

CASE REPORT

HUGE SYNOVIAL SARCOMA ARISING FROM CHEST WALL: A RARE CASE REPORT

Waddi Sudhakar¹, B. Venkata Rao², Abburi Srinivas³, S. R. Harshavarsan Majety⁴, M. Madhulika⁵

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ABSTRACT: Synovial sarcomas are the fourth most common malignant soft-tissue tumors, and typically develop in para-articular locations of the extremities. However, the occurrence of these tumors in the chest wall is rare. In this article, we report the interesting case of a 27-year-old male with spindle cell variant of synovial sarcoma arising in the anterior chest wall with a brief review of the literature.

KEYWORDS: Synovial sarcoma; chest wall; spindle cell variant.

CASE REPORT: A 27 year old male patient presented with swelling in the chest wall and anterior aspect of neck for the last one year. Except for disfigurement patient had no other symptoms. On clinical examination a 20x15 cm irregular shaped swelling over anterior aspect of chest and neck, with nodular surface and engorged veins over it. The swelling is nontender, having variable consistency and with restricted mobility.(Figure-1)

X-ray of the chest and neck showed soft tissue mass lesion.

CT scan of the chest and neck showed large subcutaneous mass from infra hyoid level to pre sternal region up to carina with a) sternal lytic lesion with anterior mediastinal invasion b) bilateral level II,III lymphadenopathy. Possibilities: 1) lymphoma 2) malignant teratoma with metastatic lymphadenopathy. (Figure-2)

True cut biopsy from the swelling suggestive of cellular soft tissue tumour mass of spindle cell type possibility of a) fibroma b) neurofibroma. Intra operatively the tumour is extending into the superior mediastinum so enblock resection of tumour along with manubrium sternum and lymph nodes done under general anesthesia. (Figure-3)

Post-operative HPE showing cellular soft tissue tumour mass with spindle cells having elongated nuclei arranged in irregular fascicles showing mild nuclear atypical features. Mitotic figures average 3-4/10 HPF. Cystic spaces with papillary projections are seen. Vascularity is prominent with medium sized irregular blood vessels associated with haemorrhages tumour shows irregular infiltrative margin, resected margins are free from tumour tissue. Features suggestive of low grade spindle cell variant of synovial sarcoma. (Figure-4) On immunohistochemistry, the tumor cells were positive for vimentin, CD99, Bcl2 and S 100 protein. These findings were consistent with synovial sarcoma of chest wall.

The postoperative course was uneventful. An adjuvant chemotherapy was prescribed but the patient was lost for review.

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DISCUSSION: Synovial sarcomas are rare malignant neoplasms of unknown histogenesis, most commonly affecting the lower extremities and frequently arises adjacent to joints or tendon sheaths.

Synovial sarcoma is a misnomer because the tumor does not arise from the synovium; it only resembles synovial tissue at light microscopy.⁽¹⁾ Synovial sarcomas rarely involve the chest wall. Synovial sarcomas constitutes approximately 5-14% of soft tissue sarcomas in different studies.^(2,3)

A part from the extremities, synovial sarcoma may arise within head and neck, esophagus, retroperitoneum and also in the thorax; mediastinum, heart, lung, pleura or pericardium with lesser frequency. Synovial sarcomas rarely involve the chest wall and there are Less than 10 cases were reported in the literature.⁽⁴⁻⁹⁾ There is a mild male predominance, the sex ratio is 1.5. It affects preferentially young patients.⁽¹⁰⁾ The symptoms of synovial sarcoma of the chest wall depend on the structures undergoing compression or invasion from the tumor. Patients may present with chest pain, cough, dyspnea, reduced breath sounds and weight loss.⁽⁶⁾

At computed tomography, synovial sarcoma that arises in the chest wall is characterized most commonly as a heterogeneously enhanced mass with well-defined margins, cortical bone destruction, tumor calcifications and tumor infiltration of the chest wall musculature.⁽¹¹⁾ Mediastinal, hilar, diaphragmatic or axillary lymph nodes are rarely involved.

Macroscopically, tumors are round or multilobular, poorly or well circumscribed. They may be either unencapsulated or surrounded by a thin fibrous capsule.⁽¹²⁾ On section, they were grey-yellow and exhibited a rather variegated appearance with cyst formation, hemorrhage, and necrosis. Gross calcification may be evident.

Synovial sarcoma is composed of two morphologically different types of cells: epithelial cells, resembling those of carcinoma, and fibrosarcoma-like spindle cells. Depending on the relative prominence of the two cellular elements and the degree of differentiation, synovial sarcomas form a continuous morphologic spectrum and can be broadly classified into the (I) biphasic type, with distinct epithelial and spindle cell components in varying proportions; (II) monophasic fibrous type; (III) rare monophasic epithelial type and (IV) poorly differentiated (round cell) type.⁽¹³⁾ Almost all morphological subtypes are characterized by a specific t(X;18) (p11.2; q11.2) chromosomal translocation.

Like other soft tissue sarcomas, synovial sarcoma's diagnosis is difficult to establish purely on the basis of histological appearance. It is even difficult in some cases without an obvious biphasic differentiation. Thence, immunohistochemical studies must be completed showing neoplastic cells diffusely immunoreactive to CK, EMA, Vimentin, Bcl-2, Actin and CD99 with focal immunoreactivity for S-100 protein and are negative for CD34 and Desmin.⁽¹⁴⁾

Treatment of choice of synovial sarcoma of the chest wall as in all soft tissue sarcomas is multimodal combination of wide-to radical resection, radiation therapy and adjuvant chemotherapy following resection and since synovial sarcoma is known to recur, a careful follow up is mandatory.^(5,6)

Synovial sarcomas might metastasize to bone, liver, skin, the central nervous system, and even breast tissue.⁽¹⁵⁾

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Prognosis is related to the disease stage and is usually poor. Young age, Her-2 expression, complete resection with clear surgical margins and response to first line chemotherapy were found as good prognostic indicators in advanced disease in different studies.⁽²⁾ In the other hand, adverse prognostic factors for synovial sarcoma include male gender, truncal as opposed to distal tumor location, lesions larger than 5 cm, high histologic grade (based on the mitotic rate and tumor necrosis), neurovascular invasion, aneuploidy, poor histological differentiation and local recurrence.⁽³⁾

CONCLUSION: Synovial sarcoma is a mesenchymal spindle-cell tumor characterized by variable epithelial differentiation. Its chest wall localization represents a diagnostic challenge because of the diverse array of competing diagnosis and rarity of incidence. Owing to its rarity and the paucity of data regarding its natural history, there are no guidelines for optimal treatment. Meanwhile, it consists on surgical resection associated to chemotherapy and/or radiotherapy.



Fig. 1: preoperative photograph



Fig. 2: CT scan showing superior mediastinal extension of the tumour

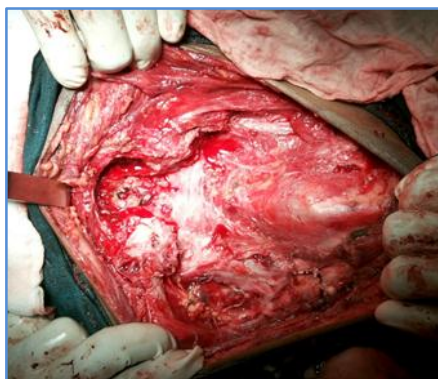


Fig. 3: Intraoperative photo en-bloc resection of the tumour along manubrium sternum

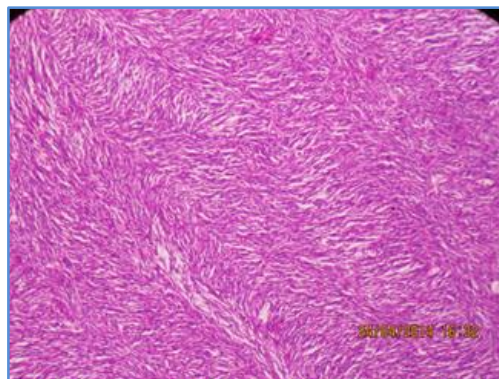


Fig. 4: HPE showing spinle cells

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AUTHORS:

1. Waddi Sudhakar
2. B. Venkata Rao
3. Abburi Srinivas
4. S. R. Harshavarsan Majety
5. M. Madhulika

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of General Surgery, A. M. C.
2. Assistant Professor, Department of General Surgery, A. M. C.
3. Post Graduate Student, Department of General Surgery, A. M. C.
4. Post Graduate Student, Department of General Surgery, A. M. C.

5. Post Graduate Student, Department of General Surgery, A. M. C.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Waddi Sudhakar,
Assistant Professor,
Department of General Surgery,
King George Hospital,
Andhra Medical College, Visakhapatnam.
E-mail: sudhakarwaddi@yahoo.co.in

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