Case Report

Solitary Fibrous Tumour of the Submandibular Region: A Rare Entity

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ABSTRACT

Solitary fibrous tumours of the head and neck region are extremely rare. The clinical diagnosis is often difficult to establish, and this lesion may be indistinguishable from other soft tissue neoplasms. An 18-year old Chinese gentleman presented with a painless right submandibular swelling which was increasing in size for eight months. A computed tomography scan showed a well-defined solid mass measuring about 2.0 x 2.96 cm in the submandibular region. The tumour was resected and was confined within its capsule. Immunohistochemical staining was strongly positive for CD34, CD 99, and vimentin and negative for desmin, smooth muscle actin (SMA), cytokeratin, S100 and CD68. The microscopic and immunohistochemical profile were compatible with solitary fibrous tumour. Distinguishing solitary fibrous tumours from various spindle neoplasms can be difficult. In view of the resemblance, immunohistochemical staining can help differentiate solitary fibrous tumour from spindle neoplasm.

Keywords: Solitary Fibrous Tumour, Submandibular

INTRODUCTION

Solitary fibrous tumours (SFTs) of the head and neck region are extremely rare and have often been misdiagnosed due to their rarity. The tumour is a spindle cell neoplasm that commonly arise in the pleura and other serosal regions. Even though more than 50% of these tumours are located in the thoracic cavity, extrathoracic tumours have been reported in various sites such as the liver, skin and the head and neck (1). In the head and neck region, SFTs have been reported in many sites, but the oral cavity is the most common site (1). Although SFTs are commonly benign, 10% to 15% of extrapleural SFTs have shown malignant characteristics in the form of metastatic disease (2). SFTs are slow growing masses that can be very difficult to distinguish from other soft tissue tumours, such as synovial sarcoma, benign fibrous histiocytoma, dermatofibrosarcoma protuberans, neurofibroma, schwannoma, fibroma, and myofibroma. The diagnosis is strongly dependent on the microscopic appearance and characteristic immunohistochemical staining for CD34 and Bcl2. Therefore, definitive diagnosis is usually made post tumour resection.

CLINICAL REPORT

An 18-year-old Chinese gentleman presented with a painless right submandibular swelling that has increased in size during the previous 8 months. The patient denied any related ear, nose, throat and constitutional symptoms. Clinically, there was a right submandibular swelling measuring 3 x 4 cm in size. The swelling was mobile and firm in constituency with normal overlying skin. There were no palpable cervical lymph nodes. The mass was bimanually palpable, with no palpable calculi and the submandibular duct opening was patent. A computed tomography scan was performed which showed a well-defined solid mass measuring about 2.00 x 2.96 cm located inferior and deep to the right mandible, with a large part situated anterior to the submandibular gland (Fig. 1). The enhancing pattern was similar to the submandibular gland, with mild central hypodensity. The preoperative diagnosis was uncertain because the fine needle aspiration cytology failed to yield any conclusive results. The patient then underwent complete removal of the tumour. Intraoperatively, the tumour was found to be confined within its capsule. It was separated from submandibular gland.

Macroscopically, the mass was firm and greyish, with a smooth external surface. It measured 40 x 35 x 15 mm. Upon sectioning and gross examination, it had a homogenous whitish appearance. Histologically, the mass is well circumscribed, consisting of spindle shaped cells within diffuse areas of collagen deposition

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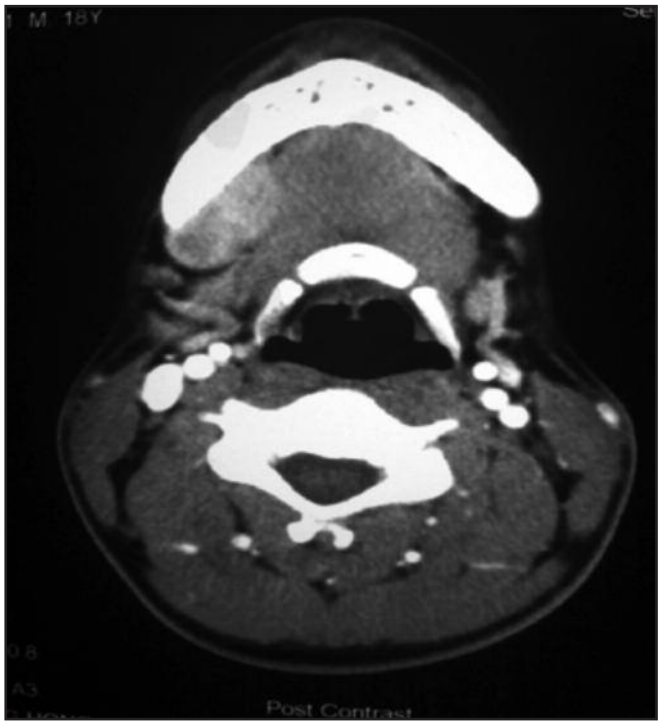

Figure 1: Contrast enhanced computed tomography scan showing a well circumscribed, soft tissue tumour adjacent to the right submandibular gland.

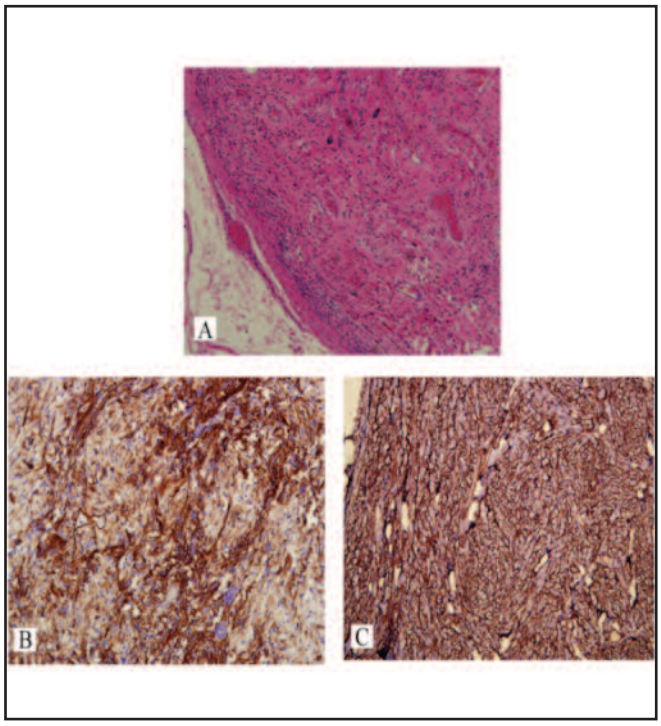

Figure 2: The submandibular mass is well circumscribed, consisting of spindle cells and many dilated and branching blood vessels (hematoxylin and eosin, A, 100x). The spindle cells are positive for CD34 (B, 200x) and CD99 (C, 100x).

(Fig. 2A). The spindle shaped cells displayed vesicular nuclei with dispersed chromatin pattern and indistinct cell borders. Many dilated vascular spaces were present within the mass, some of which formed branching patterns. No atypia, mitosis or necrosis was present.

Immunohistochemical studies showed that the spindle cells were strongly positive for vimentin, CD99 and CD34 and negative for desmin, smooth muscle actin, cytokeratin, S100 and CD68 (Fig. 2B and 2C). The gross and microscopic morphological features, supported by the immunohistochemical studies were consistent with the diagnosis of SFTs. The patient's postoperative course was uneventful, with no recurrence during 2 years follow-up.

DISCUSSION

SFTs were initially associated only with mesothelial-lined surfaces, most commonly arising from the pleura and other serosal regions. SFTs usually present as slow-growing and painless masses, as in this case. The clinical symptoms are dependent on location, usually due to compression rather than tissue infiltration. Although SFTs are commonly benign, malignant properties have been reported in masses arising from the sublingual and parotid glands.

There are no absolutely distinctive imaging features that are diagnostic of these rare tumours. Dense enhancement of the lesion on computed tomography or magnetic resonance imaging may be suggestive and should prompt inclusion of SFT as a differential diagnosis (3).

The classic histologic appearance of an SFT is characterised by a variety of growth patterns, the most typical being a random arrangement of spindle cells in a collagenous background, with a prominent vascularity that results in a hemangiopericytoma-like pattern. Mitoses and necrosis are usually absent. However, an estimated 10% -15 % of benign SFT can behave in a malignant fashion (2). Histological features used to determine malignancy include high cellularity, marked cytological atypia, frequent mitoses, tumour necrosis, and infiltrative borders-none of which were present in this case.

Histologically, a number of other soft tissue neoplasms have a striking resemblance to SFTs. Therefore, differential immunohistochemical staining helps distinguish SFTs from other soft tissue tumours, including benign fibrous histiocytoma, dermatofibrosarcoma protuberans, myofibroma, fibromas, and neurogenic tumors, and other vascular soft tissue tumours, such as hemangiopericytomas and synovial sarcomas.

In the immunohistochemical studies, the spindle cells are typically positive for vimentin, CD 34, Bc1-2 and CD 99 and negative for S-100 protein, cytokeratin, desmin, actin, and myogenin (4). Benign fibrous histiocytomas typically stain negative for CD34 and Bcl2 (3).

In contrast, CD34 and Bcl2 may be reactive in neurofibromas and schwannomas, but these tumors also stain positive for S100. Myofibromas and fibromas stain strongly positive for vimentin, smooth muscle actin, and muscle-specific actin but stain negative for CD34. Dermatofibrosarcoma protuberans is frequently CD34 positive but often Bcl2 negative (3). SFTs can be differentiated from synovial sarcoma based on the fact that over 80% of synovial sarcomas have a specific chromosomal rearrangement, t(X,18) (p11.2;q11.2) (5).

In our case, the tissue showed marked positivity for Vimentin, CD99 and CD34, while it was negative for desmin, smooth muscle actin, cytokeratin, S100 and CD68. Therefore, the microscopic and immunohistochemical profile was consistent with a benign solitary fibrous tumour.

The recommended management for benign SFT is complete surgical excision of the neoplasm, and such excision with free margins is an important prognostic factor. Recurrence of SFTs have been reported 30 years after surgical excision despite complete excision (2). Therefore, long-term follow up is essential even in cases pathologically diagnosed as benign.

CONCLUSION

Clinicians should include SFT in the differential diagnosis of tumours that arise from soft tissues of the head and neck. Distinguishing a SFT from various spindle neoplasms can be difficult. Therefore, immunohistochemical staining is important. Suggested management is complete excision of the mass with long term follow up, because of the potential for recurrence and malignant transformation.

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