Dendritic Fibromyxolipoma of the Larynx- A Case Report and Review of Literature

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ABSTRACT

Dendritic fibromyxolipoma (DFML) is an uncommon, benign soft tissue tumor. It is seldom reported in the hypopharynx. However, its endoscopic appearance may mimic an aggressive soft tissue neoplasm hence the importance of its recognition. We discuss a rare case of DFML of the pyriform fossa in a 66-year-old male who presented with progressive hoarseness. The histopathological diagnosis was made following conservative excision of the mass using carbon dioxide laser, preserving swallowing and voice.

Keywords: Dendritic Fibromyxolipoma; Lipoma; Larynx

INTRODUCTION

Lipoma constitutes the commonest benign soft tissue tumor occurring in adults. In most cases, it presents as a soft mass in the posterior neck or shoulder region, just underneath the skin **[1]**. This disease is predominant in men between the ages of 40 and 50 years. Many variants of benign lipomas have been described including angiolipoma, chondroid lipoma, myolipoma, and spindle cell lipoma **[1]**.

Dendritic fibromyxolipoma (DFML) is a rare and distinctive tumor that is considered by many as a variant of spindle cell lipoma. It is characterized by extensive myxoid change with the presence of stellate cells and dendritic processes **[2]**. Herein, we describe a case of DFML which occurred in the larynx and hypopharynx. These are unusual anatomical sites for DFML to occur. Furthermore, in this case where an elderly presents with progressive hoarseness, a high index of suspicion for a malignant tumor in the larynx and hypopharynx is the usual norm. In such cases, a biopsy procedure is considered mandatory to arrive at a histopathological diagnosis.

CASE REPORT

A 66-year-old man with hypertension presented with progressive hoarseness for 3 months. It was associated with reduced effort tolerance, noisy breathing, and shortness of breath (Medical Research Council breathlessness scale grade 3). Otherwise, no dysphagia, odynophagia, orthopnoea, or aspiration symptoms were present. The patient was working as a mechanic. He was a non-smoker and did not drink alcohol.

On examination, the patient was comfortable, not tachyponeic and no stridor or stertor heard. The overall dysphonia was grade 2 with the main component of roughness. Neck examination was normal with no scar or palpable lymphadenopathy. Intraoral examination was normal. Flexible naso-pharyngo-laryngoscopy noted the presence of a submucosal mass arising from the right pyriform fossa. The mass was covered by normal overlying mucosa, extended medially, and covered bilateral true cords. No pooling of saliva was noted and the sensations were intact.

Computed tomography (CT) scan showed a homogenous well-defined mass measuring 3.1 x 3.2 x 2.5 cm at the right pyriform fossa. The attenuation was suggestive of the fat component. No erosion of the thyroid or arytenoid cartilage was seen.

Endoscopic endolaryngeal excision of the tumour was performed. The patient was intubated using microlaryngeal tube (MLT) size 5 via video and bougie assistance. Direct laryngoscopy noted the presence of a soft, smooth-surfaced mass arising from the right pyriform fossa, extending to the right false cord, and partially obscuring the true cord (Figure 1). The mass was excised with carbon dioxide laser. The true cords, subglottic,

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Figure 1: Right pyriform fossa mass (yellow arrow) obscuring view of true cords

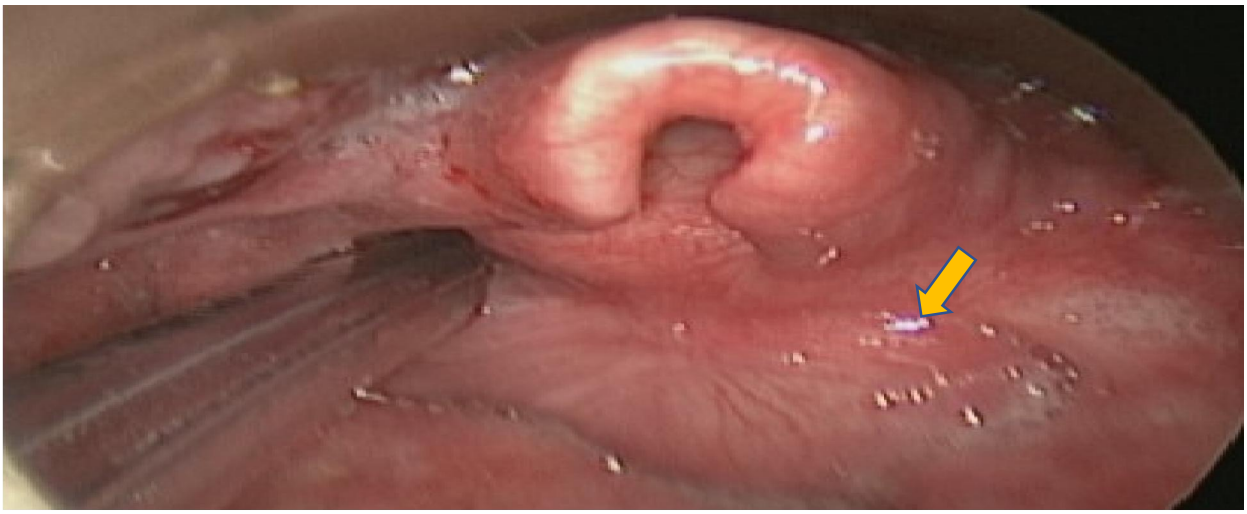


Figure 2: Endoscopic view of excision of mass

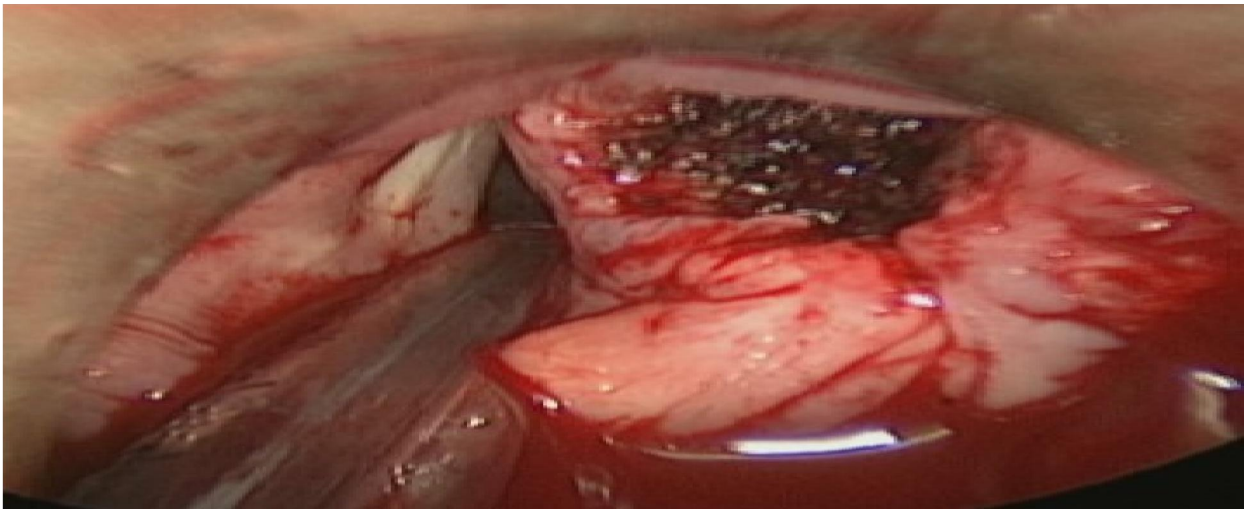


Figure 3: Histopathology showing spindle-shaped cells admixed with mature adipocytes

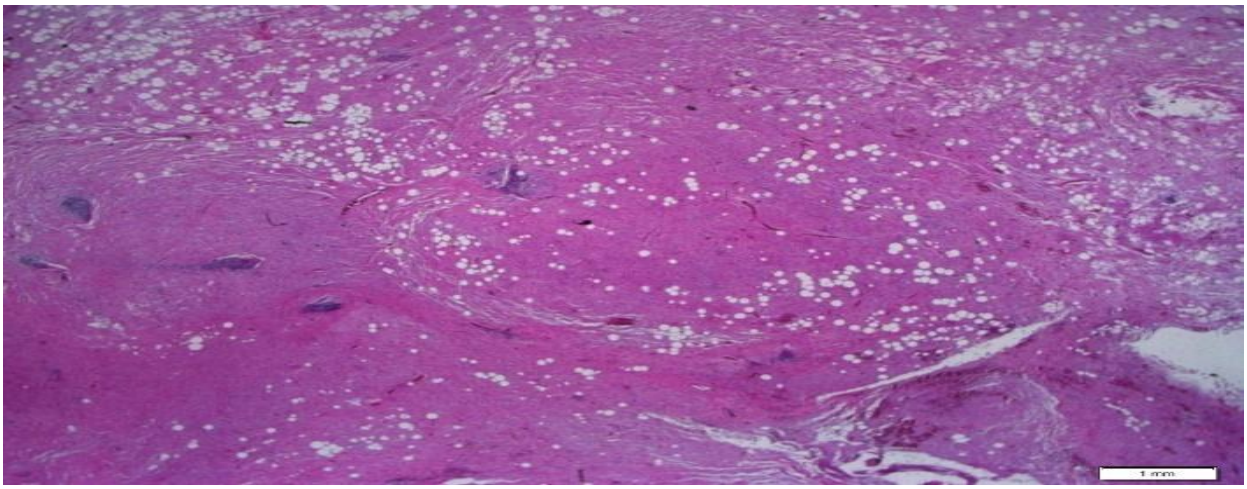
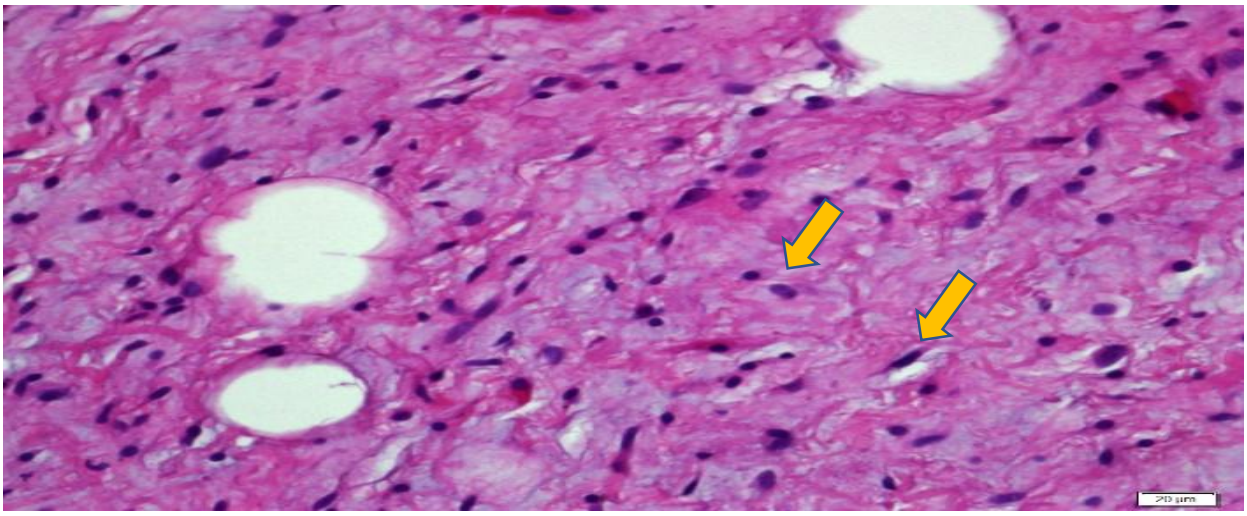


Figure 4: Histopathology slide with spindle cells identified (yellow arrows)



vallecula, and epiglottis were not involved (Figure 2).

On postoperative follow-up, the patient's voice was improved and there was no evidence of recurrence of the disease. Histopathological examination of the mass found a proliferation of spindle-shaped cells admixed with a variable amount of mature adipocytes surrounded by thick eosinophilic ropy collagen in areas (Figure 3). The spindle cells showed bland oval nuclei surrounded by a mixture of collagen and myxoid stroma (Figure 4). The features were consistent with DFML.

DISCUSSION

Lipomas of the larynx and hypopharynx are rare, with a site predilection at the false cords, aryepiglottic folds, and epiglottis **[2]**. They can present with variable symptoms of airway obstruction. In addition, the sensation of a lump in the throat, voice change, snoring, and excessive accumulation of salivary secretions may also be noted. Because lipomas are slow-growing, symptoms may span several months to years, with an insidious onset. On the contrary, our patient presented with 3 months history of hoarseness associated with reduced effort tolerance. The rapid progression of symptoms in this patient suggests the possibility of malignant disease. The endoscopic appearance of laryngeal lipomas is quite varied, ranging from a submucosal mass, as in this case, to a pedunculated, intraluminal projection **[3]**.

Diagnosis of head and neck lipoma starts with good clinical examination. Lipomas are non-painful, usually round, mobile masses with a characteristic soft, doughy feel on palpation. Although most superficial subcutaneous lipomas can be suspected with a high degree of accuracy by clinical examination alone, very large, deep-seated or infiltrating lipomas, as well as lipomas arising from unusual regions within the head and neck, require imaging for further assessment and diagnosis **[4]**. On CT images, lipomas and fibrolipomas appear as smooth or lobulated well-defined masses associated with moderate displacement of surrounding tissues **[5]**.

El-monem et al found both CT and magnetic resonance imaging (MRI) to be accurate diagnostic modalities for head and neck lipomas **[4]**. One weakness in the use of current diagnostic imaging techniques in the diagnosis of tumors of fatty tissue is that neither CT nor MRI can differentiate a lipoma from a liposarcoma **[6]**. The distinction can only be made with certainty by histopathological examination. Therefore, complete excision of lipoma is recommended to exclude a possible liposarcoma, especially in fast-growing lesions **[1]**.

DFML was initially described in 1998 by Suster et al **[7]**. DFMLs are characterized by histologic and immunohistochemical features that reminiscent of spindle cell lipoma and solitary fibrous tumor. However, its large size, prominent vascularity, and myxoid changes may be suggestive of a malignant neoplastic process. Therefore, it is usually misinterpreted as a low-grade sarcoma.

The reported age of onset ranges from 24 to 81 years, with a median age of 65 years. It has preponderance in males with a reported male to female ratio of 4:1. In this case, the age of presentation is similar to the median age reported

with a male predilection. DFML commonly presents with a subcutaneous mass at the muscular fascia of the shoulder, neck, and back. Other reported locations include the nasal tip, lip, forearm, and intramuscular location. To the best of the author's knowledge, there was only one other report by Al-Abdulsalam et al whereby DFML was found arising from the pyriform sinus **[8]**. Endoscopic laryngeal microsurgery surgery with debulking of the mass using carbon dioxide laser was performed.

Microscopically, DFML is composed of spindle cells with anastomosing blood vessels, ropy collagen, and prominent mast cells. The spindle cells have typical multiple dendritic cytoplasmic processes, owing to its name. It is best highlighted by immunohistochemistry for CD34, bcl-2, and vimentin **[7]**. CD99 positivity has been reported in a case **[9]**. Smooth muscle actin, muscle-specific actin (HHF35), desmin, S-100 protein, keratin, and EMA are negative **[7]**.

CONCLUSION

In conclusion, lipomas can present in a variety of different ways in the head and neck region, especially in the hypopharynx. CT and MRI scans help a specific pre-operative diagnosis in virtually all cases, thus enabling better treatment planning. Complete excision of lipoma is indicated to improve the patient's symptoms and exclude a malignant lesion.

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