The Journal of Laryngology and Otology
May 1992, Vol. **106,** pp. 469-471

View from Beneath: Pathology in Focus
Retro-pharyngeal liposarcoma

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Abstract
We present a case of extensive recurrence of a retro-pharyngeal liposarcoma following surgical removal 18 years previously. The surgery and pathology are discussed, and management strategies of head and neck liposarcomas are reviewed.

Introduction

Liposarcoma, next to malignant fibrous histiocytoma, is the most common soft tissue sarcoma of adult life; its incidence in the head and neck region remains exceedingly rare. In a 50-year review of the literature, only 40 primary liposarcomas of the head and neck region were found (Batsakis, 1979).

Case report

A 69-year-old housewife presented with dysphagia and extensive swelling of her neck bilaterally, which she first noticed two years earlier and which had rapidly expanded over the past 12 weeks (Fig. 1). She did not complain of any hoarseness or dysphonia.

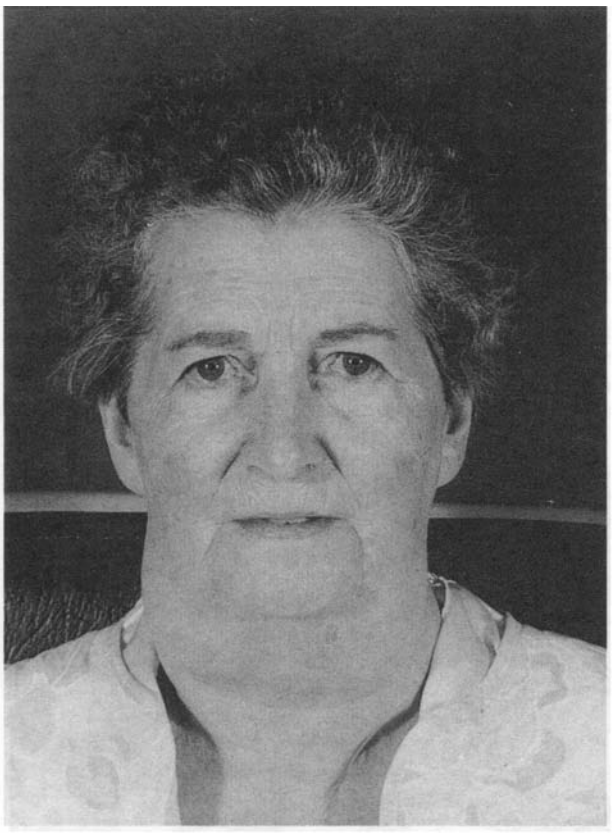

Fig 1

Eighteen years previously she developed a similar swelling of her neck and 335 g of lipomatous tissue was removed from her retro-pharyngeal space. The tissue was described as 'lipoma with bizarre pleomorphic features'. She was otherwise healthy with a previous history of pleurisy and a left hip replacement. She smoked 20 cigarettes a day and drank 20 units of alcohol per week.

On examination, she had a soft, non-tender, fixed and very diffuse extensive swelling of her neck bilaterally. Fibreoptic laryngoscopy gave the impression of a considerable retro-pharyngeal mass with a mid-line nodule of tissue projecting into the hypopharynx just above the arytenoids. Both vocal cords were seen to move normally. There was no pooling of saliva in the pyriform fossae.

CT scanning (Fig. 2) reported extensive fatty tissue of higher than normal density in the left side of the neck extending deep into the retro-pharyngeal area causing marked distortion of the larynx and pharynx which were pushed anteriorly, and extending inferiorly into the upper left mediastinum. The rapidly expanding right sided component of the lesion may have been caused by a recent haemorrhage. Chest X-ray and liver function test were normal.

A U-shaped 'apron' neck incision was made and a flap elevated with platysma and veins. A massive lobulated lipomatous tumour was found arising in the retro-pharyngeal space and extending into both sides of the neck, displacing the carotid sheaths laterally (Fig. 3). The right sided component of the tumour was exposed and dissected out, mainly extracapsular,

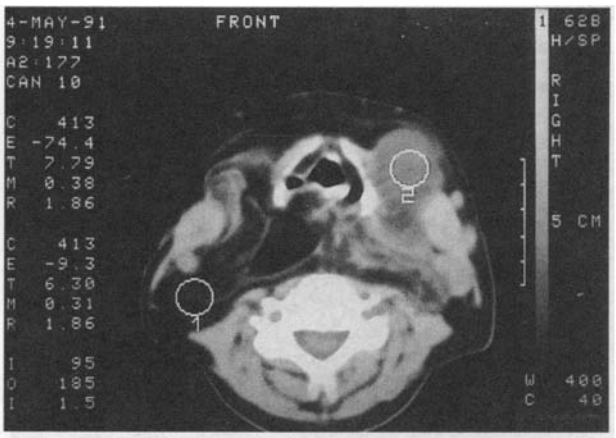

Fig 2

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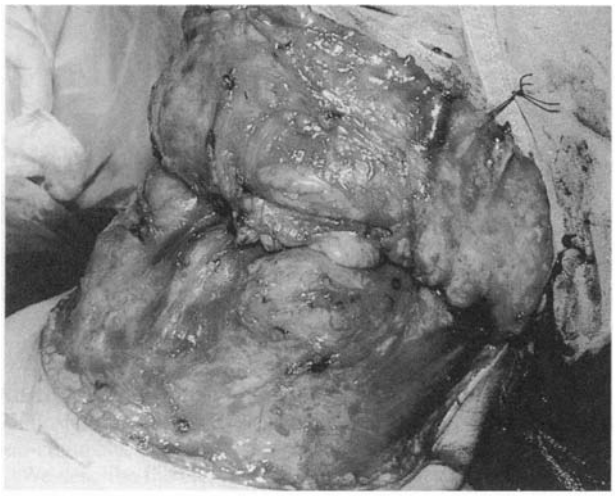

FIG 3

and removed in two large pieces. The retropharyngeal component was exposed by blunt dissection and removed. The left sided component of the lesion was much larger, extended into the upper mediastinum and was closely adherent to the pharynx and upper oesophagus. It was removed from above downwards, including the left submandibular gland. The bulk of the tumour was removed allowing access to the para-pharyngeal and upper mediastinal component, great care being taken to preserve the left recurrent laryngeal nerve. The thoracic duct was ligated and a tracheostomy performed.

The patient was noted to have a left vocal cord paralysis but otherwise made an uneventful recovery with no pneumothorax, chylous fistula, or damage to the brachial plexus. She was decannulated on the tenth post-operative day and discharged five days later.

At review one month post-surgery, she was still hoarse and described a choking episode which was relieved by the Heimlich manoeuvre. On examination, the incision and tracheostomy site were well healed and there were no obvious masses in the neck. Fibreoptic examination of the pharynx and larynx showed the left vocal cord to be still paralyzed. The pharynx and the upper third of the oesophagus appeared to be widely patent and peristalsis was visible.

She was assessed for post-operative radiotherapy, however the radiotherapist expressed reservations about the total treatment volume and possible encroachment on the spinal cord; as a result no treatment was given.

Pathology

A total of 396 g of fatty tissue was received as multiple fragments. Microscopically this consisted predominantly of apparently normal adipocytes (Fig. 4) with focal areas of increased cellularity. Within these foci, lipoblasts were present, each with an enlarged, hyperchromatic, scalloped nucleus and multiple cytoplasmic vacuoles of variable size (Fig. 5). Background vascularity was indistinct, but more apparent in the hypercellular foci. Mitotic figures and necrosis were not seen. A thin capsule of fibrovascular tissue delineated the edges of the tumour lobules. The appearances were regarded as those of a well differentiated liposarcoma.

Discussion

Liposarcomas show 'fat differentiation' and may arise *de novo* either from lipoblasts or totipotential mesenchyme within or adjacent to fascial and intramuscular areas, or rarely from subcutaneous fibroadipose tissue. However, it is now recognized that they do not arise from sarcomatous degeneration of a lipoma (Dahl *et al.*, 1982). Several histological classifications exist, the most widely used being the American Forces Institute of Pathology classification (Batsakis, 1979; Wenig *et al.*, 1990), which divides liposarcomas into four categories: (1) myxoid, (2) round cell, (3) well differentiated, (4) pleomorphic. The majority of head and neck liposarcomas are of the myxoid type followed distantly by the round cell variety.

Liposarcomas usually present as an inconspicuous painless mass, but the clinical course may vary from indolent growth to rapid aggressive local invasion. Distant metastases from the head and neck occur most frequently to the lungs, but appear to be relatively rare; 10 cases of laryngeal and hypopharyngeal liposarcomas reported by Wenig *et al.* (1990), displayed no distant metastasis with up to 40 years follow-up.

The tumour rarely occurs before the age of thirty, with median presentation in the sixth decade, and shows a slight male predilection.

CT scanning is recommended as the cornerstone in initial radiographic evaluation of lipomatous soft tissue masses. Magnetic resonance imaging features seem not to be specific (Wolfe *et al.*, 1989) and cannot differentiate benign from malignant tissue. However, MRI appears to be useful for pre-operative staging and follow-up studies (London *et al.*, 1989).

The treatment of choice remains wide surgical excision and post-operative radiotherapy. Evidence has shown an improved 5 and 10 year survival rate for combined versus single treatment regimes (Spittle *et al.*, 1989). Post-operative radiotherapy appears particularly useful for the myxoid variety.

The mis-leading pseudoencapsulation may tempt the surgeon to 'shell the tumour out', but satellite nodules around the main mass are common accounting for many local recurrences thus wide excision of the tumour free margin is critical. This may mean disruption of important structures nearby.

Chemotherapy is yet to be established as an effective adjuvant

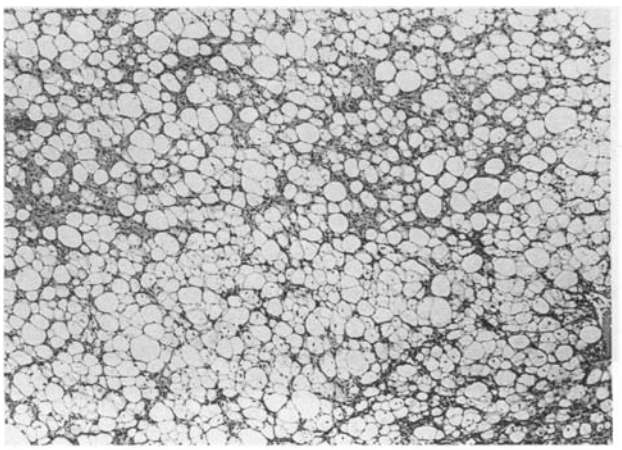

FIG 4

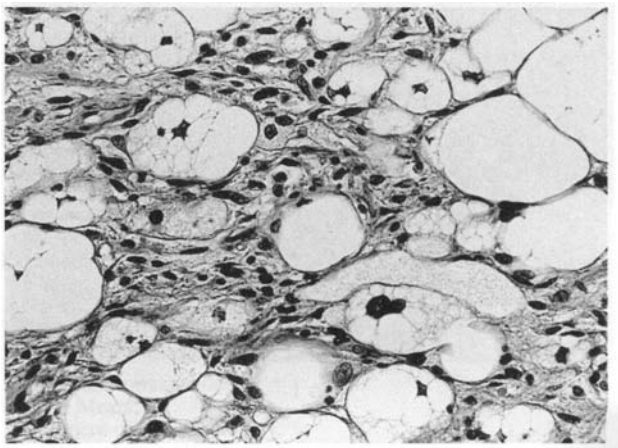

FIG 5

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therapy, but may be important for treating occult metastases (Ubayama, 1987).

More than 50 per cent of liposarcomas recur locally regardless of histological subtype. However, the 30 per cent of patients who may develop distant metastases are almost exclusively those with pleomorphic or round cell varieties. Rapid enlargement and tumour size of 12 cm or greater also show increased propensity to metastasize. Five year survival is greater than 70 per cent for myxoid and well differentiated types, but only 20 per cent for pleomorphic and round cell variants.

Conclusion

Liposarcoma of the head and neck region is a rare but important tumour. The necessity for wide surgical excision, postoperative radiotherapy and regular follow-up is strongly emphasized.

Acknowledgement

We would like to thank Miss D. McGrath for typing the manuscript.

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Key words: Head and neck tumours; Liposarcoma

https://doi.org/10.1017/S0022215100119875 Published online by Cambridge University Press