ISSN: 2690-9189

Research Article

International Journal of Orthopaedics Research

A Case Report of A Rare Tumor In Rare Site

Nalla Ambhika[1], Ranganath Ratnagiri[2], Megha Uppin[3] and Sujata Patnaik[4]*

[1]*MD, Junior Resident Department of Radiology, NIMS, India*

[2]*MS, MCh, Additional Professor Department of Surgical Oncology, NIMS, India*

[3]*MD, Additional Professor Professor of Pathology, NIMS, India*

[4]*MD, Professor of Radiology, NIMS, India*

***Corresponding Author**
Dr. Sujata Patnaik MD, Professor of Radiology, NIMS, India.

Submitted: 2024, Aug 29; **Accepted:** 2024, Sep 25; **Published:** 2024, Oct 02

Citation: Ambhika, N., Ratnagiri, R., Uppin, M., Patnaik, S. (2024). A Case Report of A Rare Tumor In Rare Site. *Int J Ortho Res, 7*(3), 01-05.

Abstract

Synovial sarcoma is a rare soft tissue tumour occurring in young ages. Most common site is extremities close to joints. Thoracic wall, chest and trunk are rare sites. Presence of calcifications, hemorrhage, solid, cystic / necrotic component and triple sign in MRI are characteristic. This is a case report of synovial sarcoma in the right supraclavicular fossa extending along chest wall. Its radiological features are described.

Keywords: Synovial sarcoma, chest tumour, rare tumours.

1. Introduction

Synovial sarcomas are rare soft tissue tumors derived from pluripotent mesenchymal cells [1]. Most of them present at 3rd to 5th decade with extremities being the most common site, usually close to the joints. Trunk, chest wall and thorax account for 7% cases [2]. Histologically there are three types- monophasic, biphasic and poorly differentiated. Monophasic type commonly presents as painless palpable mass. Synovial sarcomas are predominantly intermediate to high grade in their potential for metastatic spread. Imaging plays an important role; though not diagnostic, gives clues for narrowing down the differentials and highlights the aggressive nature of mass. It helps in surgical planning and selecting a biopsy site. Less than 10 cases of chest wall synovial sarcomas are reported in literature [3]. Because of such rarity of these tumours with its rare site of occurrence, we are reporting this case with its imaging features and reviewed the literature.

2. Case Report

A 35-year-old female presented with swelling on right side of neck close to right clavicle extending along the chest wall for about 3 months. It was associated with pain that increased on working and radiating to right upper limb. There was no history of loss of appetite or weight. She was not hypertensive or diabetic. There was no history of tuberculosis or asthma or any significant family history. On examination, the general condition was fair; she was afebrile with pulse rate of 74 per minute and blood pressure-116/70 mm Hg. Systemic examination was remarkably normal. On local examination the mass was 5x4 cms and firm; tensely cystic swelling felt behind the lateral border of right pectoralis muscle. Fullness was noted over right upper chest. The clinical diagnosis was soft tissue tumor. Hemoglobin was 8 gm/dl. Other blood tests and the PT, and APTT were normal. On fine- needle aspiration cytology (FNAC), it was reported as an epithelial neoplasm.

On chest X-ray, a vague soft tissue opacity was appreciated in right supraclavicular fossa and along right upper chest wall. There was no evidence calcification on radiograph (Figure 1).

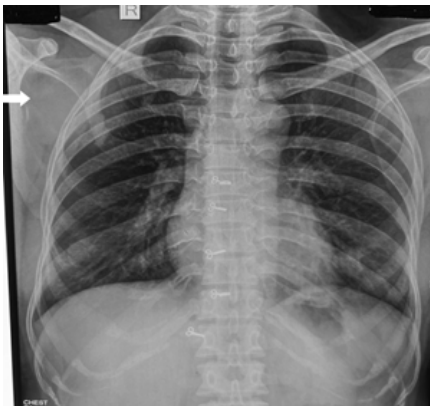

Figure 1: Chest radiograph reveals soft tissue opacity along supraclavicular fossa and along upper chest wall on right side

Following FNAC, MRI was done. Subsequently CT scan was performed for better evaluation. On CT chest, the mass was well defined oblong shaped heterogenous mass of 7x 3.5cm with areas of hypodensity and isodensity (similar to muscle). The lesion involved the Serratus anterior muscle and was on its medial aspect along the chest wall. There was minimal heterogenous enhancement. Foci of punctate calcification were appreciated within the lesion (Figure 2A and Figure 2B) and there were no bone erosions. On MRI, T1W the lesion was predominantly isointense to muscle, with foci of hyperintensity (Figure 3A) within representing hemorrhage. On T2W there was triple sign of low, Intermediate and high signal intensity with fluid level (Figure 3B). On contrast heterogenous enhancement with thick enhancing septa (Figure 3C) was noted. The possibility of malignant mesenchymal tumor was entertained.

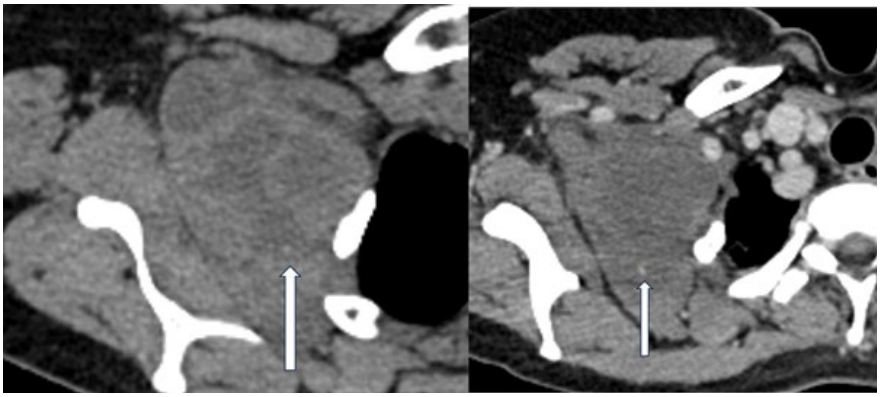

Figure 2A: Axial CT chest non-contrast scans show heterogenous mass of iso and hypodensity with foci of calcification indicated by arrow.

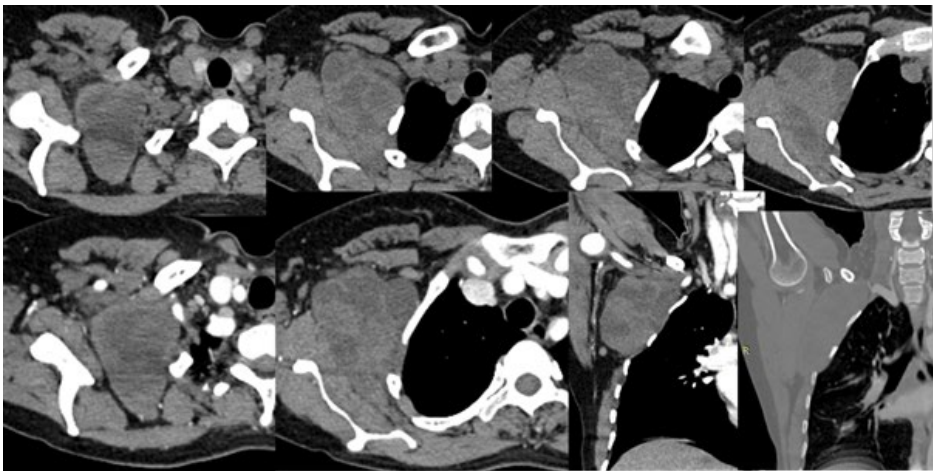

Figure 2B: Axial CT chest and coronal reconstructed images in contrast enhanced scans in soft tissue and bone window images show heterogenous enhancement and lesion in involving Serratus anterior. There is no bone erosion

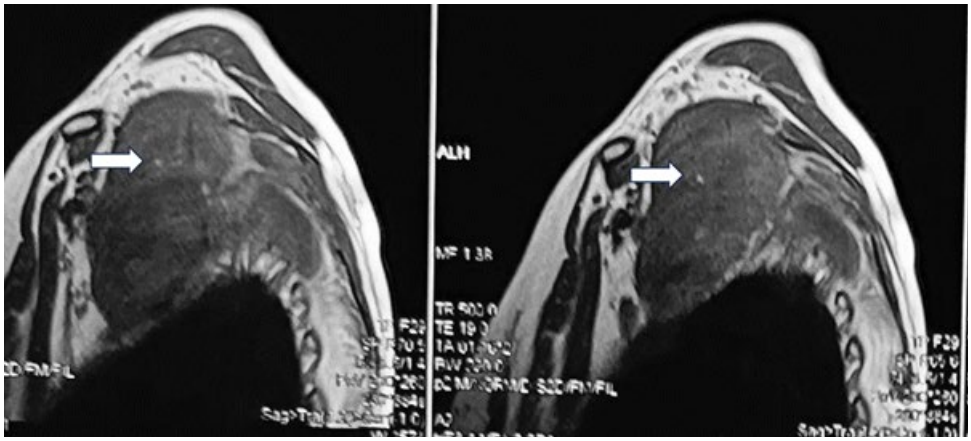

Figure 3A: Sagittal T1W image depicts, the hyperintense foci representing haemorrhage in background of mildly hypointense mass

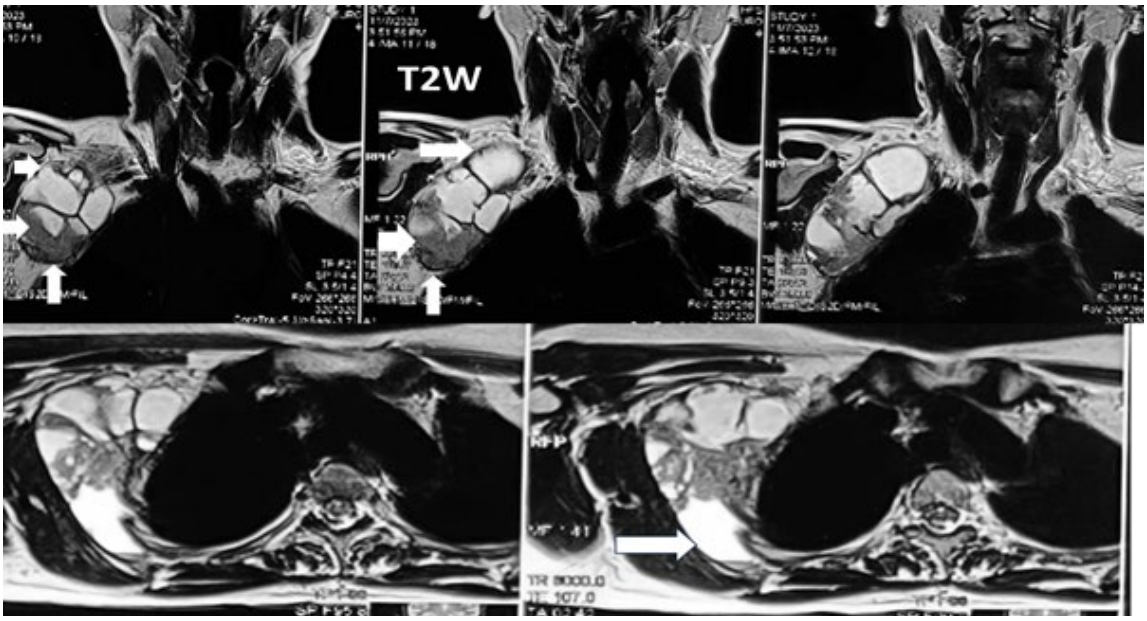

Figure 3B: Coronal and Axial T2w image clearly delineates triple sign (arrow) with fluid level

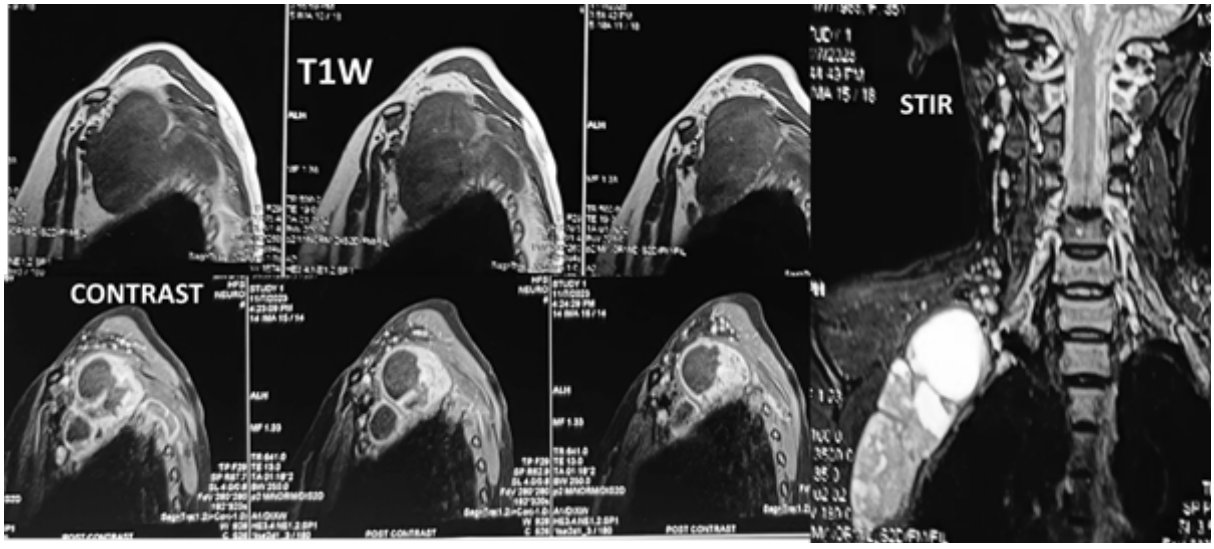

Figure 3C: Both plain and contrast enhanced sagittal T1W images show intense heterogenous enhancement. Fluid- fluid level is depicted in coronal stir image

Patient was operated and intraoperative findings were the mass arising from Serratus Anterior muscle extending from right supraclavicular region to right axilla, chest wall between intercostal muscle and subscapularis. The mass measured 9x7x3.5cms. A few enlarged nodes in right axilla with largest one -1.5x1cm at level 1 were noted. Nerve to Serratus anterior was sacrificed. Sections studied from the soft tissue mass showed a well circumscribed lesion. It comprised of spindle cell and epithelial cell components in varying proportions. The spindle cell component showed cells arranged in sheets and intersecting fascicles. The cells were spindle shaped with elongated, pleomorphic vesicular nuclei and moderate amount of eosinophilic cytoplasm with blunt ends. The epithelial component showed cells arranged in solid nests, sheets, cords and glands. The lesional cells were polygonal to columnar with round to oval pleomorphic vesicular nuclei with moderate amount of pale eosinophilic cytoplasm. Mitosis was frequent and was accounting for 8-10/10 HPF. Necrosis was not identified. Pericapsular fibroadipose tissue showed mild inflammatory infiltrate comprised of lymphocytes and plasma cells (Figure-4). One of the resected margins was involved by the tumour. 15 lymph nodes were isolated from the separately sent specimen labelled as axillary lymph nodes of which 2 lymph nodes showed evidence of metastasis (2/15). The final diagnosis was *Synovial Sarcoma, Biphasic, pT3 N1*.

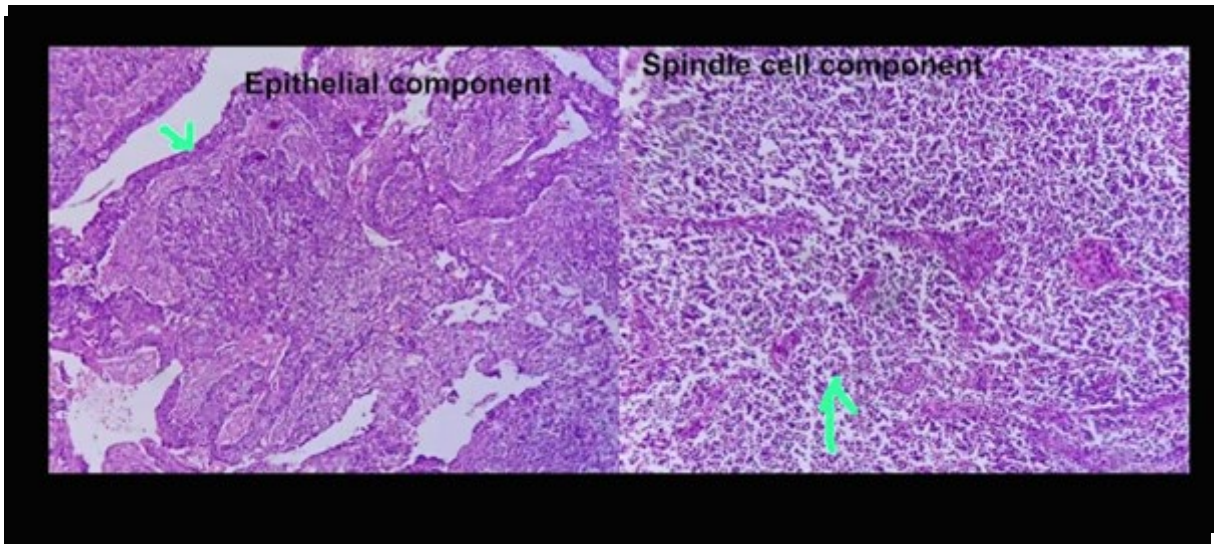

Figure 4: Histopathological Examination-Spindle cell component show cells arranged in sheets and intersecting fascicles indicated by green arrow). The cells are spindle shaped with elongated pleomorphic vesicular nuclei and moderate amount of eosinophilic cytoplasm with blunt ends. Epithelial component shows cells arranged in solid nests, sheets cords and glands (indicated by green arrow

3. Discussion

Synovial sarcoma is a misnomer as it does not arise from synovium; it resembles synovial tissue in light microscopy. It arises from as yet unknown multi potent stem cell that are capable of differentiating mesenchymal and epithelial structure and lack of synovial differentiation [4]. These tumours occur in young age. They are commonly seen close to the joints. Extremities are common sites. Less common sites are head, neck, trunk, thorax, chest wall, pelvis, retroperitoneum. Chest wall, axilla are extremely rare and is a challenge for surgical approach. According to Arco CD et al, only 5 cases of biphasic synovial sarcoma of axilla were reported previously [5].

Radiograph is normal when the tumor is small or when situated in complex areas such as in pelvis or is deep seated. Larger lesions show nonspecific findings. Calcification is seen in 30% cases [6]. These calcifications are eccentric and peripheral; rarely chondroid or osseous mineralisation is seen. Erosion of underlying bone is rare and is seen in 5% of cases [6]. Ultrasonography can evaluate the shape, margin, vascularity and composition of tumours. It also helps in biopsy. These lesions are nodular, lobulated solid mass. Heterogeneity may be seen due to necrosis, haemorrhage. On CT scan, the mass appears as solid soft tissue with attenuation similar to muscle or slightly hypodense. Areas of lower attenuation due to haemorrhage, necrosis as in our case. Smaller lesions are homogenous. Small number of cases are cystic predominantly. On contrast there is heterogenous enhancement. Margins may be well defined or irregular. Calcifications and bone erosions are better depicted on CT. The mass observed in our patient was well defined all round except medially and was inseparable from Serratus anterior and abutting the Subscapularis and intercostal muscles. On MRI the lesions are T1-heterogenous lobulated with signal similar to or slightly more than muscle. On T2, it is heterogenous. Triple sign was described by Jones [7]. Low intermediate and high signals are due to fibrous tissue, solid cellular component and haemorrhage /necrosis. Triple sign is seen in 25-57% cases [7]. It was also observed in our case. MFH also demonstrates triple sign. Areas of haemorrhage appear as high signal on T1W images or demonstrates fluid level. Margins are well defined in majority of cases and ill-defined irregular in 9-47% cases [7]. As in neurogenic tumour *split fat sign* is observed when

the tumour arises in intermuscular plane. On contrast there is heterogenous enhancement. Sometimes there are thick septate (>3mm) showing enhancement. Prognosis of synovial sarcoma is poor and five -year survival is 38-76% [8]. There are certain features which indicate bad prognosis- absence of calcification, more cystic component with haemorrhage, necrosis with triple sign and size of tumour >5cms.

The tumor in our case, after excision was 9 x 6 x 3.5cms in size and on HPE was biphasic and one of resected margin is involved by tumor. In general prognosis is poor in synovial sarcoma. Size >5cms, biphasic histopathology and positive resected margin were additional bad prognostic features in our case. Lungs, lymph nodes and bones are the common sites of metastases. Lung nodules shows calcification. Surgery is the treatment of choice. Wide local excision of mass or amputation are surgical options depending on situation. Chemotherapy is given for residual or metastatic disease. Radiation is important for microscopic positive margins. With treatment the size of mass decreases but there is increase in necrosis. Oedema surrounding the tumour may appear with treatment.

4. Conclusion

Synovial sarcoma is a rare soft tissue tumour occurring in young persons. Most common site is extremities close to joints. Thoracic wall, chest and trunk are rare sites. In our patient the tumour was in a rare site. Presence of calcifications, cystic component and triple sign on MRI are characteristic features. Though rare, synovial sarcoma should be considered in differential diagnosis of any soft tissue mass. More cases have to be studied for better characterisation of this rare lesion.

References

1. Gopalakrishnan, V., Amini, B., Wagner, M. J., Nowell, E. N., Lazar, A. J., Lin, P. P., ... & Araujo, D. M. (2017). Synovial sarcoma of the head and neck: a single institution review. *Sarcoma, 2017*(1), 2016752.
2. Deshmukh, R., Mankin, H. J., & Singer, S. (2004). Synovial sarcoma: the importance of size and location for survival. *Clinical Orthopaedics and Related Research®, 419,* 155-161.
3. Fekih, L., Boussoffara, L., Fenniche, S., Abdelghaffar, H., Akrout, I., Ayadi, A., & Megdiche, M. L. (2011). Rare primary chest wall sarcoma: the synovialosarcoma. *Revue des Maladies Respiratoires, 28*(5), 681-685.
4. Ruggiero, A. (2004). Synovial sarcoma. *Orphanet Encyclopedia March.*
5. Díaz del Arco, C., & Fernández Aceñero, M. J. (2017). Biphasic axillary synovial sarcoma diagnosed by preoperative fine-needle aspiration cytology. *Diagnostic Cytopathology, 45*(9), 857-860.
6. Kransdorf, M. J., & Murphey, M. D. (2006). *Imaging of soft tissue tumors.* Lippincott williams & wilkins.
7. Jones, B. C., Sundaram, M., & Kransdorf, M. J. (1993). Synovial sarcoma: MR imaging findings in 34 patients. AJR. *American journal of roentgenology, 161*(4), 827-830.
8. Bergh, P., Meis-Kindblom, J. M., Gherlinzoni, F., Berlin, Ö., Bacchini, P., Bertoni, F., ... & Kindblom, L. G. (1999). Synovial sarcoma: identification of low and high risk groups. *Cancer, 85*(12), 2596-2607.