Solitary Fibrous Tumor of Thigh: A Case Report

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Solitary fibrous tumor (SFT) is an uncommon mesenchymal tumor and is classified as an Intermediate (rarely metastasizing) fibroblastic/myofibroblastic tumor (WHO 2013). It occurs as slow growing masses at any location in deep soft tissue, abdomen/pelvis, pleura, the extremities and other sites. We present a case of a 60-year-old woman with a slow growing swelling of the right thigh. On magnetic resonance imaging, a large lobulated, heterogeneously enhancing septated lesion was noted. Wide excision of the lesion showed a well-circumscribed tumor with gray white and mucoid areas. Light microscopic examination showed a circumscribed, partially encapsulated tumor composed of hypercellular areas with intervening hypocellular, hyalinized areas were seen and tumor cells showed diffuse strong CD34 positivity. Based on histopathology and immunohistochemistry (IHC), a final diagnosis of SFT was offered. SFTs belong to a spectrum of neoplasms from benign to malignant which can occur at myriad sites in the body. IHC forms the basis for confirmatory diagnosis. To preventmislabeling as sarcoma, a high degree of suspicion and radio-histological correlation is warranted to arrive at a conclusive diagnosis.

Keywords: CD 34 antigen, Desmin, Immunohistochemistry, Thigh, Vimentin

INTRODUCTION

Solitary fibrous tumor (SFT) is an uncommon mesenchymal tumor and is classified as an intermediate (rarely metastasizing) fibroblastic/myofibroblastic tumor (WHO 2013) encompassing a spectrum of lesions including the pleura based SFT at one end with its predominantly fibrous component and “hemangiopericytoma” with its characteristic vascular pattern at the other end. The majority of these neoplasms are benign in course, seen in adults with equal sexual predilection and occur as slow growing masses at any location in deep soft tissue, abdomen/pelvis, pleura, the extremities and other sites. We present a case of a 60-year-old woman with a slow growing swelling of the right thigh.

CASE REPORT

A woman aged 60 years presented with gradually increasing swelling over the right thigh, associated with pain and in frequent moderate grade fever for the last 2 years.

On physical examination, a hard mass measuring 8 cm × 10 cm was noted, located on the anterior aspect of right thigh. Computerized tomography showed a heterogeneous enhancing lobulated lesion involving the anterior compartment muscles of right thigh (vasti medialis and intermedius muscle). On magnetic resonance imaging, a large lobulated, heterogeneously enhancing septated lesion with altered signal intensity having calcification and blood fluid level was noted in the flexor compartment of right mid-thigh with no evidence of infiltration of adjoining femur.

An incision biopsy done could only provide a histopathological diagnosis of benign spindle cell tumor of soft tissue as hypocellular areas had been sampled. Subsequently, wide excision of the lesion was performed.

Grossly, a tumor weighing 120 g with overlying skin was received, which on cut section, showed a well-circumscribed tumor measuring 12.5 cm × 6.5 cm × 6.5 cm with gray white and mucoid areas, cystic spaces and hemorrhagic specks (Figure 1).

Light microscopic examination showed a circumscribed, partially encapsulated tumor composed of hypercellular areas with short splitting fascicles, focal herringbone pattern of spindle-like and stellate cells with indistinct cytoplasm, ropy collagen interspersed with areas exhibiting...
tissue culture pattern of plump epithelioid to round cells with minimal nuclear atypia, vesicular chromatin, 1-3 inconspicuous nucleoli and mitosis (1/10 high power fields [HPF]) (Figure 2a-c).

Intervening hypocellular, hyalinized areas were seen along with thick walled vessels with vascular wall hyalinization. Extensive areas of collagenization were noted and focal areas of hemorrhage. Spindle areas showed infarction, degenerative changes; epithelioid cell areas showed scattered bizarre cells, multinucleated tumor giant cells, rosettes and microcystic spaces (Figure 2d).

On immunohistochemistry (IHC), the tumor cells showed diffuse strong CD34 positivity. BCL2, vimentin (cytoplasmic), CD99, CK (focal) were also positive (Figure 3a-d). Smooth muscle actin (SMA), S-100 and epithelial membrane antigen (EMA) were negative. Ki67 index was low (3%) (Figure 4a and b). A final diagnosis of SFT was offered histopathologically.
DISCUSSION

Extrapleural SFTs are commonly seen as an incidental finding in pleura and present as slow growing masses, commonly associated with hypoglycemia in other soft tissue locations. These tumors are associated with a recurrent rearrangement involving chromosome 12q, forming NAB2-STAT6 fusion oncogene.1,3
Radiographic findings show large, lobulated, heterogeneously enhancing tumor with high vascularity and prominent collateral feeding vessels, as seen in the present case, or a visible fatty component.4

According to the new WHO 2013 classification, SFT hemangiopericytoma, lipomatous hemangiopericytoma and giant cell angiofibroma are all clubbed under the “extrapleural SFT” category.5

Grossly, these tumors are circumscribed, often lobulated/ nodular lesion,1-4 1-40 cm in diameter,2 with a median size of 10.3 cm, with gray-white to red-brown cut surface.5 Histopathology shows a lesion of moderate cellularity, composed of spindled to round-fusiform cells arranged in a patternless manner with extensive areas of hyalinization, a ramifying network of blood vessels with sinusoidal vessels demonstrating staghorn appearance and large blood vessels with thick coats of collagen. Pseudovascular spaces, giant cells, cystic change may be noted. Sharply demarcated poorly differentiated areas may be seen. Necrosis and marked cellularity may also be seen.2 The above-mentioned features were also seen in the present case. The tumor cells show CD34, CD99, BCL2, SMA and EMA positivity and are negative for S-100 and desmin. Cytokeratin may show variable positivity.5,6

With the discovery of recurrent cytogenetic abnormality involving the long arm of chromosome 12 in SFTs, a new IHC marker, STAT6, has been shown to have nearly 100% specificity.5

Most tumors are histologically benign, with 10-20% showing an aggressive course. The malignant behavior is difficult to predict but is seen to be associated with a tumor size >15 cm, increased cellularity, pleomorphism, high mitotic count (>4/10 HPF) and evidence of metastasis.5,6 5 years disease-specific survival rates vary in different studies from 60% to 93%.2,7 Pleural lesions are also noted to have higher rate of metastasis.2

A differential diagnosis of deep fibromatoses and monophasic synovial sarcoma were considered on histopathology. Both these tumors show prominent vascularization with spindled to epithelioid cells in a collagenous matrix. Deep fibromatoses show glistening white; coarsely trabeculated surface on macroscopic examination and IHC demonstrates a strong vimentin expression with variable muscle-specific and smooth muscle actin.8 Rare cells may also express desmin and S100 protein; while synovial sarcoma is cytokeratin positive and CD34 negative, thus aiding in narrowing down the diagnosis in our case.6,8

Owing to the uncertain course of this neoplasm, long-term follow-up is recommended.4

CONCLUSION

SFTs belong to a spectrum of neoplasms from benign to malignant which can occur at myriad sites in the body. High vascularity and nodularity form the core to diagnosis on radiology, a patternless pattern to fascicles of spindled and stellate cells with herringbone pattern of vessels on HPE; IHC forms the basis for confirmatory diagnosis. The occurrence of hypo and hypercellular areas may cause misdiagnosis on trucut biopsy, depending on the area sampled. To prevent mislabeling as sarcoma, a high degree of suspicion and radio-histological correlation is warranted to arrive at a conclusive diagnosis.

REFERENCES