



## Myxoid synovial sarcoma: An unusual clinical and pathologic presentation

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### ABSTRACT

Myxoid Synovial sarcoma (SS) is a unique, uncommon, malignant variant of Synovial sarcoma with unusual clinical and pathologic presentation. Recognition of synovial sarcomas with massive myxoid feature is important because these can easily be mistaken for other myxoid soft tissue neoplasms. Here, we report a rare case of myxoid synovial sarcoma in a 28-year-old male who presented with slow growing localized tumor in the anterior aspect of his left forearm. This tumor was circumscribed and showed a well-defined capsule. The cut surface was gray, soft and myxoid. Histological examination showed proliferation of spindle cells in the predominantly myxoid stroma, including uniform spindled cells with fascicular growth patterns, collagenous stroma and hemangiopericytoma-like vascular patterns.

Small areas of glandular components were also obtained after many serial sections of the tumor, with features more typical of synovial sarcoma.

Our case strongly highlights that to make a correct diagnosis, clinical correlation and proper histomorphological examination along with immunohistochemical studies is essential.

**Keywords:** Synovial Sarcoma, Presentation, Soft Tissue, Myxoid

### INTRODUCTION

Synovial Sarcomas (SS) represent one of the rare soft tissue sarcomas, which typically arise near the knee and ankle joints of children and young adults. However, it is also known to occur around other joints, such as shoulder and hip.<sup>1</sup> Histologically, synovial sarcoma is either biphasic or monophasic. Focal myxoid change in synovial sarcoma is not uncommon. The presence of predominantly myxoid stroma has rarely been reported. This was not defined until 1999. Recognition of synovial sarcomas with marked myxoid feature is important because these can easily be mistaken for other myxoid spindle cell neoplasms.<sup>2</sup>

Because of the atypical morphology of cells, with their myxoid nature, they can be misdiagnosed as sarcoma especially Myxoid Liposarcoma, Extraskeletal Myxoid Chondrosarcoma or Hemangiopericytoma.<sup>3</sup>

In addition to these, the biphasic nature of this raises some diagnostic problems, especially when it arises in the extremities or on an unusual site. In such cases, carcinosarcoma and glandular malignant peripheral nerve sheath tumor (MPNST) are to be considered in the differential diagnosis.<sup>4</sup>

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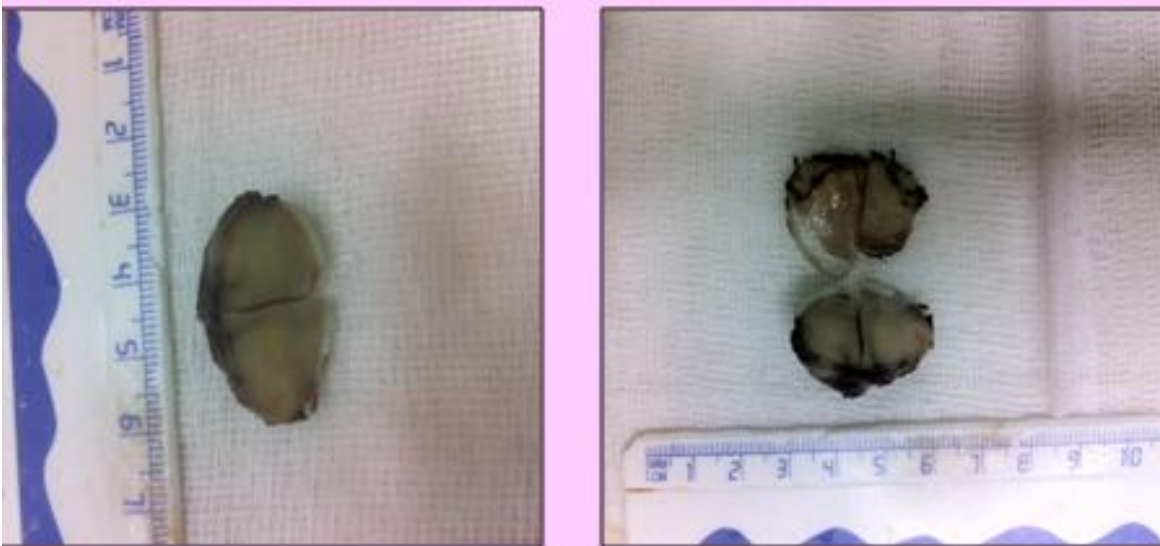
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### THE CASE

A 28-year-old male presented with a painless mass on the anterior aspect of his left forearm, which grew slowly in 2 years. His medical history was noncontributory. Physical examination revealed a firm, mobile, non-tender mass with smooth surface on the anterior aspect of the left forearm, with limitation of motion on clenching the hand. A working diagnosis of giant cell tumor was made clinically, by finding a movable lump in typical location of ganglion or lipoma, with less possibility of malignant mass due to its slow growth.

On wide excision, a 3 cm mass was found, covered with many vessels and located deeply within the tendon, with rounded regular border and smooth surface.

Grossly, the tumor measured 3x2x2cm, weighed 7 gms and was a well circumscribed, lobulated and capsulated oval mass (Fig. 1). The cut surface was grayish-white homogenous.



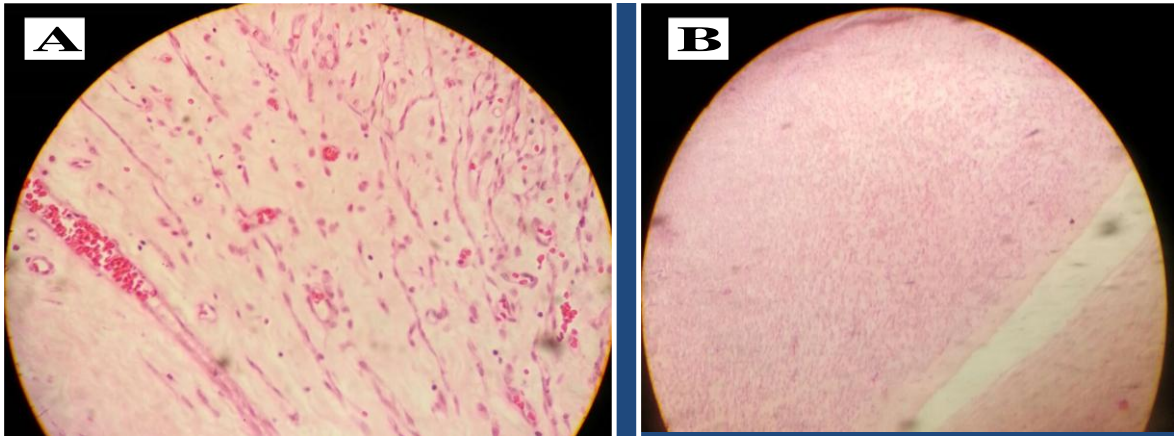
**Fig 1** An egg-size firm oval-shaped, gray-white solid mass and a thin obvious capsule were identified. Cut surfaces revealed whitish nodular solid and myxoid surface.

Histologically, the tumor showed atypical proliferation of spindle cells in the myxoid stroma. We found myxoid change in more than 60% of the sections. In the myxoid areas, spindled tumor cells had tapered nuclei and poorly-defined cytoplasm. The chromatin was finely dispersed. Well-defined capsule was noted around the tumor. (Fig 2)

A focal area of vascular Hemangiopericytoma-like pattern was noted. Mitotic count was 3 to 4 per 10

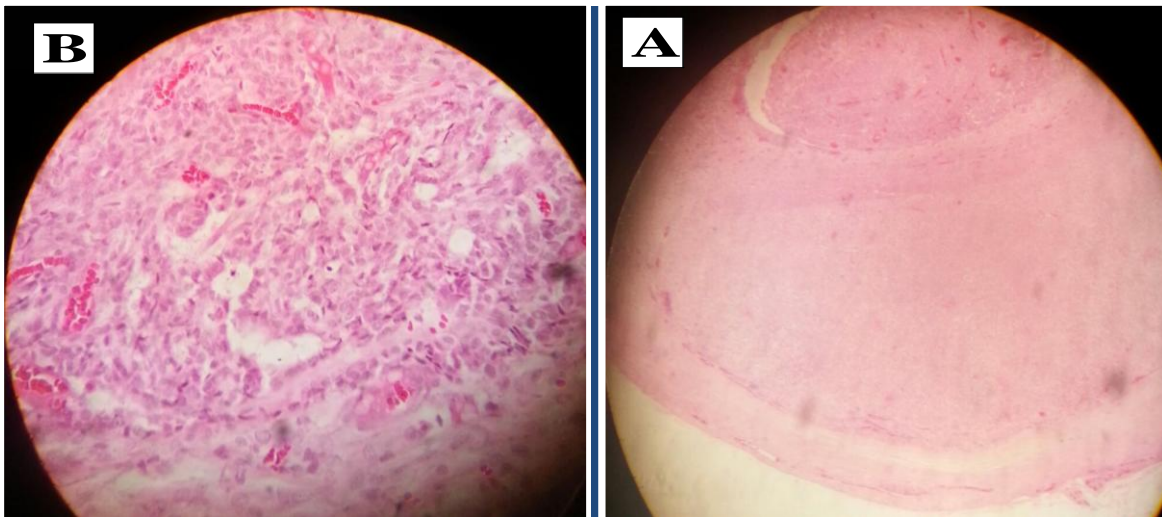
high-power fields, scrutinized in the most cellular areas. Further sections from the tumor showing focal glandular differentiation were focally observed. (Fig. 3) No necrosis or capsular invasion was detected.

The diagnosis was corroborated by a panel of Immunohistochemical (IHC) stains, which were performed in the regional laboratory.



**Fig 2 Synovial sarcoma, H&E Section**

- A. A Typical Proliferation of Spindle Cells in the Predominantly Myxoid Stroma**  
**B. Well Defined Capsule is Noted Around the Tumor with Obvious Cleavage**



**Fig 3 Synovial sarcoma, H&E Section.**

- A. Low Power Showed Focal Areas of Vascular Hemangiopericytoma-like pattern was noted in a Cellular Background**  
**B. Synovial Sarcoma exhibiting a classic biphasic spindle cells and gland-like appearance with intervening congested vascular channels**

### DISCUSSION

Synovial sarcoma represents the third most common soft-tissue sarcoma. The term myxoid synovial sarcoma was first described in 1999 by *Krane et al.* as a condition in which myxoid stroma constitutes greater than 50% of the studied surface area.<sup>3</sup> *Moffatt et al.* reported the cytological features of synovial sarcoma with predominantly myxoid stroma.<sup>5</sup> Serial sections of the tumor in study [Fig. 1]

show myxoid changes in more than 60% of the stroma.

The myxoid material in synovial sarcoma is of unknown origin. It has been suggested that myxoid material may be associated with a degenerative process or a product of stromal fibroblasts.<sup>2</sup>

### Age and Sex

This tumor is commonly seen among adults and has male preponderance. The age of the patient in this case was 28 years.

### Site of the Lesion

Synovial sarcoma arises adjacent to joints or tendon sheaths, with predilection for the lower extremities. Its histogenesis is unknown and is most certainly not a synoviocyte.<sup>6</sup> Reflecting a non-joint origin, less than 10% of synovial sarcomas are intra-articular.<sup>6</sup>

### Histopathology

Several histological types of synovial sarcoma are recognized: *biphasic* type, with distinct epithelial and spindle cell components in varying proportions; *monophasic* fibrous type; rare *monophasic epithelial* type and *poorly differentiated* (round cell) type.<sup>4</sup>

Myxoid areas are usually inconspicuous, but there are isolated reports indicating the possibility of a more abundant myxoid stroma.<sup>7</sup> Synovial sarcoma reveals initial painful symptoms due to compression. Therefore, it is crucial to combine imaging and pathology to establish the diagnosis.

Because of the relative chemosensitivity of synovial sarcoma, it is important that they be distinguished from other myxoid spindle cell sarcomas. The histologic differential diagnoses of synovial sarcoma with extensive myxoid feature include aggressive angiomyxoma, MPNST, myxofibrosarcoma, low grade fibromyxoid sarcoma, myxoid leiomyosarcoma, fibromyxolipoma, myxoid liposarcoma and myxoid malignant melanoma.

Generally, the use of ancillary techniques helps in correct diagnosis. A panel of immunohistochemical markers can be very useful in separating myxoid synovial sarcoma from other myxoid spindle cell tumors. In fact, myxoid synovial sarcoma is usually immunoreactive for cytokeratin or EMA and less often positive for S-100 and CD99. In addition, bcl-2 protein has been reported in 75-100% of synovial sarcomas, typically in a strong and diffuse fashion.<sup>2</sup> In particular, this neoplasm is characterized by the chromosomal translocation t(x;18)(p11;q110) and is

thought to be of multipotent mesenchymal cell origin.<sup>5, 8</sup>

### CONCLUSION

Myxoid synovial sarcoma is a rare variant of Synovial sarcoma with morphological features that mimic other myxoid spindle cell tumors, e.g. myxoid liposarcoma, extra skeletal myxoid chondrosarcoma and hemangiopericytoma.

This rare histologic variant of synovial sarcoma is important because it can easily be mistaken for other myxoid spindle cell neoplasms, potentially resulting in suboptimal therapy.

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