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Journal of Experimental Pathology Case Report

Liposarcoma of Laryngeal Vallecula

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Abstract

Liposarcoma is a malignant mesenchymal tumor originating from adipose tissue. Soft tissue liposarcomas are quite common in the adult population, whereas a primary liposarcoma in the larynx is exceedingly rare. Liposarcomas of the larynx have a high risk of local recurrence but seldom metastasize. The prognosis of these tumors is better than that of non-laryngeal liposarcomas. We present a case of a sixty-year old male with laryngeal liposarcoma which was diagnosed as an epiglottic cyst clinically.

Keywords: Larynx, Liposarcoma, Head and neck

Introduction

Liposarcoma (LS) is the most common soft tissue sarcoma (20% of the tumors in adults). This neoplasm was first described by Virchow in 1857 and has been well documented thereafter. Liposarcoma is a common soft tissue malignant tumor and is often found in the lower extremities and retro peritoneum. Only about 5.6% of liposarcomas are found in the head and neck and most of these tumors arise from the soft tissues of the neck [1-3].

The origin of liposarcomas is controversial, these tumours are thought to arise de-novo rather than from malignant transformation of a pre-existing benign tumor. Most cases do not have any specific etiology. Although trauma is often implicated as an inciting agent, it is unclear whether it is a true etiologic factor. Genetic alterations are becoming increasingly recognized as causative factors and have been used to improve the accuracy of subtyping combined with morphology [4,5]. In all sites including outside the head and neck, liposarcoma is the most common tumor among sarcomas of the soft tissue and is categorized into four subgroups: atypical lipomatous tumor/well-differentiated liposarcoma (WDLS), dedifferentiated liposarcoma (DDLS), myxoid liposarcoma, and pleomorphic liposarcoma [6,7].

Due to the similarities in presentation of lipomas and liposarcomas, a clinical diagnosis is often difficult to make. Thus, initial management is the same for both these lesions and involves complete excision for diagnostic and therapeutic purposes.

In this article, we present a rare liposarcoma involving the laryngeal vallecula in a sixty year old male which was clinically diagnosed as a cyst.

Case Report

A sixty-year-old male presented to the outpatient department with a complaint of difficulty in swallowing. Intraoral examination was non-contributory. Direct laryngoscopic examination, showed a smooth surfaced polypoid mass lesion measuring approximately 8 cm in greatest dimension involving the left vallecula. A provisional diagnosis of a cyst arising from left vallecula was made. Considering the benign and innocuous nature of the lesion, an excision of the lesion was planned. No cervical lymph nodes were detected during the extra-oral examination. There was no significant medical history.

We received a Polypoid mucosa covered mass measuring approximately 8 cm in greatest dimension. Cut surface of the mass was lobulated yellowish and fatty. The histopathologic sections showed a mass composed of mainly adipocytes with variability in adipocyte size, focal myxoid and collagenous stroma and atypical stromal cells with scattered lipoblasts. The microscopic

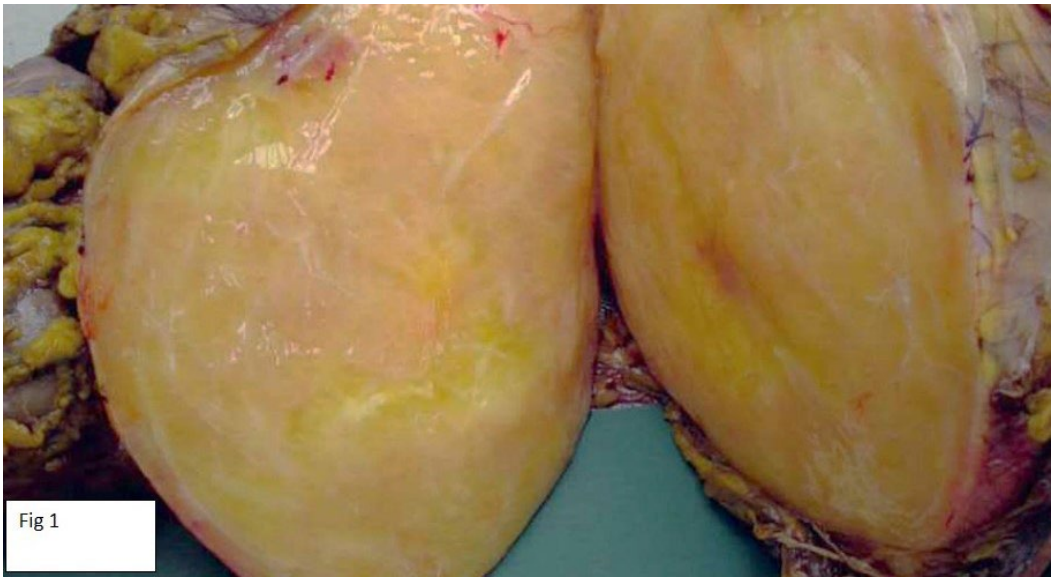

Figure 1: Gross picture of specimen.

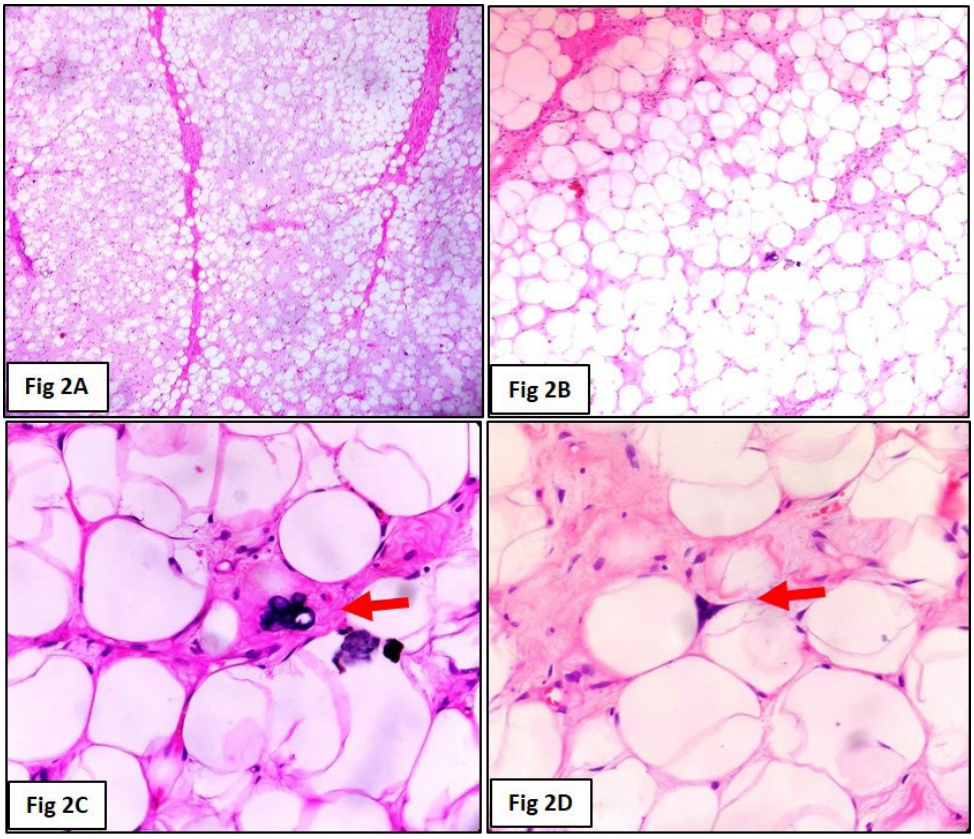

Figure 2: A) Photomicrograph showing composed of variably sized adipocytes and scattered lipoblasts. (HE- 100x). **B)** Photomicrograph showing adipocytes separated by fibrous strands. (HE-200x). **C)** Photomicrograph showing atypical stromal cells. (HE-400x). **D)** Photomicrograph showing pleomorphic stromal cells. (HE- 400x).

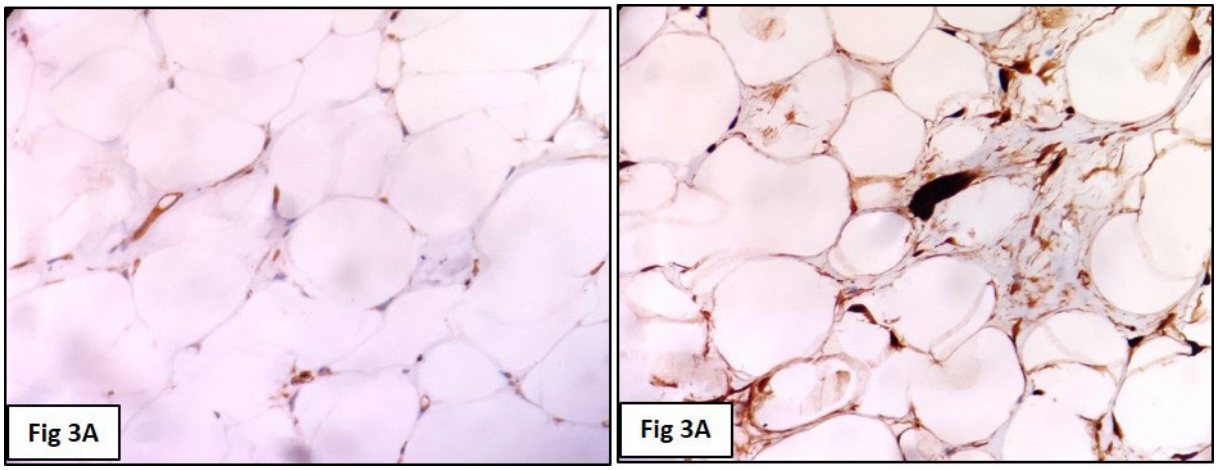

Figure 3: Immunohistochemistry images. **A)** Photomicrograph showing atypical spindle cells immunopositive for CDK-4. (400x). **B)** Photomicrograph showing atypical spindle cells immunopositive for MDM-2. (400x).

examination revealed few mitotic figures (0-10 per high-power fields). No necrosis was observed (Figures 2A-2D). Immunohistochemical analysis for MDM-2 and CDK-4 was performed and revealed nuclear staining in atypical spindle cells (Figures 3A and 3B). A final diagnosis based on the histological appearance and immunohistochemical profile of atypical lipomatous tumor/well-differentiated liposarcoma was rendered. All resected margins were clear and the distance from closest resected margin was 0.1 cm. Thereafter, the patient is on regular follow up for the last one year and is disease free till now.

Discussion

Liposarcoma is one of the most common types of soft tissue sarcoma. It is a malignant neoplasm recapitulating fat. It may recur locally but usually does not metastasize unless dedifferentiated [5,7,8].

Liposarcomas occur most frequently in the 5th and 6th decades of life, predominantly in men. Approximately 4% of liposarcomas occur in the neck; among these tumors, the most frequent sites of involvement are the neck (28%), larynx (20%), and pharynx (18%). The case being discussed had a demographic profile in accordance with the literature.

In the head and neck region, liposarcomas are typically low-grade and detected at an early stage due to symptoms and thus have a better prognosis and disease specific survival than liposarcomas found in other sites. Symptoms like dysphagia, hoarseness, choking, and dyspnea due to significant airway obstruction are produced by laryngeal, supraglottic and hypopharyngeal liposarcomas, while retropharyngeal liposarcomas cause obstructive sleep apnea [8]. Similar signs and symptoms were observed in the present case also.

According to the current WHO classification of soft tissue tumours, adipocytic tumours are classified as follows:

Intermediate (locally aggressive) –

Atypical lipomatous tumor/ Well-differentiated liposarcoma (WDLS)

Malignant –

Dedifferentiated liposarcoma

Myxoid/round cell liposarcoma

Pleomorphic liposarcoma

Mixed-type liposarcoma

Liposarcoma, not otherwise specified [6]

Lipoma-like WDLS were also termed as Atypical Lipomatous Tumour (ALT), a term which was introduced in 1974. Thus, in the 2002 WHO classification, it was recognized that atypical lipomatous tumor and well-differentiated liposarcoma are essentially synonymous entities. These lesions are classified as locally aggressive tumors with intermediate malignant potential. ALTs share similar histologic features with lipoma-like WDLS and typically occur in subfascial locations of the extremity and trunk (this term is not used for retroperitoneal lipomatous tumours) [4-10].

Myxoid liposarcoma is the most common subtype, associated with a 5-year survival rate of 77-95%. This high-level survival is likely because the tumor is slow-growing and tends not to metastasize; although local recurrence is common. A liposarcoma diagnosis is usually delayed because of the indolent nature of tumor, though symptoms differ according to the location and size, and patients often have no complaints in the early stage of disease [5-10].

The radiologic imaging may resemble lipomas in which more than 75% of the tissue volume consists of fat. There is extensive overlap between the characteristics of benign lipomatous tumors and those of well-differentiated liposarcomas, and non-invasive differentiation may be impossible. ALT/WDLS are composed of mature adipocytes with significant variation in cell size and focal nuclear atypia. They typically show scattered atypical stromal cells with hyperchromatic nuclei embedded within mature adipose tissue. This morphologic appearance is in keeping with a lipoma-like WDLS. Some important features that may help distinguish well-differentiated liposarcomas include the patient's age (>60 years), large lesion size (>10 cm), presence of enhancing thick septa (>2 mm thick), presence of non-adipose mass like areas, and a low proportion of fatty content (<25% of tumor volume) [4,5].

The differential diagnosis of a pharyngeal mass includes benign and malignant entities such as fibrovascular polyp, lipoma, nodular fasciitis, carcinoma, sarcoma, lymphoma, and melanoma. Discriminating liposarcoma from lipoma by histology alone can be difficult, as the cytologic atypia of a well-differentiated liposarcoma can be subtle. Indeed, the majority of hypopharyngeal/esophageal WDLS in the past have been considered reactive polyps rather than malignant neoplasms.

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Detecting MDM2 or CDK4 amplification by FISH is a sensitive and specific method for distinguishing liposarcoma from benign lipoma. MDM2 and CDK4 are located on chromosome 12q13-15, an area that is amplified in well-differentiated and dedifferentiated liposarcoma. Immunohistochemistry for MDM2 and CDK4, when strong and diffuse, can be used as a surrogate for FISH with reasonable diagnostic efficacy [13-15].

For both Liposarcomas of the whole body and Head and Neck, surgical resection is the standard treatment. Although the tumor appears well encapsulated, the fascia surrounding it is not a true enveloping layer so the surgeon could leave microscopic residual disease behind. For this reason, excision should be as wide and meticulous as possible. Regional lymph node dissection is usually not recommended because node metastases are rare. In the head and neck, the extent of tumoral excision is tempered by the close proximity of vital neurovascular structures, resection of which may result in severe morbidity [3].

Radiotherapy (RT) has an undefined role in the treatment of laryngeal liposarcoma and is a matter of debate. Some authors suggest that adjuvant RT would be useful in the following situations: close or positive margins, high tumor grades, and the myxoid variant. In fact, adjuvant RT is considered to decrease the recurrence rate after surgery. The role of chemotherapy has not gained much attention in the literature and is still in the experimental phase. In our case the patient did not receive post-operative RT [8].

Liposarcomas rarely develop in the head and neck region. The mainstay of treatment is surgical excision and the prognosis is largely determined by the histological grade and the clinical stage. Complete excision is usually sufficient to cure low-grade tumors (well differentiated and myxoid). However, high-grade tumors (pleomorphic and round cell) are more aggressive and have a worse outcome despite surgery and postoperative radiotherapy.

The prognosis of laryngeal liposarcomas appears to be more favourable compared to liposarcomas affecting other regions. This feature seems to be related to the low incidence of high-grade tumours and to the rarity of metastases [3-10].

Prognosis is chiefly dependent upon histologic grade and site. The well-differentiated and myxoid variants, the two most prevalent types, have the best outcomes while the de-differentiated types (arising from atypical lipomas or well-differentiated liposarcomas), pleomorphic, and round cell types do not fare as well. Liposarcomas of the head and neck tend to be well-differentiated (44.5%) or myxoid tumours (35.5%) [5,6].

Furthermore, pleomorphic and round cell liposarcomas are also much more likely to metastasize compared to well-differentiated or myxoid types. Pleomorphic subtypes are the highest grade and carry the worst prognosis amongst all the five subtypes. Golledge et al. found that neck, pharyngeal and oral liposarcomas had the poorest outcomes while those in the scalp, laryngeal area and face had a much better outcome. They also found that oral liposarcoma had the poorest outcome with a five

year survival of 50%. However, other authors have noted that the small size of oral liposarcomas resulted in better prognosis [4,9,10-15].

Conclusion

Liposarcomas are classified into three biological types encompassing five subtypes based on morphological features and cytogenetic aberrations. These five subtypes have different biology, growth rates, and patterns of behavior. Surgical resection is the mainstay of curative treatment. The present case provides an example that duration of symptoms and clinical appearance of the mass are not reliable indicators of benign or malignant potential in lipomatous tumors of the larynx. Because of this diagnostic ambiguity, appropriate management requires complete excision of the mass and histopathologic evaluation supplemented by Immunohistochemical studies.

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