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Extrapleural Solitary Fibrous Tumor (SFT) at Dorsum Foot - A Rare Case Report.

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ABSTRACT

Solitary fibrous tumor is a benign tumor. But it has tendency for local recurrence as well as metastasis. WHO classifies it under the category of fibroblastic/myofibroblastic tumor. It most commonly occurs in pleura and very rare in extra-pleural site such as sub-cutaneous or deep soft tissue of extremity, head and neck, mediastinum, retroperitoneum, etc. Histologically SFT show patternless hypercellular and hypocellular areas of fibroblast proliferation, collagen deposition, hemangiopericytoma like areas with myxoid change or fibrosis. This tumor needs to be differentiated from other spindle cell tumors with immunohistochemistry. It is a rare entity with only a few cases reported. We report a case of 60 years old male with well circumscribed painless, swelling dorsum foot. It was clinically diagnosed as inverted papilloma, but microscopy was consistent with typical features of SFT. This was confirmed by CD 34 & CD99 positivity. This case is being presented for its rare location.

Keywords: Solitary fibrous tumor, hemangiopericytoma, Collagen, patternless.

INTRODUCTION

Solitary fibrous tumor is an intermediate rarely metastasizing tumor. It is classified by WHO under the category of fibroblastic/myofibroblastic tumor.[1] It usually occurs in the pleura, but extra-pleural sites are also reported.[1-4] Occurrence at extra-pleural sites is rare and soft tissue sites account for 0.6% of all soft tissue tumors.[4,8] Extra-pleural sites include sub-cutaneous or deep soft tissue of extremity, head and neck, mediastinum, retroperitoneum, etc. Extra-pleural SFT is seen in age group of 20-70 years.[1] Like pleural SFT, extra-pleural SFT are usually benign, but 10% recur locally or metastasize.[4,7].

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Histologically, typical SFTs show a patternless architecture characterized by a combination of alternating hypocellular and hypercellular areas separated from each other by thick bands of hyalinized, somewhat keloidal collagen and hemangiopericytoma-like vessels. Myxoid change, areas of fibrosis are commonly observed.[1,3] Immunohistochemistry for CD34, CD99 and bcl2 help for confirmatory diagnosis of SFT. Only a few case reports document the occurrence of extra-pleural SFT. We report a case of 60 years old male. He presented with well circumscribed painless swelling on dorsum of foot and was clinically diagnosed as inverted papilloma. Histologically, it showed typical features of SFT. Von Gieson stain confirmed collagen and Immunohistochemistry showed positivity for CD34 and CD99.

CASE REPORT

We present a case of 60 years old male patient who came in surgery outpatient department with complaints of painless swelling on dorsum of left foot. On examination, swelling was well circumscribed, soft to firm in consistency and non-tender. Clinically, it was diagnosed as inverted papilloma and was advised excision. The swelling was excised and sent for histopathological examination in our department of pathology. On gross examination, there were two skin covered soft tissue pieces together measuring 1x1 cm and the material was whole passed.

On microscopic examination, there was seen epidermis and dermis. Epidermis showed papillomatous hyperplasia. Upper dermis was unremarkable and lower dermis showed

hypercellular and hypocellular areas of fibroblast proliferation, hemangiopericytoma like areas along with extensive keloid like collagen and focal myxoid change. Histopathological features were consistent with solitary fibrous tumor. Cytochemistry was done with Von-Gieson stain, which stained collagen part of tumor as red. Final confirmation was done by immunohistochemistry using CD34 & CD99. CD 34 showed weak focal positivity and CD99 showed strong diffuse positivity. Correlating H& E, Von-Gieson stain and immunohistochemistry findings; final diagnosis of solitary fibrous tumor was made. (See figures A-E).

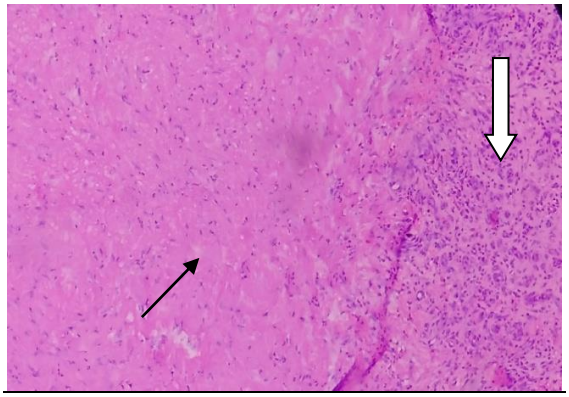

Figure A: 10X Microscopic view (H & E) showing hypercellular (thick arrow) and hypocellular (thin arrow) areas.

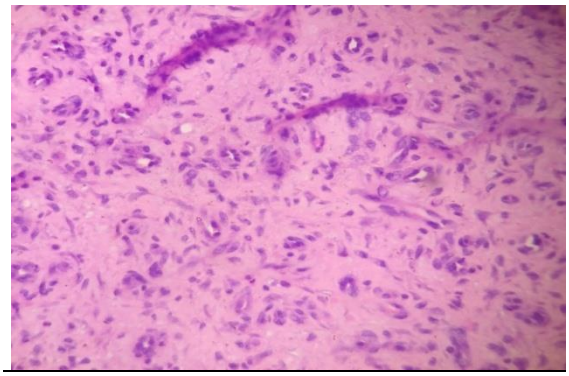

Figure B: Low (100x) and high power (400x) views of hemangiopericytoma like areas.

Figure C: 400X Microscopic view of Von-Gieson's stain showing collagen as red.

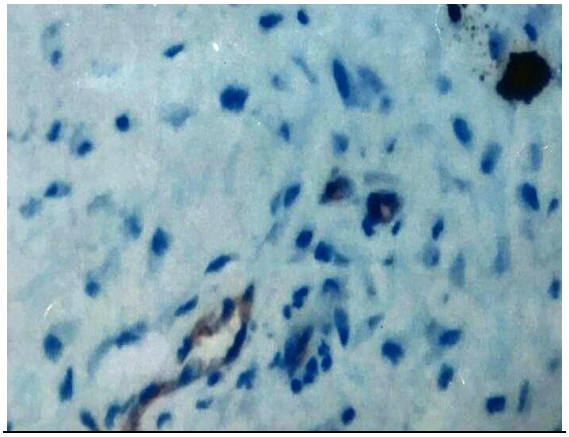

Figure D: Immunohistochemical staining for CD34 showing focal positivity.

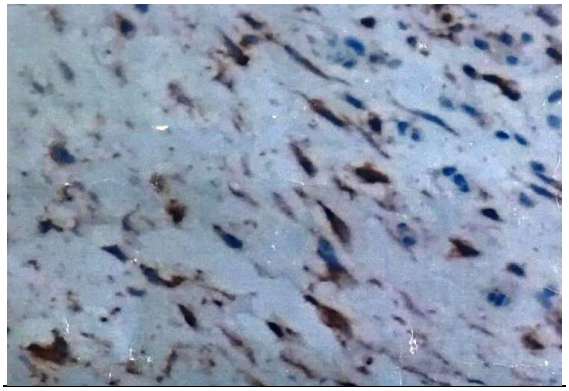

Figure E: Immunohistochemical staining for CD 99 showing strong diffuse positivity.

DISCUSSION

Soft tissue SFT is a rare entity with only a few cases reported. Clinically, extra-pleural solitary fibrous tumor cause symptoms related to tumor size and location. Systemic symptoms have also been reported such as hypoglycemia, arthralgia, osteoarthritis and clubbing. These symptoms subside upon tumor resection.[6] In our case, there was painless well circumscribed swelling on dorsum of left foot with no systemic features. Clinically it was diagnosed as inverted papilloma.

SFT needs to be differentiated from various other spindle cell lesions such as, hemangiopericytoma, smooth muscle tumor, GIST, nerve sheath tumor and fibrous histiocytoma. Histopathology helps in this differentiation. Collagen is not seen in hemangiopericytoma. Fascicular growth pattern is seen in smooth muscle tumors. Frequent palisading is seen in nerve sheath tumors and fibrous histiocytoma shows predominantly storiform pattern as well as histiocytic differentiation.[1]

Histologically, SFT shows patternless architecture with alternating hypocellular and hypercellular areas separated from each other by thick bands of hyalinized, somewhat keloidal collagen and hemangiopericytoma-like vessels. Myxoid change,

areas of fibrosis are commonly observed.[1,3] Our case was histologically consistent with these typical features of SFT.

Cytochemistry further helps (as adjunctive) in differentiating SFT from other spindle cell tumours e.g. Von Gieson stain helps to stain collagen as red.

IHC is essential in differentiating SFT from other spindle cell neoplasms. Hemangiopericytoma, nerve sheath tumor, fibrous histiocytoma show focal CD34 positivity. GIST and SFT show diffuse CD34 positivity. Smooth muscle tumors are desmin positive. GIST is positive for CD117 and DOG1.[1] Positivity for CD34, bcl-2 and CD99 indicate SFT.[3,4] In our case, tumor showed positivity for CD34 and CD99.

Behaviour of SFT is unpredictable. Some histologically aggressive tumors are benign, while some morphologically benign tumors show aggressive behaviour. 10-13% recur and metastasize.[7] They may also recur or metastasize even after complete surgical excision. Therefore complete surgical excision and long term follow-up recommended.[5]

CONCLUSION

SFT is a benign tumor, which is capable of aggressive behaviour in the form of local recurrence as well as distant metastasis. It usually occurs in pleura, where it's mostly benign. But extra-pleural locations also exist which are rare. Only few case reports suggest such existence. Extra-pleural SFT may show unpredictable behaviour. Histologically, both pleural and extra-pleural SFT are similar. Histopathology, cytochemistry and immunohisto-chemistry help in differentiating SFT from other spindle cell tumors. Treatment is by complete surgical excision, but because of unpredictable behaviour, long-term follow up is necessary.

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