patient inability to extubate following the direct laryngoscopy persuaded us to choose an emergency laryngectomy as the best course. At the time the diagnosis was fibrosarcoma which is known to be radioresistant. A tracheostomy would have decreased the chances of obtaining a complete clearance due to the very high reported incidence of peristomal recurrence (Keim *et al.*, 1965; Baluyot *et al.*, 1971). Also there would be the technical difficulties along with the problems of infection.

One of the authors (A.L.P.) found that patients undergoing laryngectomy as an emergency were generally very satisfied with the results and were strongly motivated towards obtaining clearer oesophageal speech. Emergency laryngectomy, although a decision not to be taken lightly, has a definite place in the treatment of laryngeal tumours.

An important lesson to be learnt from this case that during the course of investigation of a patient with a previous history of functional symptoms, it is important to be aware of the possibility of serious new pathology. Such a history should neither obviate the need for thorough investigation nor overshadow judgement.

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