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Case Report

Pathology Section

A Tumour in Disguise in the Right Palm- Monophasic Synovial Sarcoma

VASUGI GRAMANI ARUMUGAM[1], SANDHYA SUNDARAM[2], ANITA RAMESH[3], PRATHIBA DUVURU[4], SRINIVASA RAJAPPA[5]

ABSTRACT

Synovial sarcoma is one of the poorly differentiated malignant soft tissue tumour occuring commonly among young adults in the extremities. We report a 50-year-old female presenting with a soft tissue mass in the right palm. On examination, a single firm and non tender swelling was noticed adjacent to the thenar muscles. Radiology suggested a benign soft tissue lesion. The swelling, clinically thought to be a lipoma, was excised and sent for histopathological examination. Microscopy showed a highly cellular tumour arranged in nests, cords and pseudo glandular pattern separated by dense fibrocollagenous tissue. An interesting and baffling finding was the presence of a distinct mucin vacuole in many of the tumour cells. A diagnosis of soft tissue sarcoma with epithelial features was considered and a panel of immunohistochemical stains done. Tumour cells showed strong positivity for cytokeratin 7, vimentin, EMA & Bcl2. CD 99 and S100 were focally positive. CD 34 and CEA were negative. In view of the above microscopic and immunohistochemical findings, a diagnosis of monophasic synovial sarcoma of epithelial type was rendered. This case is being documented for the rare morphological appearance of mucin vacuoles in a monophasic epithelial type synovial sarcoma.

Keywords: Epithelial type, Malignant soft tissue tumour, Mucin vacuoles

CASE REPORT

A 50-year-old female presented with a soft tissue swelling in right palm for one year duration. She had no other associated symptoms like pain, difficulty in moving fingers or other co-morbidities. On examination, it was a firm, non tender swelling located adjacent to the thenar muscles between the ring and middle finger of right hand [Table/Fig-1,2]. Movements were not restricted. X-ray revealed a soft tissue shadow with scalloping of metacarpals. MRI was also done and reported as being suggestive of a benign soft tissue lesion. The swelling clinically thought to be a lipoma was excised and sent to histopathology.

Grossly, it was multiple grey white soft tissue bits amounting to 5x3x1 cm. The cut surface was grey white to grey yellow. Microscopy showed a cellular tumour arranged in nests, cords and pseudoglandular pattern separated by dense fibrocollagenous tissue [Table/Fig-3]. Individual cells were plump with pleomorphic nuclei with vesicular chromatin.

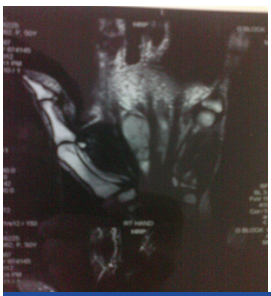
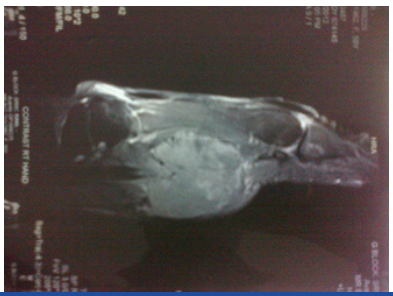
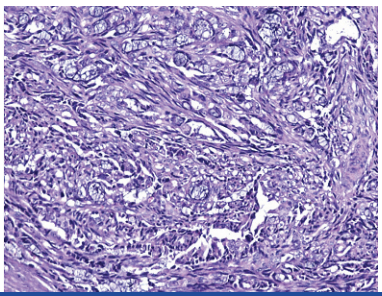
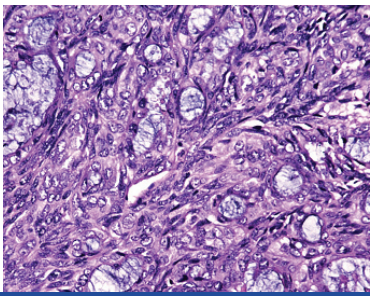
Cytoplasm was moderately abundant and eosinophilic [Table/Fig-4]. An interesting finding was the presence of distinct mucin vacuoles in many of the tumour cells which was better appreciated by periodic acid Schiff special stain [Table/Fig-5]. Scattered mitotic figures were seen. A diagnosis of soft tissue sarcoma with epithelial features was considered and a panel of Immunohistochemical markers was done for categorization.

In view of the above microscopic and immunohistochemical findings [Table/Fig-6], a diagnosis of monophasic synovial sarcoma of epithelial type with mucin vacuoles was rendered. After the initial excision our patient has completed 5 cycles of chemotherapy and is doing well. Follow up at the end of one year did not show any recurrence or metastasis.

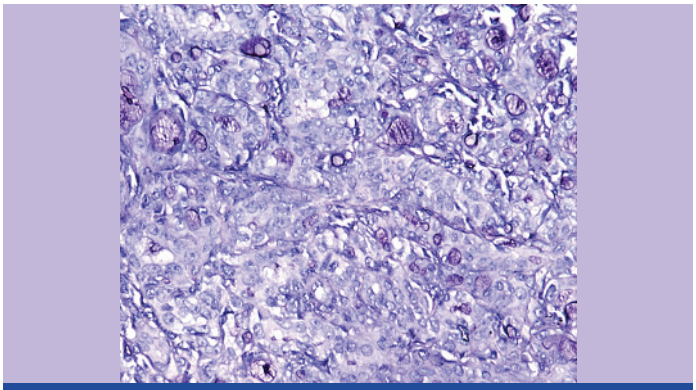
DISCUSSION

Synovial sarcomas are deep seated soft tissue tumours usually seen in young adults with a male preponderance. The most common presentation is that of a palpable, deep seated swelling or mass associated with pain or tenderness. Synovial sarcomas occur predominantly in the extremities followed by the head and neck region. Most of the tumours are firmly attached in and around tendon or joint capsule [1]. It has also been described at various anatomical sites including heart, pleuropulmonary region, kidney, prostate, liver, mediastinum, retroperitoneum, gastro intestinal tract, etc. Our case presented as an innocuous soft tissue mass in the right palm over the thenar muscles.

Presence of tumour in upper extremities, constitutes approximately 10% to 15% of all cases and are distributed among the forearm-wrist region, shoulder, elbow-upper arm region, and hand. A total of 107 cases of synovial sarcoma presenting in hand has been reported by Lotz et al., in a study [2].

[Table/Fig-1]: MRI coronal view- soft tissue mass lesion seen in the plane between the flexor tendon sheath of ring and middle finger. **[Table/Fig-2]:** MRI contrast- Thinning and scalloping of the third and fourth metacarpals. **[Table/Fig-3]:** Cellular tumour with cells arranged in pseudoglandular pattern H&Ex40. **[Table/Fig-4]:** Cellular tumour with mucin vacuoles H&Ex100.



[Table/Fig-5]: PAS positivity in the mucin vacuoles within tumour cells x200.

Cytokeratin-7 [Table/Fig-7], Vimentin, EMA [Table/Fig-8]	Strongly positive
Bcl-2 [Table/Fig-9]	Diffusely positive in tumour cells
CD 99, S-100	Focally positive
CD 34, CEA	Negative
Ki 67 labelling index [Table/Fig-10]	40%

[Table/Fig-6]: Panel of Immunohistochemical markers done and inference.

On radiology, most synovial sarcomas appear as round or oval lobulated swellings or masses of moderate density. The most striking radiological feature found in 15-20% of synovial sarcomas is the presence of multiple small, spotty radiopacities caused by focal calcification and less frequently bone formation [3]. The present case showed thinning and scalloping of the third and fourth metacarpal bone, however calcification was not observed. Grossly, it is a well circumscribed firm, grey white to yellow lesion completely or partially covered by a smooth glistening pseudocapsule.

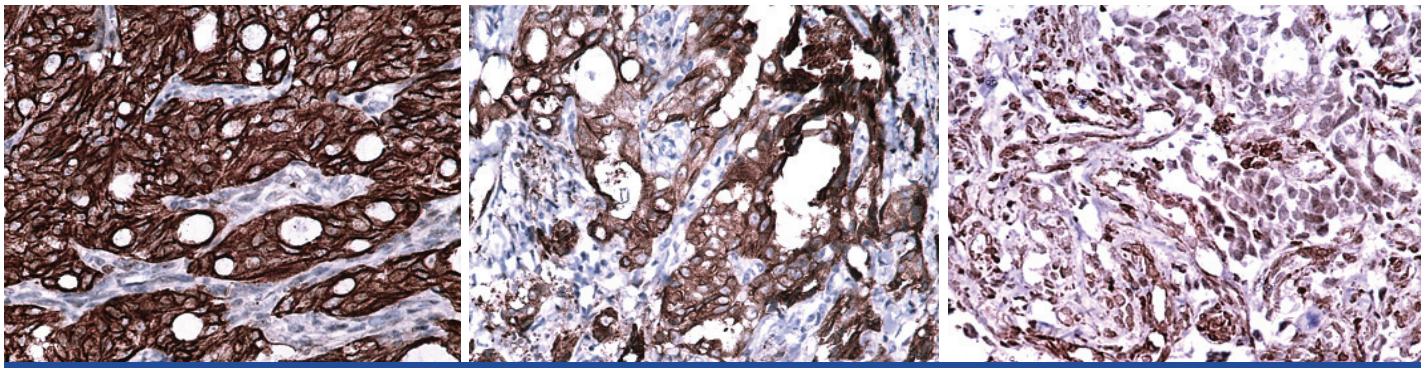
According to the prominence of the cellular elements and the degree of differentiation, histologically, they are divided into the biphasic type, with epithelial and spindle cell components, monophasic epithelial type, monophasic fibrous type and poorly differentiated (round cell) type [4].

The present tumour was a monophasic epithelial type synovial sarcoma which is a rare variant. Additionally, it also showed mucin vacuoles in the cytoplasm. Therefore a differential diagnosis of metastatic mucin secreting adenocarcinoma had to be ruled out. Thorough investigations revealed no primary mucin secreting tumour elsewhere in the body. Other important differential considerations were malignant melanoma, and adnexal tumours. Epithelioid mesenchymal tumours should also be considered, including epithelioid sarcoma and epithelioid MPNST. However immunohistochemical staining with positivity for cytokeratin, Epithelial Membrane Antigen (EMA) and diffuse Bcl2 positivity was more in favour of a synovial sarcoma. Mucinous, gland in synovial sarcoma must be recognised to avoid misdiagnosis of metastatic carcinoma [5].

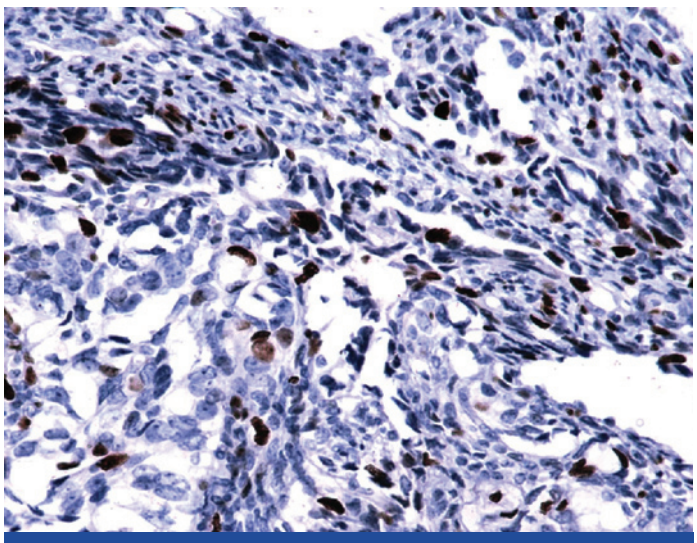
Immunohistochemically, synovial sarcomas stains positively for Cytokeratin 7, 8/18, 19, AE1/AE3, EMA, CD99/O13, Vimentin (spindle cells), Bcl2 [6]. TLE-1 is a recent immunohistochemical marker found to be positive in 97% cases of synovial sarcoma [7].

Translocation t(X;18)(p11;q11) : SYT-SSX1 genes are detected in 90% cases. Also, t(X;18)(p11.21;q11) : SYT-SSX2 fusion genes may also be an associated finding. P16/INK4A gene deletion is seen in 74% of them. RT-PCR is highly sensitive than FISH in detecting the SYT: SSX1/ SSX2 fusion genes [7].

The gold standard diagnostic test for synovial sarcoma is by demonstrating the Translocation {t(x:18), (p11.2,q11.2)} by fluorescence in situ hybridization, reverse transcriptase polymerase chain reaction, or cytogenetics. TLE1 encodes a transcription



[Table/Fig-7]: IHC for CK showing strong positivity x200. **[Table/Fig-8]:** Tumour cells show strong immunopositivity to EMA x200. **[Table/Fig-9]:** IHC showing diffuse immunopositivity to Bcl-2 x200.



[Table/Fig-10]: Ki-67 staining showing high proliferative index x200.

factor that is found to be overexpressed in synovial sarcomas. Hence TLE1 is considered as a bio-marker for distinguishing synovial sarcoma from other soft tissue malignancies by gene and tissue microarray studies [8].

Good prognostic factors include young age, tumour size less than 5 cm, complete resection with clear surgical margins, presence of mast cells microscopically and response to first line chemotherapy. On the other hand, male gender, 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 are considered as poor prognostic factors [9].

Synovial sarcoma is an aggressive malignant tumour; the 5-year survival rate hasbeen reported to be approximately 70% to 80%, and the 10-year survival rate is 50%.

Recurrence has been reported in about 8% to 60% within 2 years after the first treatment. Multimodal combination of wide-to radical resection, radiation therapy and adjuvant chemotherapy following

resection is the treatment of choice for synovial sarcomas. Follow up is mandatory because of the high incidence of recurrence [10-12]. Synovial sarcomas may metastasize to bone, liver, skin, brain, adrenals and even breast tissue [13,14].

Authors	Reference	Patient details	Histologic type	Prognosis
Khandeparker SS Gaurish S, Avinash J et al.,	[15]	30/F, swelling right palm	Monophasic synovial sarcoma spindle cell type	Good, no recurrence one year follow-up
Laila C, Ikram B et al.,	[16]	23/M, Painless cystic nodule left hand	Monophasic synovial sarcoma spindle cell type	Good, no recurrence one year follow-up
Casal D, Isabel A et al.,	[17]	63/F, Painful firm mass right hand	Monophasic synovial sarcoma Fibrous variant	Died of recurrence after 12 years
Harjai MM, Bal RK, et al.,	[18]	6/M, Slow growing mass left hand	Synovial sarcoma with predominantly epitheloid pattern	Good, No recurrence due to favourable prognostic factors
Present case		50/F, Painless mass right palm for one year	Monophasic synovial sarcoma epitheloid type with mucin vacuoles	Good, no recurrence one year follow-up

[Table/Fig-11]: Similar case reports in literature.

Similar case reports of synovial sarcoma presenting in hand reported in the literature have been tabulated in [Table/Fig-11].

CONCLUSION

The case has been documented to highlight the behavior of this rare variant of monophasic synovial sarcoma epithelial type with mucin vacuole in the palm of hand. Knowledge of the varied histological features of synovial sarcomas will prove useful in making a precise diagnosis and understanding the biology of this interesting tumour.

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PARTICULARS OF CONTRIBUTORS:

1. Post Graduate, Department of Pathology, Sri Ramachandra Medical College and Research Institute, Chennai, India.
2. Professor, Department of Pathology, Sri Ramachandra Medical College, Chennai, India.
3. Professor, Department of Oncology, Sri Ramachandra Medical College and Research Institute, Chennai, India.
4. Professor, Department of Pathology, Sri Ramachandra Medical College and Research Institute, Chennai, India.
5. Associate Professor, Department of Orthopedics, Sri Ramachandra Medical College,Chennai, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Sandhya Sundaram,
2A, Abirami Meaddowland, 32 Park Road, Anna Nagar West, Padi, Chennai-600050, India.
E-mail: sandsrid@gmail.com

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