

Synovial Sarcoma -A Very Rare Chest wall Tumor

Dr. Rajendra Prasad Potlabathini¹, Dr. Srinivasan Somalwar²

¹Associate Professor, ²Assistant Professor, Osmania Medical College, Hyderabad. Telangana state, India

*Corresponding author

Dr. Srinivasan Somalwar

Email: somalwar73@gmail.com

Abstract: An 18 Year old girl presented to the thoracic surgery department with a left sided chest wall swelling of 10 years duration. On physical examination the lesion appeared to be a benign lesion. The patient was investigated and she underwent resection of the swelling. The Histopathology report of the resected specimen revealed it to be a synovial sarcoma. Synovial sarcoma of the chest wall is extremely rare, with only few cases described in literature.

Keywords: Synovial sarcoma, chest wall, Histopathology

INTRODUCTION

Among primary tumors of chest wall, cartilaginous tumors, Desmoids tumors, and Fibrous dysplasia are more common [1]. Synovial sarcomas account for approximately 6% of thoracic tumors and commonly involve joints of the arm, neck or leg. They occur rarely on thoracic wall and have been reported as few occasional case reports in literature [1, 3-7]. We hereby describe a case of synovial sarcoma originating from back of the chest wall.

CASE REPORT

An 18 Year old lady was admitted to the thoracic surgery department with a chest wall swelling (fig1) of size 12x6 cms on the left poster lateral thoracic wall. She was relatively asymptomatic for the last 10 years and presently complained of pain over the swelling since the last 6 months. There was no history of trauma to the chest wall. Skin over the swelling was pinch able and side to side mobility was present but restricted. There was no vertical mobility. Size of the swelling decreased on contraction of serratus anterior and latissimus dorsi muscles. The swelling was firm in consistency on palpation. Vitals signs were stable, there was no pallor. Local or generalized lymphadenopathy was absent. Lung fields on Chest X-ray were normal. (Fig-2) Computed Tomography Scan chest (Fig-3)

showed a well-defined heterogeneous soft tissue density mass 7x3.4 cm lesion in the posterior thoracic wall deep to the latissimus dorsi and inferior to the serratus anterior muscles. Intra thoracic extension and calcification were absent. A diagnosis of benign spindle cell tumor was made on a pre-operative FNAC. The patient underwent surgery after taking informed consent. En block surgical resection with 5 cm free circumferential tumor margin was done under general anesthesia.

Histopathology report: Macroscopically the tumor had a firm feel, was well circumscribed and cut section showed grayish white appearance with tiny cystic areas measuring 1 cm to 1.8 cms.

Multiple sections taken from the cystic swelling revealed a well circumscribed tumor comprised of spindle cells arranged in short fascicles and interlacing bundles. Focal peritheliomatous arrangement of cells was noted. Individual cells were spindle to elongate with mildly pleomorphic plump ovoid nuclei with vesicular chromatin and prominent eosinophilic micro nucleoli. Mitotic activity was sparse. Stroma was scanty with multiple cystic spaces (fig 8, fig 9 and fig 11).



Fig – 1



Fig-2



Fig-3



Fig-4



Fig -5



Fig -6



Fig -7

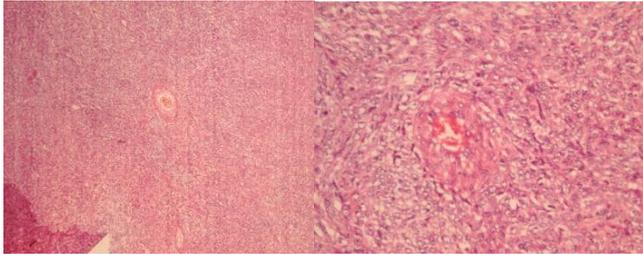


Fig-8

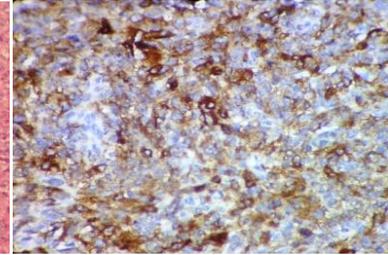


Fig -9

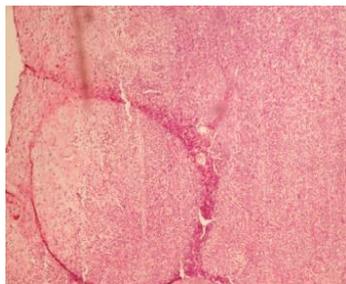


Fig-11

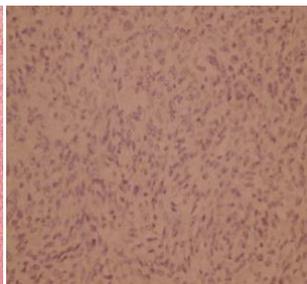


Fig-12

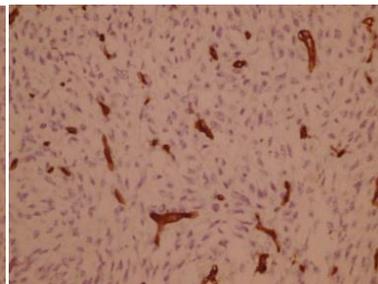


Fig-13

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Pancytokeratin (fig 10), EMA, S100, Vimentin, Bcl 2 staining showed cytoplasmic positivity in tumor cells.

CD 99 showed strong membrane positive staining. Desmin (fig 13) was negative in tumor cells. Ki 67 labeling index was 3% (fig 12).

POST OPERATIVE RADIO THERAPY

1 month after the surgery Patient was put on local radiotherapy to the back of the chest with a of dose 200 CG (2Gy)fractions per day for 30 days amounting to a total dose of 6000 CG (60Gy) over 6 weeks to avoid local recurrence.

POST OPERATIVE FOLLOWS UP:

Patient was reviewed every6 months after the surgery for detection of local and distant metastases by Chest X Ray and chest CT scan. There was no evidence of local and distant metastases for the first 2 years after surgery.

DISCUSSION:

Synovial sarcoma (also known as: malignant synovioma [1] is a rare form of cancer which usually occurs near the joints of the arm, neck or leg. The name "synovial sarcoma" was coined early in the 20th century, as some researchers thought that its microscopic similarity to synovium, and its propensity to arise adjacent to joints, indicated a synovial origin; however, the actual cells from which the tumor develops are unknown and not necessarily synovial [2]. Primary synovial sarcomas are most common in the soft tissue near the large joints of the arm and leg but have been documented in most human tissues and organs, including the brain, prostate, and heart. Synovial sarcoma represents about 8% of all soft tissue sarcomas [3] and about 15-20% of cases occur in adolescents and young adults [4]. The peak incidence is before the 30th birthday and male to female ratio is 1.2:1. Macroscopically they have non-specific appearances. They are well or poorly defined heterogeneous masses with frequent areas of hemorrhage and necrosis. They are divided histologically into the following four sub types: biphasic (20-30%), monophasic fibrous (50-60%),

poorly differentiated (15-25%) and monophasic epithelial (very rare). Cytogenetic aberration of the t(X;18) translocation is highly specific and is seen in over 90% of cases [1-4]. Treatment of choice of synovial sarcoma of the chest wall, as in all soft tissue sarcomas, is a multimodal combination of wide-to radical resection, radiation therapy and adjuvant chemotherapy following resection. Since synovial sarcoma is known to recur, a careful follow up is mandatory [5]. Synovial sarcomas might metastasize to bone, liver, skin, the central nervous system, and even breast tissue. Prognosis is related to the disease stage and is usually poor [6]. Young age, Her-neu 2 expression, complete resection with clear surgical margins and response to first line chemotherapy were found to be good prognostic indicators in advanced disease in different studies [7]. On 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.

CONCLUSION

Synovial sarcomas of chest wall are very rare tumors. In our case the tumor size was >5cm, histologically it was a low grade tumor with no local lymph nodes spread or distant metastases. The TNM Classification was T2N0M0 hence wide local excision of tumor and adjuvant post-operative local Radiotherapy was given. A careful follow up every 6 months for 2 years was done. No evidence of local and distant metastases was detected.

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