Dendritic Fibromyxolipoma Of Larynx

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Abstract

Dendritic Fibromyxolipoma (DFML) is an uncommon, benign soft tissue tumour. 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 report a rare case of dendritic fibromyxolipoma of the pyriform fossa in a 66-year-old male who presented with hoarseness. The best approach in managing this case will be discussed.

Introduction

Lipomas represent the most common soft tissue tumours in adults. Spindle cell lipoma (SCL) is an uncommon variant characteristically presents as a mass in the posterior neck and shoulder region of men between the ages of 40 and 50. Dendritic fibromyxolipoma (DFML) is a rare and distinctive tumour that is considered by many as a variant of spindle cell lipoma. It is characterized by extensive myxoid change and the presence of stellate cells with dendritic processes.

Case report

A 66-year-old man with underlying hypertension presented with worsening hoarseness for the past 3 months. It was associated with reduced effort tolerance, noisy breathing and shortness of breath. There was no dysphagia, odynophagia, orthopnoea or aspiration symptoms. He was a non-smoker and non-alcohol consumer.

On examination, he was not tachypnoeic with no stridor or stertor. Overall dysphonia grading was grade 2 with main component of roughness. Neck and intralaryngeal examination was normal. Flexible nasolaryngopharyngoscopy noted 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 and sensation was intact. Computed tomography (CT) scan showed a homogenous well-defined mass measuring 3.1x3.2x2.5cm at the right pyriform fossa. The attenuation was suggestive of fat component. No erosion of the thyroid or arytenoid cartilage was seen. Direct laryngoscopy finding noted presence of a soft, smooth surfaced mass arising from the right pyriform fossa extending to the right false cords, partially obscuring the true cord (Figure 1). The mass was excised with carbon dioxide laser endoscopically. The true cords, subglottic, vallecula and epiglottis was not involved (Figure 2).

Post operatively his voice had improved. No evidence of recurrence of disease following up. Histopathological examination reported as proliferation of spindle-shaped cells admixed with variable amount of mature adipocytes surrounded by thick eisinosplenic raphy collagen in areas (Figure 3). The spindle cells showed bland oval nuclei surrounded by mixture of collagenized and myxoid stroma (Figure 4). The features were in keeping of dendritic fibromyxolipoma.

Figure 1: Right pyriform fossa mass (blue arrow) obscuring view of true cords
Figure 2: Endoscopic view after excision
Figure 3: Spindle-shaped cells admixed with mature adipocytes
Figure 4: Spindle cells (yellow arrows)

Conclusions

• 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.

Discussion

Lipomas of the larynx and hypopharynx are rare, with a site predilection at the false cords, aryepiglottic folds and epiglottis [1]. 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 saliva 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 a possibility of a 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 [2]. Lipomas are non-painful, usually round, mobile 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, required imaging for further assessment and diagnosis [3]. On CT images, lipomas and fibrolipomas appeared as smooth or lobulated well-defined masses associated with moderate displacement of surrounding tissues [4]. El-moneim et al found both CT and magnetic resonance imaging (MRI) to be accurate diagnostic modalities for head and neck lipomas. One weakness in the use of current diagnostic imaging techniques in the diagnosis of tumours of fatty tissue is that neither CT nor MRI can differentiate a lipoma from a liposarcoma [5]. 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 [6].

Dendritic fibromyxolipoma (DFML) has been initially described in 1998 by Suster et al. [7] It was characterized by histologic and immunohistochemical features that reminiscent of spindle cell lipoma and solitary fibrous tumour. The reported median age is 65 years. It has proven recurrence in mass with ratio of 4:1. DFML commonly presents with a subcutaneous mass at the muscular fascia of the shoulder, neck and back [8].

The microscopic feature of DFML composed of spindle cells with anastomosing blood vessels, collagen and prominent mast cells. The spindle cells have typical multiple dendritic cytoplasmatic process, 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].

References


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