Case Report

Recurrent solitary fibrous tumor in distal lower extremity: An extremely rare entity

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Abstract

Solitary fibrous tumor (SFT) represents a spectrum of mesenchymal tumors, encompassing tumors previously termed hemangiopericytoma, as having intermediate biological potential. Though they can occur at any site, lower distal extremity is a rare site and recurrence in it is extremely rare. Behavior of SFT is unpredictable. Histomorphology and clinical follow-up have poor correlation. The most important single indicator of clinical outcome is complete excision of the tumor at the time of primary presentation. Tumors with positive margins require close follow-up for several years owing to the potential for late local recurrence.

Key words: CD 34, fibrous, recurrence, spindle cells

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Introduction

Solitary fibrous tumors (SFTs) were formerly thought to be limited to mesothelial covered surfaces (Pleura). Now, they have been described at almost every anatomic location and usually occur in the elderly. Extra pleural SFT accounts for 0.6% of all soft-tissue tumors.[1] Curiously soft-tissues of extremity are among the rarest site of occurrence. Most occur in the proximal lower extremity and tend to have more malignant potential.[2,3]

Case Report

This was a case of a 15-year-old boy who was admitted with 6 months history of recurrent painless mass (8 cm × 4 cm) on the upper 1/3 anteromedial aspect of left leg [Figure 1a Arrow]. Patient had undergone prior surgical resection of mass from the same site, one year prior to the current presentation. Clinical examination and laboratory test were otherwise non-contributory except for the soft-tissue tumor. Repeated fine-needle aspirations were hemorrhagic and hypo cellular. Microscopy showed occasional small cluster and dispersed spindle cells with evenly distributed chromatin. A diagnosis of spindle cell lesion was suggested. Patient's previous tumor from the same site was classified as SFT based on histomorphology and immunohistochemical (IHC) studies. Patient underwent surgical removal of the mass. Surgery and post-operative period were uneventful.

Grossly, a well-delineated (7.5 cm × 3 cm × 1.5 cm) white mass was received for histopathological examination [Figure 1b]. Histomorphology of routine hematoxylin and eosin stained slides was similar to previous resected tumor and showed haphazardly arranged spindle cells (pattern less growth), dense collagen and areas of marked hyalinization. Spindle cells have oval to elongate bland vesicular nuclei with a moderate amount of cytoplasm. No significant nuclear pleomorphism or mitotic activity was noted [Figure 1c and d]. Many thin walled blood vessels were seen [Figure 1e]. Microscopic foci of mature adipose tissue were seen [Figure 1f]. Surgical margins were free of tumor tissue [Figure 2a]. Tumor cells were negative for actin, desmin, epithelial membrane antigen (EMA), S100 and strongly positive for CD34 immunostains [Figure 2b-f]. A diagnosis of SFT was made.

Discussion

SFT represents a spectrum of mesenchymal tumors encompassing tumors previously termed as hemangiopericytoma. They are composed of subset of

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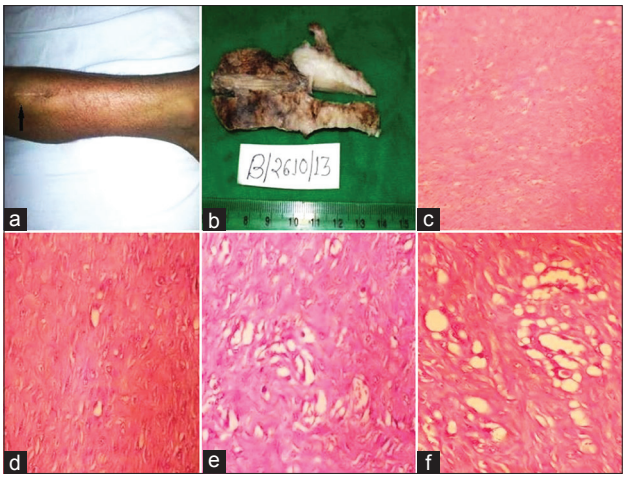

Figure 1: (a) Swelling and scar on upper 1/3 of lower distal extremity. (b) Tumor. (c and d) Solitary fibrous tumor (SFT) - Spindle cells with collagen (H and E, ×100 and ×400). (e) Vascularity in SFT (H and E, ×400). (f) Microscopic foci of adipose tissue in SFT (H and E, ×400)

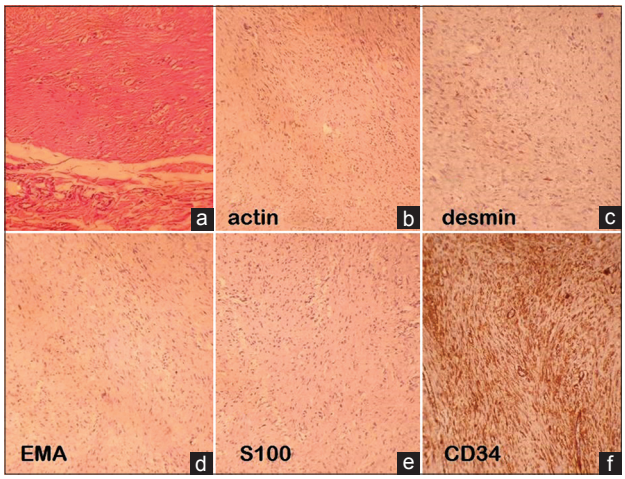

Figure 2: (a) Solitary fibrous tumor (SFT) with free surgical margin (H and E, ×400). (b-f) Negative staining of SFT with actin, desmin, epithelial membrane antigen, S100 and strong positivity with CD34 (IHC, ×100)

fibroblast like cells. They are classified as tumors of intermediate biologic potential with low risk of metastasis and have relatively indolent course under the 2002 World Health Organization classification scheme.[4] Most commonly SFT present during 5th and 6th decade of life and there is no significant sex predilection.[2]

Though previously thought to be confined to pleura, extra thoracic SFT by now have been reported at almost every anatomic location. Tumors in extremities still represent a rare entity of soft-tissue neoplasms.[5] Number of reported cases in the extremities are few, according to Fakui *et al*.[6] There were only 28 cases reported, including their two cases involving shoulder and thigh. The common sites in extremities from which tumor have been reported include thigh, popliteal fossa, flank, neck, shoulder, deep groin and gluteal region. Our patient was a young boy and had recurrent lesion at upper 1/3 of lower distal extremity.

Due to overlapping of histomorphological features of SFT with other soft-tissue tumors in extremities, precise pathological characterization necessitates to evaluate specimen for proper diagnosis and for detection of malignant features. Classic SFT show pattern less arrangement of spindle cells in dense collagen with prominent vascularity that result in hemangiopericytoma like pattern.

SFT at times can be confused with many benign and malignant lesions having prominent pericytic vascular pattern such as synovial sarcoma, mesenchymal chondrosarcoma, juxtaglomerular tumor and fibrous histiocytoma.

However, patternless arrangement of spindle cells in dense collagen helps distinguishing SFT from other soft–tissue, which happened in our case. Unusual features such as pseudo vascular spaces, cystic change and giant cells can confuse the histology of SFT. These features were not seen in our case.

Tumor cells in SFT are usually negative for actin, desmin, cytokeratin, EMA, S-100. However, focal immunopositivity with cytokeratin, actin, S-100 and EMA may be seen. Tumor cells are strongly positive for CD 34 in most cases.[4] However, its expression can be lost in high-grade tumors or tumors with repeated recurrence.[7] SFT may present with malignant behavior and local recurrence or metastasis. Pathological criteria for malignant SFT is based on high cellularity, >4 mitotic figures/10 hpf and hemorrhage/necrosis.[8] Local recurrence is defined as tumor involving the tumor bed or a scar of a previous surgery.

In a study by Vallat-Decouvelaere *et al*.[9] suggested atypical histological features such as nuclear atypia, areas of increased cellularity, necrosis and >4 mitotic figures/10 hpf as predictive for clinical malignant behavior of the tumor and local or distant relapse in 80% of cases. Gold *et al*.[1] in their study observed that primary tumor size of >10 cm and positive surgical resection margin correlated with unfavorable clinical outcome. Factors that predispose to local recurrence are: Tumor size >10 cm, malignant component on histology or microscopic positive surgical margin. In our patient, though atypical/malignant features were not seen in prior specimen, possible cause of recurrence may be incomplete removal.

Behavior of SFT is unpredictable. Though malignant histopathological features remain an indicator of poor clinical outcome, it should be emphasized that histological appearance does not correlate well with the clinical behavior in SFT.

A more recent study of large series of extra pleural SFT with a long follow-up period reported by Cranshaw *et al*.[10] observed that they behave clinically in the same manner similar to high grade soft-tissue sarcoma with a high rate of recurrence, metastatic spread and overall poor prognosis. But the most important single indicator of clinical outcome is complete excision of tumor at the time of primary presentation.[11] Late recurrence is one of the clinical characteristic of SFT. Treatment of choice is complete surgical resection with disease free margins.Adjuvant radiotherapy and chemotherapy may be used in malignant variants. Prolonged follow-up is advisable.

Conclusion

Recurrent SFT in distal lower extremity is extremely rare. Careful histological examination and IHC studies are essential for definitive diagnosis. Behavior of SFT is unpredictable. Treatment of choice is complete surgical extirpation with disease free margin.

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