

Solitary Fibrous Tumor Of The Deep Soft Tissues Of The Neck

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Citation

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Abstract

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm. Since its initial description as arising from the pleura, SFT has been reported at a wide range of anatomic sites. However, report of SFT in deep soft tissue of the neck is very rare. The authors present a case report of a rare atypical solitary fibrous tumor of the deep soft tissue of the neck. A 69-year-old woman presented with 1 year history of an enlarging, firm mass in the left posterolateral region of the neck. The tumor was characterized by a mesenchymal proliferation with areas of varying cellularity, a mild cellular atypia and hemangiopericytoma-like vessels. The tumor cells were positive for CD34, bcl-2 and CD99 and negative for EMA, cytokeratins (MNF116, CAM 5.2, AE1/AE3), S-100 protein and smooth muscle actin. The picture was coherent with atypical solitary fibrous tumor. At 2 years after surgery the patient was well with no evidence of recurrence.

INTRODUCTION

SFT is a rare mesenchymal neoplasm originally described in the pleura [1] but more recently reported at a wide range of anatomic sites including two cases in deep soft tissue of the neck [2,3]. This tumor is characterized histologically by a variety of growth pattern and can be confused with other benign or malignant neoplasms. This report describes a case arising from the deep soft tissue of the left posterolateral region of the neck.

CASE REPORT

A 69-year-old woman presented with 1 year history of an enlarging, painless mass in the left posterolateral region of the neck; the lesion was rapidly increasing in size in the last 4 months. She had only a medical history of hypertension. On examination the lump was ovoid (10x8 cm), non-tender and firm. There were no other lumps in the neck or axilla. Chest X-ray was normal. Blood tests were within normal limits.

Fine needle aspiration cytology (FNAC) showed no evidence of malignant cells.

A CT and MR scan showed a 8 x 5cm vascularized mass with high signal intensity on T2-weighted images and a low T1 signal intensity occupying the posterior cervical triangle from the clavicle to the posterior border of the mastoid. The mass expanded the scalenes muscles without infiltration.

A 4-vessel arteriogram was performed. The feeding vessels appeared to arise from several branches of the left vertebral artery.

A left cervical incision was performed. The tumor was dissected from beneath the trapezius muscles and other surrounding muscular masses. Frozen-section evaluation revealed a mesenchymal neoplasm not otherwise specified. Definitive histopathological examination showed a SFT.

Macroscopically the tumor was a lobulated mass measuring 7x5x3cm with a cystic yellow cut surface separated by white fibrous tissue.

Microscopically the tumor was characterized by a mesenchymal proliferation with areas of varying cellularity and hemangiopericytoma-like vessels (Figure 1). A mild cellular atypia was present and mitotic activity was < 2 mitoses/10 HPF. The tumor cells were positive for CD34, bcl-2 and CD99 and negative for EMA, cytokeratins MNF116 (Figure 2), CAM 5.2, AE1/AE3, S-100 protein and smooth muscle actin.

Figure 1

Figure 1: The “tumor” is characterized by a mixture of spindle cells with tapered nuclei, disposed in hyper- and hypocellular areas, in which bundles of sclerotic collagen here and there separate the groups of proliferating elements; the spindle cells are haphazardly arranged, but in areas may be oriented in a vague storiform pattern. (E.E: 100x)

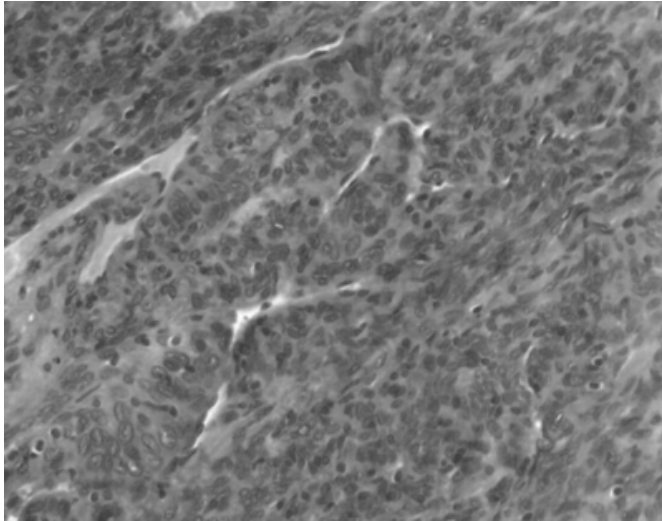
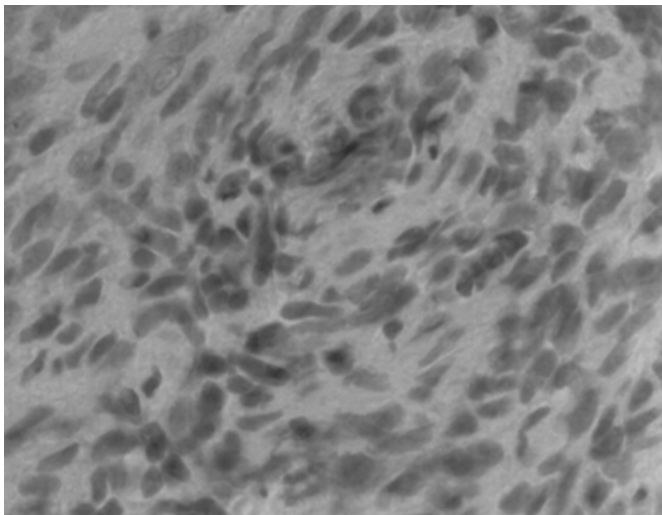


Figure 2

Figure 2: The proliferating elements appear immunohistochemically MNF116 negative (400x)



The pattern was coherent with atypical solitary fibrous tumor.

The patient had an uneventful post-operative recovery and at 2 years follow-up was well with no evidence of recurrence.

DISCUSSION

SFTs of soft tissues, first described in 1931 [1], are rare spindle cell tumours which occur mainly in adults and can be either benign or malignant. Originally described in the

pleura, more recently they have been described at extrapleural sites such the parenchyma of the lungs, abdominal cavity, orbit, thyroid gland, upper respiratory tract, nose and paranasal sinuses, salivary glands, parapharyngeal space, epiglottis, tongue, cranial nerve and soft tissues [4,5,6,7,8,9,10,11,12,13,14].

SFTs occurring in the soft tissues present as a well-circumscribed subcutaneous or deep rubbery mass from 1 to 6 cm. The cut surface is usually pale or firm. They are unencapsulated and characterized by various growth pattern with foci of sclerosis and a mixture of cellular spindle areas alternating hyper- and hypo-cellular areas; the presence of rope-like collagen separating the tumour cells is distinctive. Blood vessels may be prominent and often have thickened walls with a focally hemangiopericytoma-like vascular pattern [15]. Mitotic figures are uncommon; necrosis and invasion of surrounding structures are not seen.

Diagnostic criteria have recently been elaborated [16] and immuno-histochemical and ultrastructural studies have shown cells closely resembling fibroblasts in differentiation [5]. The majority of SFTs have been immunoreactive for CD34 which is expressed in hematopoietic stem cells, endothelial progenitor cells and vascular endothelial cells with idiosyncratic positivity in several non-vascular lesions; authors believe that its expression is a definitive marker of this tumor [17]. SFTs are negative for epithelial, neural and smooth muscle markers.

The incidence of aggressive behaviour has been described in 20 per cent of pleural tumors; almost all extrapleural SFTs have pursued a benign clinical course [18] but extrathoracic SFTs seem to have an increased risk of local recurrence [19] and a malignant potential is described [20]. However the clinical behaviour is unpredictable because some “benign” tumors have behaved in an aggressive manner and some with a “malignant” appearance have behaved in a benign manner.

Proper diagnosis of these tumors is essential to avoid a confusing diagnosis with a variety of benign and malignant neoplasms: hemangiopericytoma, schwannoma, mesothelioma, sarcomatoid carcinoma, fibrosarcoma, synovial sarcoma, leiomyosarcoma and leiomyoma.

Treatment of choice is conservative surgery and resectability is the most important prognostic factor [21,22]; a careful and long-term follow up is required.

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