

Pathology in Focus

Well-differentiated liposarcoma of the epiglottis

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Abstract

Liposarcomas of the larynx are very rare. A review of the English literature revealed only 28 published reports of tumours in this anatomical location. Diagnosis requires a high index of suspicion and careful histologic analysis. We present a case of a well-differentiated liposarcoma of the epiglottis, the tenth reported case at this laryngeal subsite. Initial biopsy specimens showed histological characteristics of a liposarcoma, which facilitated provision of optimal surgical treatment after careful analysis of published literature.

Key words: Epiglottitis; Laryngeal Neoplasms; Liposarcoma

Case report

A 51-year-old male presented to the emergency room with symptoms of increased difficulty in breathing and a change in voice. Additionally, he had experienced progressive dysphagia for approximately 4 months and positional dyspnoea, which was worse in the supine position. He was a heavy cigarette smoker, and had hypertension and adult onset diabetes mellitus. Flexible fiberoptic laryngoscopic examination showed a large yellowish-tan mass that appeared to arise from the base of tongue and obstructed most of the oropharyngeal airway. The remainder of the head and neck examination was essentially normal. An emergency tracheostomy was performed along with a direct laryngoscopy and biopsy. The mass obscured most of the larynx, was not friable, and did not bleed on biopsy. A post-operative computed tomography (CT) scan showed a mass measuring $5 \times 5 \times 3.5$ cm arising from the epiglottis with a tissue attenuation coefficient similar to that of lipomatous tissue (Figure 1). The initial biopsy specimen showed histological features suspicious of a liposarcoma. The patient subsequently underwent an anterior pharyngotomy with wide excision of the tumour. The pharynx was entered through the vallecula after freeing the suprahyoid musculature from the hyoid bone. Operative findings showed the sessile tumour to arise from the epiglottis. Microscopically, the tumour infiltrated the surrounding soft tissue in some foci and was well delineated in the remaining areas. It was composed of adipose tissue separated by fibrous septa of variable thickness (Figure 2a). The fibrous septa contained numerous spindle cells, many of which were atypical and hyperchromatic. The adipose tissue was mostly mature and contained foci of myxoid changes. The adipose tissue also contained atypical spindle cells and rare lipoblasts (Figure 2b). Necrosis and mitotic figures were not present. These findings confirmed the diagnosis of a well-differentiated liposarcoma. The surgical margins were free of tumour. The patient was decannulated and discharged home three



FIG. 1

Axial CT scan showing a mass with a tissue attenuation coefficient similar to that of lipomatous tissue filling the oropharynx.

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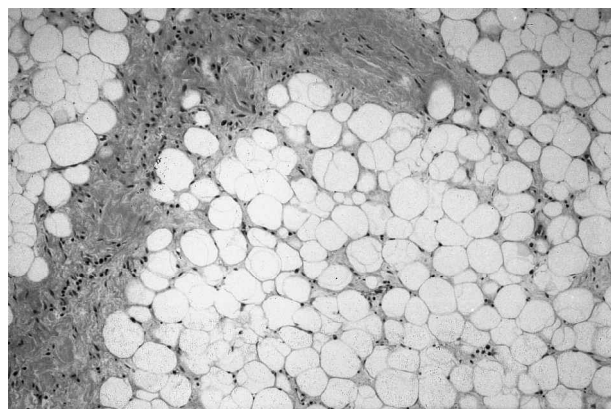


FIG. 2a

Fibrous septa separating adipose tissue. (H&E; $\times 100$).

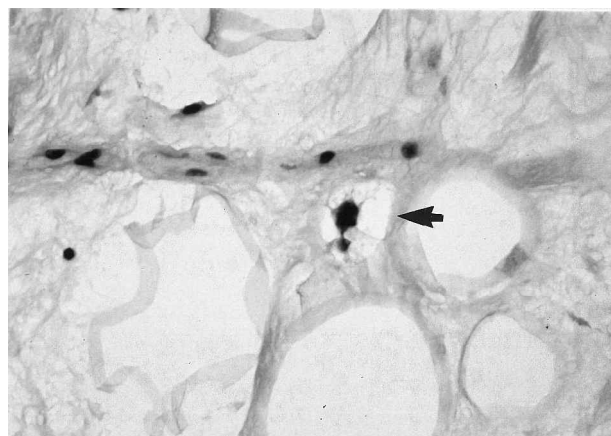


FIG. 2b

Adipose tissue containing lipoblast (arrow). (H&E; $\times 400$).

days after surgery. He remains free of disease 18 months after the procedure.

Discussion

Liposarcoma is the second most common soft-tissue sarcoma found in adults. Liposarcomas occur in the head and neck with a frequency of three to six per cent.¹ The most common head and neck site is the soft tissue of the neck. Liposarcomas of the larynx are extremely uncommon. Review of the English literature shows only 28 cases of liposarcoma of the larynx reported.^{2,3} We are reporting the 29th case of this laryngeal tumour.

The majority of patients with liposarcoma of the larynx are male, with only three cases being reported in females. Patients' age ranged from 28 to 83 with a median age of 55. Smoking has been suggested as an environmental factor in the development of this tumour;⁴ no other factors or basis for male predilection have been identified. A review of 27 cases revealed the most common presenting symptoms in order of frequency to be dysphonia, dysphagia, and respiratory symptoms.² The supraglottis has been identified as the most common site of tumour presentation, with a predilection for the epiglottis and aryepiglottic folds. To date there has been no report of this tumour arising in the subglottic region. Our case can be considered a typical presentation of this tumour in that he was male, a heavy smoker, had symptoms of respiratory obstruction, and an epiglottic lesion.

Four histologic types of liposarcomas exist, of which the most common are the well-differentiated and myxoid tumors. The well-differentiated type is often confused with

a simple lipoma. In our patient, although the tumour appeared and felt like a lipoma on direct laryngoscopy and biopsy, histopathological examination revealed atypical lipocytes, raising the suspicion of a sarcomatous tumour. Well-differentiated liposarcomas are frequently not recognized until they recur, and the original diagnosis of lipoma is reviewed. This problem is attributed to the subtle changes of liposarcoma, i.e. pleomorphism, infiltrative growth, and lipoblasts that may not be recognized on biopsy material.⁴ It was further noted that identification of lipoblasts often requires an exhaustive search of the tumour specimen.⁴ The term atypical lipoma or atypical lipomatous tumour is currently used for tumours showing morphologic features of well-differentiated liposarcoma but occurring in a superficial location. For tumours occurring in a vital organ, such as the larynx, the term well-differentiated liposarcoma is preferred because inadequate excision and subsequent recurrence may result in increased morbidity and mortality.¹ The other two types of liposarcoma are the pleomorphic and round cell tumours, which have been reported to behave aggressively. Well-differentiated liposarcomas of the larynx tend to recur locally and have no reported metastasis. Other subtypes have been reported to metastasize, although infrequently.⁵⁻⁷

Surgical excision is the accepted modality of treatment for liposarcomas of the larynx, although the extent of resection remains controversial. Reported therapeutic procedures have ranged from simple endoscopic excision to supraglottic laryngectomy.⁸ As there have been no cases of cervical metastasis, surgical treatment of the neck is not advocated.^{2,4,8} A review of the literature suggests an approximately 95 per cent recurrence rate with simple endoscopic excision, which can be reduced to less than 10 per cent by performing a more extensive procedure.² We had the opportunity to review the literature prior to definite surgery and hence elected for an anterior pharyngotomy approach. Nevertheless, this particular tumour would not have been amenable to endoscopic excision due to its large size. A wide excision of the tumour including a part of the epiglottis gave us disease-free margins. The patient presented in this case report has been followed clinically and remains disease-free 18 months later. Long-term surveillance for recurrence in these patients is necessary. One recent review article found the average time from initial resection to first recurrence to be 3.9 years.² Post-operative adjunct therapy was not given to this patient. There is no justification for post-operative radiation therapy after wide surgical excision of the tumour.⁴ Re-excision with post-operative radiation therapy has been advocated for control of local recurrences.³ A high index of suspicion and knowledge of the clinicopathological features of this tumour allowed us to effectively treat this patient and minimize surgical risks.

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