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Case Study

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# POORLY DIFFERENTIATED SYNOVIAL SARCOMA: REPORT OF TWO CASES WITH UNUSUAL PRESENTATION

**Prajwala Gupta[1], Deepika Gupta*[1], Minakshi Bhardwaj[1], Aastha Gupta[2]**

[1]Department of Pathology, Atal Bihari Vajpayee Institute of Medical Sciences and Dr. RML Hospital.
[2]Department of Dermatology, Atal Bihari Vajpayee Institute of Medical Sciences and Dr. RML Hospital, New Delhi.

***Corresponding Author: Deepika Gupta**
Department of Pathology, Atal Bihari Vajpayee Institute of Medical Sciences and Dr. RML Hospital, Delhi.
**Email ID:** deepikagupta351@gmail.com,

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## ABSTRACT

Synovial sarcomas are rare malignant soft tissue tumors affecting mainly young adults and presenting as a slow growth mass located in the deep soft tissues of extremities, near the joints. The poorly differentiated forms may be indistinguishable from other round cell tumors based on the morphology alone or at times by immunohistochemical studies. Here in, we describe 2 unusual presentations; first one as an extremely rare case of poorly differentiated synovial sarcoma of the foot metastasizing to inguinal lymph nodes and other case presenting as a localised swelling in the palm. Fine needle aspiration cytology (FNAC) with immunohistochemistry (IHC) was helpful in the early diagnosis and differentiating it from its closest differential Ewings sarcoma.

**KEYWORDS:** Immunohistochemistry, synovial sarcoma, FNAC, Ewings sarcoma, metastasis.

## INTRODUCTION

Synovial sarcoma (SS) is a rare malignant soft tissue tumor and it comprises 5.6 % to 10 % of all soft tissue sarcomas.[1] The tumor usually occur predominantly in the extremities, where they tend to arise in the vicinity of large joints, especially the knee region and are intimately related to tendon sheaths, bursae, and joint capsules.[2]

SS presenting in foot and palm are rare and even rarer to present with metastasis.[3],[4]We herein describe 02 cases of poorly differentiated SS with extremely unusual presentations wherein fine-needle aspiration cytology (FNAC) with immunohistochemistry (IHC) was helpful in early diagnosis.

## CASE REPORT 1

A 28 yr old female presented with rapidly increasing swelling on medial side of left foot since 1month and left sided inguinal lymph node swelling for past 15 days. On local examination, the foot swelling was lobulated measured 4x5 cm,surface ulcerated, firm to hard in consistency, fixed and non tender [Figure1]a. The left sided inguinal lymph node was discrete measured 1.5x1.5cm, firm in consistency, fixed and non tender. FNAC smears from inguinal lymph node revealed high cellularity comprising tumor cells arranged in loosely cohesive clusters, fragments and many scattered singly. The cells were round to oval with moderate anisonucleosis, granular chromatin, inconspicuous nucleoli and moderate amount of pale cytoplasm in a background of lymphoid cells [Figure 2]a. Biopsy from foot mass was done and histopathological examination revealed tumor composed of sheets and nests of round to ovoid cells with hyperchromatic nuclei and scant amount of eosinophilic to clear cytoplasm in a stroma showing necrosis and myxoid changes [Figure 2]b. The IHC revealed positivity for Vimentin, PanCK, CD99 [Figure 2]c and BCl-2 [Figure 2]d. Final diagnosis was of SS with metastatic deposits in ipsilateral inguinal lymph node. Patient was referred to Oncology centre for further treatment after which the patient was lost to follow up.

## CASE REPORT 2

A 42-year-old male presented with a soft tissue swelling in right palm for the past 4 months. He also complained of pain and difficulty in moving fingers. On examination, the swelling measured 4x4cm, firm, non mobile, non tender and located near the base of thumb in the thenar region [Figure 1]b. Magnetic resonance imaging (MRI) of hand revealed a lobulated mass lesion in the region of thumb involving thenar muscles and abutting bones of left thumb [Figure1]c. On Doppler study, tumor showed high vascularity. Based on the radiological findings, differential diagnosis of high flow AV malformation and soft tissue sarcoma was suggested. FNAC done from palm swelling revealed high cellularity comprising round to oval tumor cells arranged in large cellular fragments, clusters, groups, vague gland like arrangement and many singly scattered. These cells had granular chromatin, inconspicuous to prominent nucleoli and scant to moderate amount of cytoplasm.Frequent mitosis were also noted [Figure 2]e. Cytomorphological Features were suggestive of small round cell tumor (SRCT) and in conjunction with overall clinical presentation, following

differentials of poorly differentiated SS and extraskeletal Ewing's sarcoma were considered. Cell block was made and sections revealed sheets of round to oval tumor cells with granular chromatin, inconspicuous to prominent nucleoli and scant cytoplasm [Figure 2]f. IHC was positive for CD99[Figure 2]g, Vimentin and Bcl-2 [Figure 2]h. CD34, Pan CK, EMA were negative. Poorly differentiated SS was given as the final diagnosis which was subsequently confirmed on histopathology. After the initial excision the patient completed 4 cycles of chemotherapy and was doing well thereafter. Follow up at the end of 6 months did not show any recurrence or metastasis.

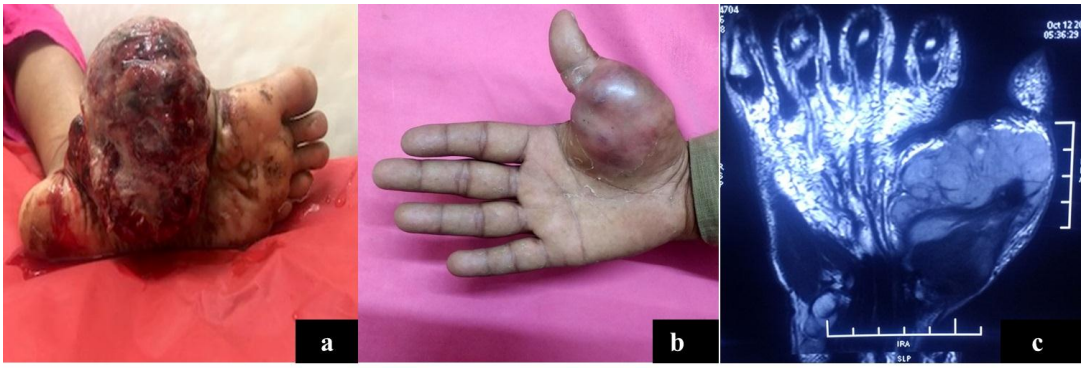


**Figure 1: (a) Large lobulated soft tissue mass over left foot. (b) Palmar aspect of the right hand shows large mass in the thenar region. (c) Magnetic Resonance Imaging (MRI) shows lobulated mass lesion involving thenar muscle and abutting the bones of thumb.**

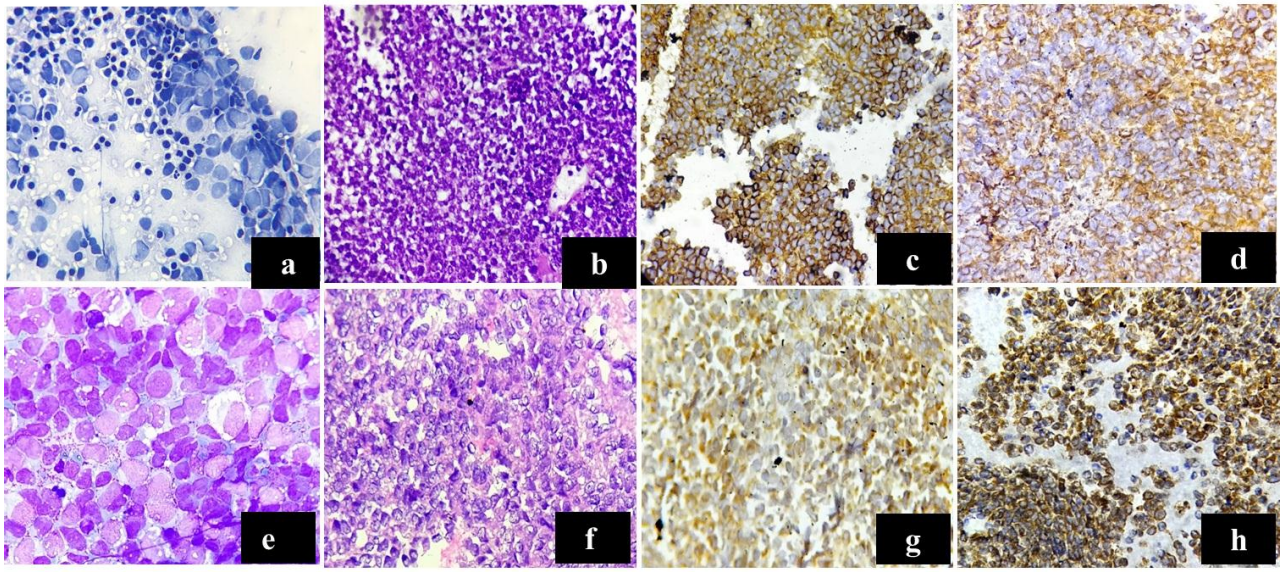


**Figure 2: (a) Cytology smear shows round to oval tumor cells seen singly and in clusters with background of scattered lymphocytes (Pap, 400X). (b) Section revealing sheets of tumor cells (Hematoxylin & Eosin, 400X). (c) Immunohistochemistry shows positivity for CD99. (d) Tumor cells show positivity for BCl2 (DAB, 400x). (e) FNA smear shows sheets of round to oval tumor cells (Giemsa, 400X). (f) Cell block section reveals sheets of tumor cells (Hematoxylin & Eosin, 400X), (g) Immunohistochemistry shows positivity for CD99. (h) Tumor cells are diffusely positive for BCl2 (DAB, 400x).**

## DISCUSSION

SS are malignant, high-grade, soft-tissue neoplasms and is more prevalent in adolescents and young adults 15 to 40 years of age with mild male predominance. Synovial sarcomas occur predominantly in the extremities, with the predilection for lower extremities followed by the head and neck region. It has also been described at various anatomical sites including heart, pleuropulmonary region, kidney, prostate, liver, mediastinum, retroperitoneum, gastro intestinal tract, etc.[2] SS involving palm and foot are rare[3],[4] and meagre case reports are there in the English literature.

SS have three main histologic patterns: Biphasic synovial sarcoma (BSS); monophasic synovial sarcoma (MSS); and poorly differentiated synovial sarcoma (PSS).[4] The BSS consists of epithelial and spindle cell components, whereas the MSS has uniform spindle cells and are difficult to distinguish from other spindle cell neoplasms such as fibrosarcoma, sarcomatoid squamous cell

carcinoma, malignant spindle cell melanoma, malignant solitary fibrous tumor, atypical carcinoid tumor, malignant peripheral nerve sheath tumor [MPNST], ES and leiomyosarcoma.[5] The PSS may mimic SRCT and ES is its closest differential;especially in palm and foot.[3]

Immunohistochemically, synovial sarcomas stains positively for Cytokeratin 7, 8/18, 19, AE1/AE3, EMA, CD99/O13, Vimentin and Bcl2. Transducer like enhancer of split 1(TLE-1) is a immunohistochemical marker found to be positive in 97% cases of synovial sarcoma and is a transcriptional corepressor that is overexpressed in synovial sarcoma.[6] PSS and other spindle cell malignancy can be differentiated on IHC. Fibrosarcoma characteristically does not show EMA immunoreactivity. Leiomyosarcomas show immunoreactivity for desmin and SMA. MPNST shows focal S100 immunoreactivity and ES shows PAS positivity and is negative for Bcl2. In ES, the tumor cells show a diffuse, complete, cytoplasmic membranous staining pattern for CD99, SS, in contrast shows cytoplasmic, granular to incomplete membranous positivity for MIC2/CD99.[6]

Cytogenetic and molecular genetic analysis is strong adjunctive method for confirming a diagnosis of SS. Chromosomal translocation t(X; 18)(p11.2;q11.2), which results in the expression of a chimera SYT-SSX transcript is considered as gold standard for diagnosis of SS.[7] However, molecular analysis was not done in our case due to unavailability and financial constraint.

Poor prognostic factor for SS include truncal as opposed to distal tumour location, lesions larger than 5cm, high histologic grade (based on the mitotic rate and tumour necrosis), neurovascular invasion, aneuploidy, poor histological differentiation and local recurrence.[8]

Multimodal combination of wide-to radical resection, radiation therapy and adjuvant chemotherapy following resection is the treatment of choice for synovial sarcomas.[9],[10]

## CONCLUSION

SS location in the palm of the hand and foot are exceedingly rare and lymph node metastasis is even rarer. To the best of our knowledge, no such case of lymph node metastasis from SS of foot has been reported in the English literature and our case highlights the same for the first time.

FNAC of Poorly differentiated SS can mimic as SRCT and differentiating it from ES poses a diagnostic challenge. However, ancillary testing with cell block and IHC prove valuable in resolving the diagnosis in such unusual presentations.

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