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Evans Tumor: A Rare Case Report.

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ABSTRACT

Sarcomas of the chest wall are uncommon. Here we present a rare case of low grade fibromyxoid sarcoma in a 44 year old male with swelling in the chest wall since childhood. Fine needle aspiration cytology revealed features suggestive of chondroid lesion. Wide local excision of the lesion was done and sent for histopathology which was reported as low grade fibromyxoid sarcoma (Evans tumor). The patient was discharged and the postoperative period was uneventful.

Keywords: Sarcoma, chest wall, Evans tumor

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Case Presentation

Here we present a 44 years old male admitted to the surgical department with the complains of swelling over left upper chest wall, just below left shoulder. Swelling was present since childhood, and it increased from the size of a peanut to the size of a cricket ball over past 6 months. There was no history of fever, pain over swelling or history of any restriction of movement of left upper limb.

On examination, there was 8x8x4 cm hemispherical swelling in the left anterior chest wall occupying deltopectoral groove. Upper border was just below lateral end of clavicle. Lower border extended up to the roof of axilla. Laterally, the swelling extended up to medial part of upper third of arm. Swelling was non tender, firm to hard in consistency, smooth surface, not fixed to skin and mobility appeared restricted on contracting pectoralis major muscle.

The following investigations were done:

Fine needle aspiration cytology was suggestive of benign chondroid lesion.

Chest X-ray and USG Abdomen showed no demonstrable metastases.

MRI – Well defined heterogenous soft tissue mass found adherent to the pectoral fascia- suggestive of soft tissue tumour.

A wide local excision was planned. Intraoperatively, tumour was found to involve superficial fibres of pectoralis major muscle and hence tumour was excised in toto with the entire left pectoralis major muscle and sent for histopathological examination. Primary closure of the wound after placing drain was done. Post operative period was uneventful. Post operatively adjuvant radiotherapy was given.

Histopathology

Well encapsulated nodular grey white tumour measuring 10x10x6 cm involving underlying fascia and not infiltrating muscle bundle. Cut margins and deep margins were free from tumour invasion. Multiple sections studied from tumour shows hyper and hypo-cellular areas of bland spindle cells in myxoid background.

Suggestive of Low Grade Fibromyxoid Sarcoma.

DISCUSSION

Low Grade Fibro-Myxoid Sarcoma (LGFMS) is a distinctive type of fibrosarcoma. It is a rare tumor whose exact incidence is not known. It is common in young and middle aged males in fourth decade of life. It is a malignant soft tissue neoplasm. Despite being benign histologically it has high potential of metastasis. This tumor was first reported by *Harry Evans* in 1987 – hence known as **Evan's Tumour.** Its cellular origin is unknown but presumed to be a mesodermal derivative. Exact etiology is unknown and there are no known risk factors.

It is usually a painless slow growing mass occuring in trunk and proximal extremities. It can occur rarely in chest wall, head and retroperitoneum. Majority occurs in subfascial deep locations. In pediatric populations it tends to be smaller, superficial and easier to resect. It typically has a long indolent course. Possibility of local recurrence and distant metastasis is very high. Distant metastasis can occur even decades after initial presentation, more commonly in lungs. etiology is unknown and there are no known risk factors.

Two Sub-types are recognised:

- Classical low grade fibromyxoid sarcoma
- Low grade fibromyxoid sarcoma with giant collagen rosettes

It is difficult to diagnose on fine needle aspiration cytology or even on core needle biopsy. It is usually detected in excision biopsy of the tumor. Histologically it is composed of bland spindle shaped cells in a whorled pattern arranged in alternating myxoid and collagenized areas. Immunohistochemically thses tumors are positive for vimentin, CD 34 and negative for desmin SMA and S100. More than 90% exhibit balanced translocation between t (7:16), resulting in a fusion gene FUS/CREB3L2.

Imaging:

MRI – Investigation of choice. It determines vascularity, relation to vessel and fascial planes. CT usually done to rule out lung secondaries.

Wide surgical excision is the treatment of choice. Amputation can be considered when extremity is involved. Radiotherapy is given in post operative period to prevent local recurrence and distant metastasis.

Follow-up

Long term follow up is vital because metastasis can occur after a long interval – as long as 40 years. In a recent study, local recurrence was noted in 54%, metastases in 6% and death from disease in 2%[2]. But no study till date has recommended a protocol for follow-up.

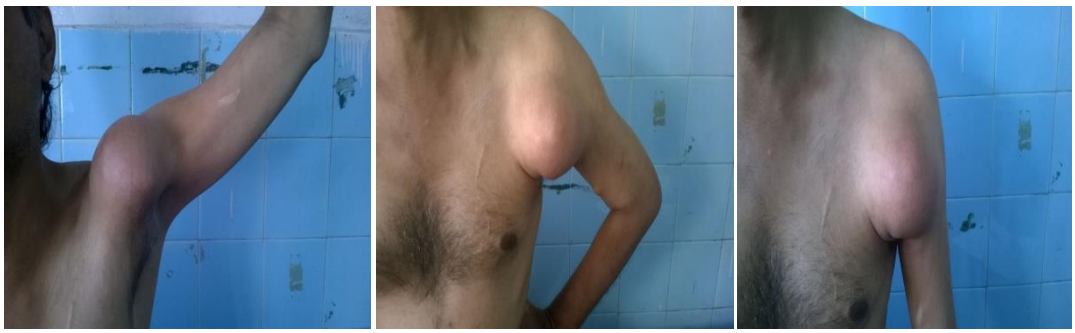

Figure 1: Clincial photograph showing soft tissue swelling arising from the chest wall

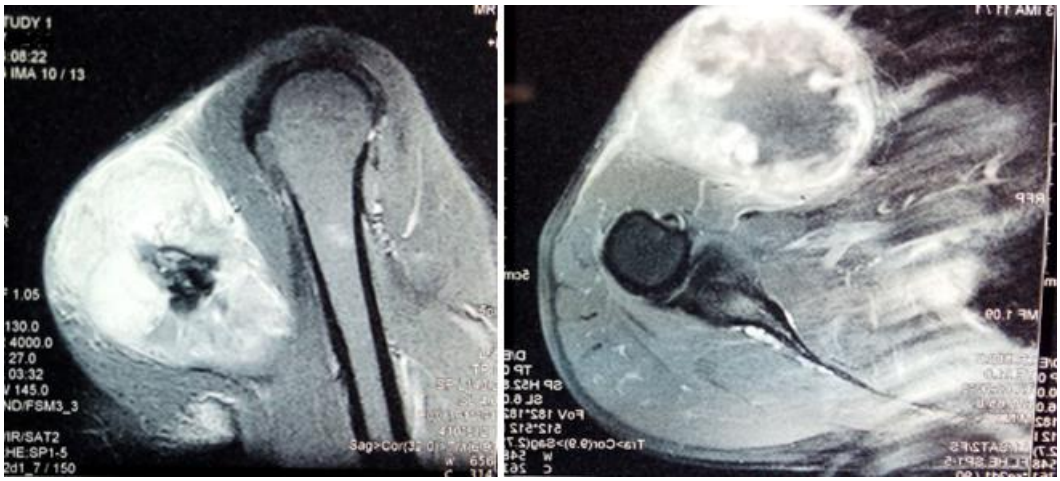

Figure 2: MRI – Well defined heterogenous soft tissue mass found adherent to the pectoral fascia, suggestive of soft tissue tumour

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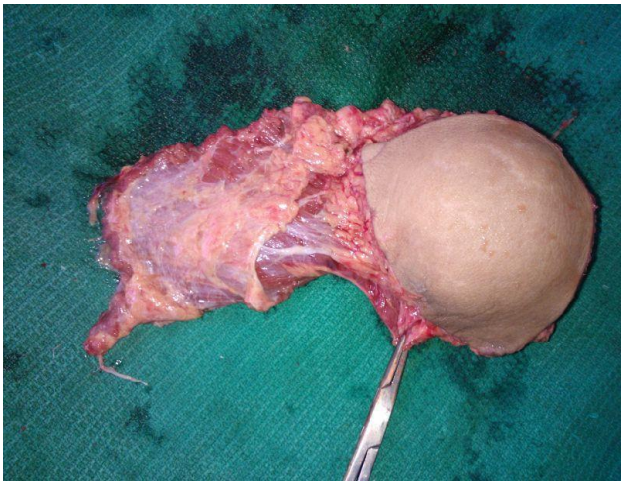

Figure 3: Excised specimen

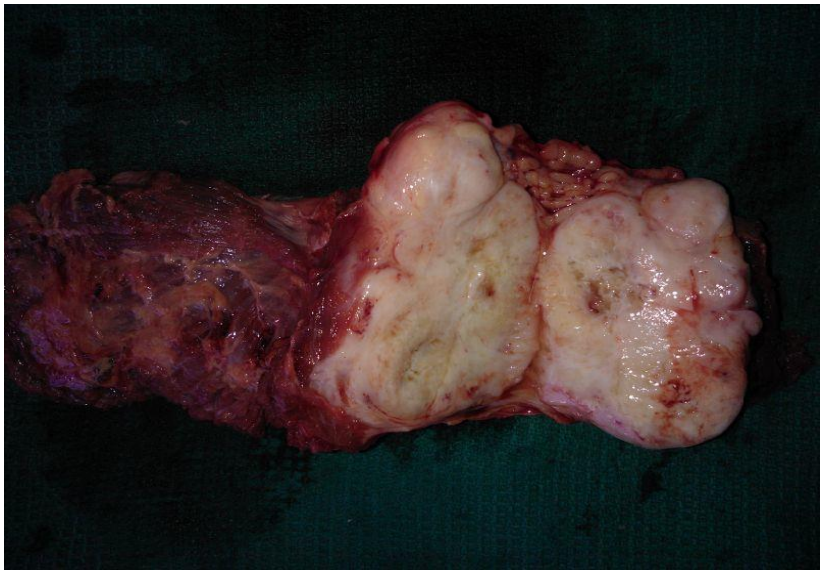

Figure 4: Cut section of the tumor showing a circumscribed grey white mass

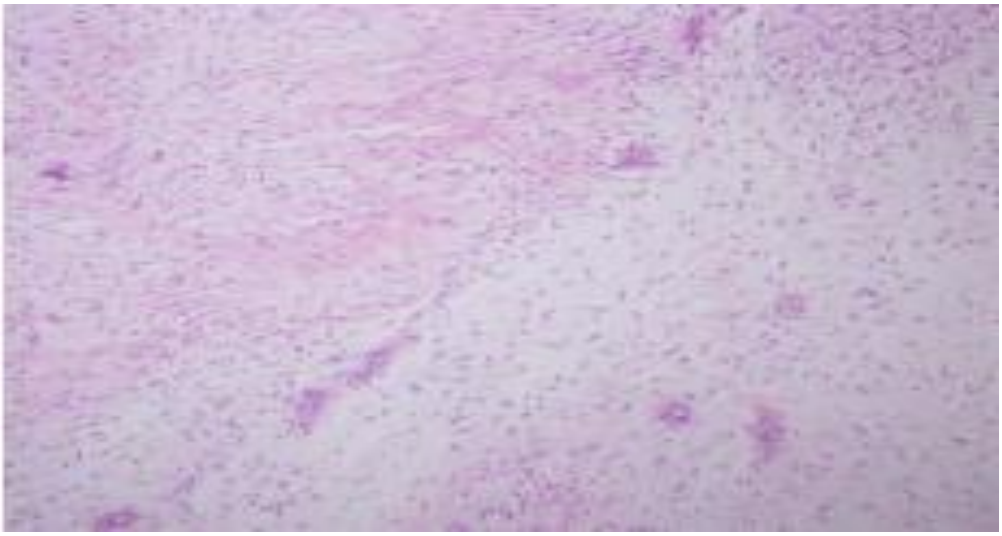

Figure 5: H&E showing hyper and hypocellular areas of bland spindle cells in a myxoid background

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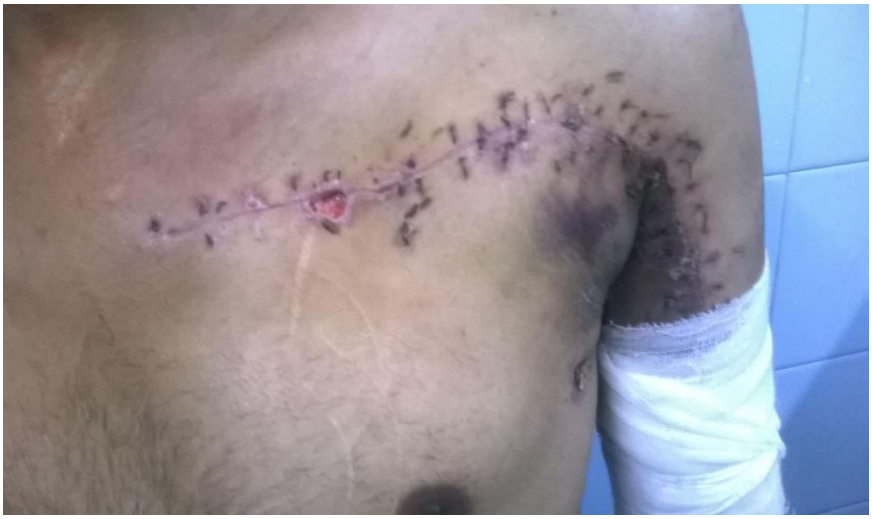

Figure 6: Post operative photograph

REFERENCES

[1] Evans HL. Am J Clin Pathol 1987; 88:615.
[2] Wu X, Petrovic V, Torode IP, et al. Pathology 2009; 41(2): 155-60
[3] Arnaoutoglou C, Lykissas MG, Gelalis ID, et al. J Orthop Surg Res 2010; 5: 49.
[4] Goodlad JR, Mentzel T, Fletcher CD. Histopathology 1995; 26(3): 229-37.
[5] Billings SD, Giblen G, Fanburg-Smith JC. Am J Surg Pathol 2005;29(2):204-10