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**CLINICAL RECORD**

# Well-differentiated liposarcoma of the epiglottis

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## Abstract

*Objective*: Liposarcomas of the larynx and its sub-sites are rare. Within this group of tumours, well-differentiated liposarcomas of the epiglottis have been only sporadically reported. In view of its infrequent presentation, difficulty still exists over the diagnosis of this pathological entity, together with uncertainty over its exact accepted management.

*Method*: We present the case of a well-differentiated liposarcoma of the epiglottis, and we review the associated English-language literature.

*Results*: The reported patient underwent multiple attempts at surgical treatment prior to establishment of a firm diagnosis. In light of the diagnosis and other mitigating factors, a conservative approach was adopted.

*Conclusion*: A high index of clinical suspicion and detailed histological analysis are required when encountering a recurrent soft tissue lesion of the larynx. In the presented case, a multidisciplinary approach and conservative management plan were adopted, based on a holistic management approach and a review of the published literature.

**Key words:** Epiglottis; Laryngeal Neoplasms; Liposarcoma

## Introduction

Malignant soft tissue tumours are infrequent in the head and neck region. We present the case of a rare, well-differentiated liposarcoma in the epiglottic sub-site of the larynx.

## Case report

An 80-year-old, Caucasian woman presented with a 3-month history of globus and hoarseness of her voice. Her past medical history consisted of well-controlled hypertension. She was a non-smoker and an occasional drinker.

This patient was well known to the ENT department having initially presented 20 years prior with similar symptoms. An epiglottic lesion had been identified and excised under general anaesthetic. Over the next 20-year period, she had several recurrences and underwent a total of 6 separate excisions. The histological diagnosis had differed on each occasion as follows: benign epiglottic neuroma, epiglottic epidermal cyst, benign schwannoma, fibroepithelial polyp, epiglottic neurofibroma and pedunculated papilliferum.

### *Management*

Fibre-optic examination revealed a bulky tongue base and a lesion of the epiglottis. Panendoscopy revealed a bi-lobed, firm, pale mass in the pre-epiglottic space, adherent to the lingual surface of the epiglottis and measuring $30 \times 15 \times 10$ mm (Figure 1). Microlaryngoscopy and excision biopsy of the lesion were performed, and the excised tissue was sent for histopathological analysis. The procedure significantly improved the patient's symptomatology.

The patient was extensively discussed at the local head and neck multidisciplinary team meeting. The pathologists involved in the case had great difficulty in arriving at a conclusive diagnosis. In view of the previous lack of a consistent diagnosis and recurrence of the lesion, the sample was sent for a second opinion and further immunohistochemical analysis. Following conclusive diagnosis, we opted to manage the patient conservatively, taking into consideration the patient's age, the time frame of disease recurrence, and the likely co-morbidities associated with a more extensive procedure. The patient was followed up with close out-patient observation, including repeated flexible nasendoscopy.

### *Pathology*

Histologically, the mass was lined by intact benign squamous epithelium, and in several areas the underlying stroma contained a proliferation which was partly spindle-celled and partly adipocytic (Figure 2). This showed a variety of different appearances, but in most areas consisted of relatively bland spindle cells with delicate nuclei, set within a collagenous stroma and intermixed with fat cells including multi-vacuolated, non-atypical lipoblastic forms (Figure 3). However, in one area the proliferation was largely adipocytic and contained scattered, atypical lipocytes and lipoblasts (Figure 4).

Taking into account the 'deep' site of the lesion, its proven tendency to recur locally, and its infiltrative nature, the diagnosis was considered to be a lipomatous tumour, much of which showed the morphology of the so-called 'spindle cell liposarcoma' variant.

No evidence of de-differentiation (i.e. abrupt transition to a high-grade, non-lipogenic sarcoma) was identified, although this was borne in mind as a potential future risk associated with the tumour, as was the possibility of further local recurrence.

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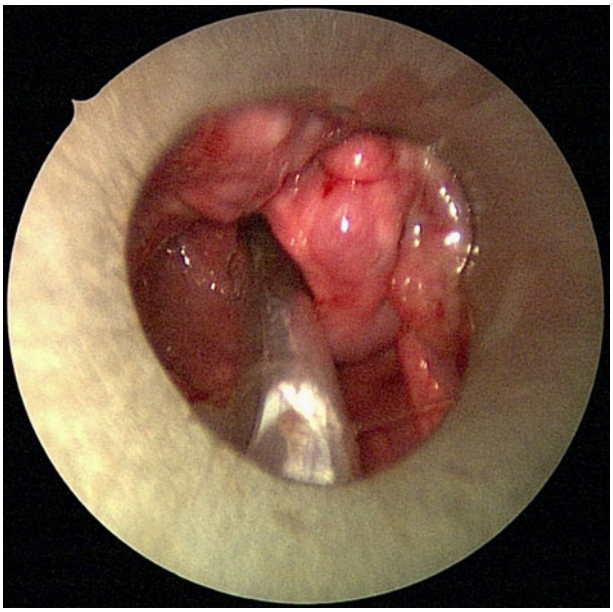

FIG. 1
Microlaryngoscopic view of the lesion.

## Discussion

Liposarcomas are known to be the most common soft tissue tumours in the body. However, only 3–6 per cent of all liposarcomas occur in the head and neck region. Of this small proportion, only a few cases of liposarcoma of the larynx have been reported in the English-language literature, with the majority being histologically classified as well-differentiated liposarcomas.[1] A review of the literature showed that previously reported cases occurred primarily in men and were confined to the supraglottic region.[1] Well-differentiated liposarcomas are difficult to diagnose since they can be histologically confused with simple lipomas.[2] Identifying the subtle changes associated with liposarcoma requires a high index of clinical suspicion by the histopathologist, and entails an extensive search of the specimen. In fact, morphologically well-differentiated liposarcomas occurring in more superficial planes are referred to as atypical lipomas.[2,3] The term well-differentiated liposarcoma is preferred in the larynx due to the tendency of the lesion to recur.[2,3] Certainly, one common feature of all previously reported cases is local recurrence despite complete surgical excision, as seen in our case. Distant metastasis has been reported, but there is debate as to whether this was due to the multi-centric nature of the disease or due to true metastasis.[4]

The evidence for the role of imaging in diagnosis is unclear. The most commonly used imaging modality described in previous case reports is computed tomography.[3,5] Other reports describe the use of thyroid isotope scanning,[6] antero-posterior laryngography[4] and barium swallow.[5] However, these investigations only show the presence of a lipomatous mass in the larynx, with no specific defining features.[3] In cases with a clear lesion on outpatient examination, the value of delaying diagnostic laryngoscopy and biopsy should be carefully weighed against the utility of imaging which may not add any useful clinical information.

Evidence from case series and individual case report reviews suggests that surgery is the treatment of choice.[1]

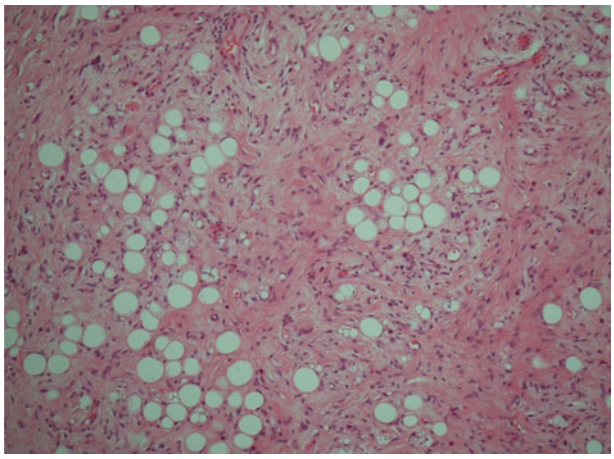

FIG. 3
Photomicrograph showing bland-looking spindle cells with delicate nuclei, set in a collagenous stroma, and intermixed with fat cells including multi-vacuolated, non-atypical lipoblastic forms. (H&E; ×100)

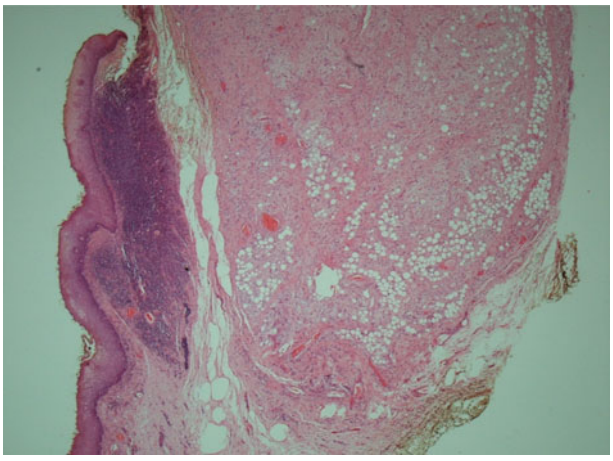

FIG. 2
Photomicrograph showing a circumscribed lesion consisting of spindle cell proliferation intermixed with fat cells. (H&E; ×40)

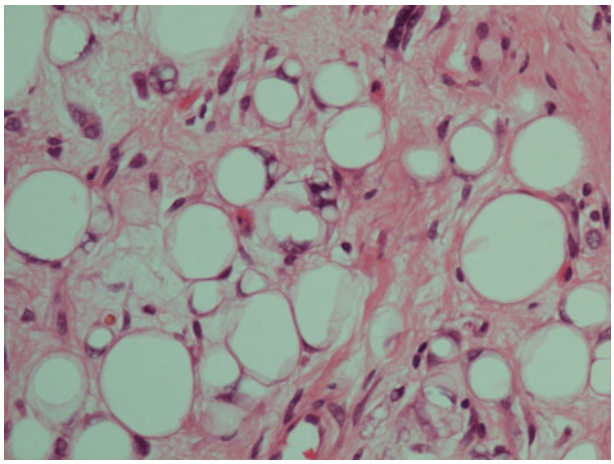

FIG. 4
Photomicrograph showing predomination of atypical lipocytes and lipoblasts; lipoblasts are particularly evident in the centre of the microscopic field. (H&E; ×400)

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The optimum extent of surgery remains unclear, since a conservative approach leads to high recurrence rates. Modalities of surgical treatment range from laryngectomy[6] to wide local excision. The extent of the surgery is also dictated by the size of the original mass.[3] In addition, it is difficult to assess the adequacy of complete resection, due to the macroscopic appearance of the tumour as well as the necessarily piecemeal nature of tissue removal.[1] However, more extensive surgery is associated with higher morbidity rates. The rates of metastatic disease and tumour-related mortality are low.[1,4] Morbidity rates, however, are high due to the tendency of the tumour to recur, requiring repeated excision.[1,5] Elective surgery to the neck has not been advocated by any author.[4] In view of this, local excision seems to be the favoured approach.

- **The diagnosis of liposarcoma should be considered for a recurrent laryngeal soft tissue lesion**
- **Well-differentiated liposarcomas are better known as atypical lipomas in the more superficial planes**
- **In the larynx, they tend to recur**
- **Wide local excision is the usual treatment**
- **In the presented case, simple debridement was successful**

Post-surgical radiotherapy has been advocated in cases of liposarcoma of the limbs and trunk.[9] In the head and neck region, and particularly the larynx, the role of radiotherapy remains unclear, and this therapy carries a high risk of morbidity. A 1978 study by Kindblom *et al*. did not report any benefit of radiotherapy.[7] This contradicted Krausen and colleagues' earlier statement that wide local excision and postoperative radiotherapy was the treatment of choice.[4] It has subsequently been shown that adequate surgical excision of the tumour tends to be curative.[8] Hence, there is currently no justification for the use of radiotherapy for laryngeal liposarcoma.

## Conclusion

Liposarcoma should be considered within the differential diagnosis of a recurrent soft tissue lesion of the larynx. Well-differentiated liposarcomas are better known as atypical lipomas in the more superficial planes. In the larynx, they have a tendency to recur.

Wide local excision of the tumour is the usual treatment of choice. In the presented case, simple debridement of the lesion was successful, with minimal morbidity.

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