

Case Report

Low grade fibromyxoid sarcoma (Evans tumour) of the arm

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ABSTRACT

A low-grade fibromyxoid sarcoma is a rare soft tissue tumour that has a tendency to develop in the deep soft tissue of young adults with potential for local recurrence or distant metastasis. We describe a 40-year-old female patient with a low-grade fibromyxoid sarcoma of the shoulder that had been excised twice in the past and then had recurred after a few months. A wide resection of this tumour with flap reconstruction was performed followed by radiation to the area. The patient had no evidence of local recurrence or distant metastasis at 2 years after surgery.

KEY WORDS

Evans tumour; low-grade fibromyxoid sarcoma; shoulder

INTRODUCTION

Low grade fibromyxoid sarcoma (LGFMS) is an uncommon tumour and was described first by Evans in 1987 and hence is also known as Evans tumour. We treated this unusual tumor in a 40 year old lady with wide excision and local reconstruction using a latissimus dorsi flap followed by radiotherapy which is the recommended treatment for this condition.

CASE REPORT

A 40-year-old female, presented with a large swelling over the left shoulder since 6-8 months. It was associated with pain and foul smelling discharge from the swelling. There was a history of the rapid increase in the size

of the swelling over 2 months. The swelling had been excised twice in the past, 20 years ago the first time, and 17 years back before recurring at the same site presently. No histopathology details of the prior excision were available.

Local examination revealed a tender, firm mass of 16 cm × 6 cm × 10 cm overlying the left shoulder and upper arm with areas of necrosis and serosanguinous discharge. The mass was bleeding on touch.

The surrounding skin was macerated. The scar from a previous surgery was seen over the left shoulder [Figures 1 and 2]. The shoulder girdle examination was within normal limits. There was no evidence of axillary lymphadenopathy. Systemic examination was normal.

Magnetic resonance imaging (MRI) examination revealed a 17 cm × 6 cm × 10 cm. Tumour in the subcutaneous plane superficial to the deltoid muscle. It was a well-circumscribed, multilobulated, solid mass. The mass was predominantly hypointense on T1 and moderately hypointense on T2. Prominent vascular channels were observed within the tumour [Figure 3].

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10.4103/0970-0358.138973



Figure 1: Necrotic tumour with scar of previous surgery



Figure 2: Areas of haemorrhage and necrosis on the tumour

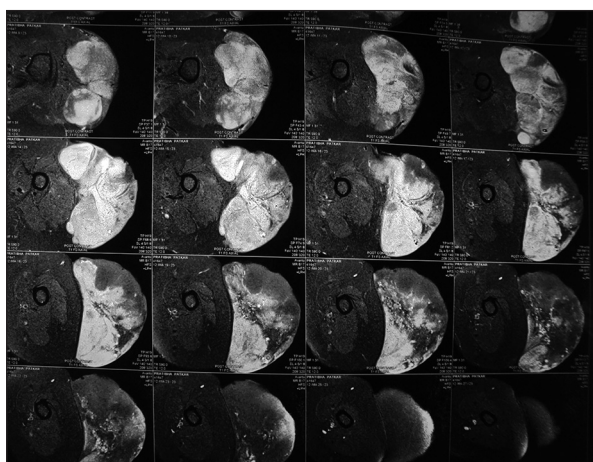


Figure 3: Magnetic resonance imaging scan showing tumour superficial to the deltoid muscle

A wide local excision of the mass with frozen section confirmation that the margins and base were free of tumour; primary reconstruction with a latissimus dorsi musculocutaneous flap based on thoracodorsal pedicle was performed.

Grossly, the excised tumour was a well-circumscribed, lobulated round, and firm mass. The cut surface was yellowish-white and fibrous to myxoid with areas of haemorrhage and necrosis. The histopathology report stated that it was a low-grade fibromyxoid sarcoma (LGFMS), which has assumed a high-grade character in multiple areas of the tumour.

The optical microscopy examination demonstrated a spindle cell tumour composed of hypo to moderately cellular areas. The tumour showed an arcuate branching capillary meshwork. The tumour cells were stellate to spindle-shaped and separated by myxoid stroma in the hypocellular areas. Areas of infarction were noted. There were areas of hypercellularity, which are high grade and border on round cell morphology [Figure 4].

Tumour cells were immunonegative for S-100 protein, CD34, smooth muscle actin (SMA), desmin, calponin, cytokeratin (CK), epithelial membrane antigen (EMA), and glial fibrillary acidic (GFA).

Patient was given radiotherapy (6000 cGy in 30 fractions over 44 days) to reduce the chances of local recurrence which was well-tolerated. She has been recurrence free at the end of 2 years [Figure 5].

DISCUSSION

Low grade fibromyxoid sarcoma has been defined as a cytologically bland malignant neoplasm with alternating fibrous and myxoid stroma of low-grade/low malignant potential.^[1] This uncommon entity was described first by Evans.^[2,3] Since then many cases have been reported of this rare tumour in the paediatric population.^[4]

The tumours vary in size from 1 to 23 cm.^[5,6] The median size of the tumour was 9.4 cm as studied.^[6] These tumours are classically situated in the trunk and lower extremities. The most common tumour locations were the shoulder area, thigh, and inguinal area. However, some tumours have been rarely noted in other areas like the mesentery and intracranial location.^[7,8] There is a case of LGFMS in the thyroid gland.^[9] The Aarhus Sarcoma Registry found a 64% greater propensity of LGFMS among females.^[10]

These tumours show alternating fibrous and myxoid stroma. Grossly, the tumour appears as a circumscribed mass, but microscopic infiltration may be present. Tumour cells are small, with scanty eosinophilic cytoplasm, round to ovoid

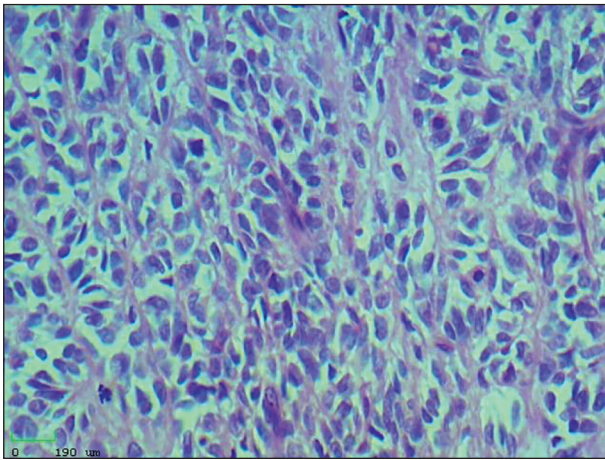


Figure 4: H and E, $\times 100$

nuclei and absent nucleoli. Although, focal cytologically atypical areas of high cellularity, increased mitotic activity, nuclear hyperchromatism, and necrosis may be found, tumour cells are usually characterised by absent to sparse mitotic figures, nuclear anaplasia, or necrosis.^[11]

On immunohistology, the staining is positive for vimentin and MUC4, and negative with antibodies, such as desmin, keratin, S-100 protein, EMA, CD34, and CD31. In our case, the pathology report suggested the tumour with mixed fibrous and myxoid areas and hypocellular stroma which is classical of LGFMS. The immunohistochemistry report suggested that the tumour cells were immunonegative for S-100 protein, CD34, SMA, Desmin, Calponin, CK, EMA, and GFAP. In a clinical study at the Aarhus sarcoma centre in Denmark, the investigators used the FUS break apart gene translocation testing using fluorescent *in situ* hybridisation technique. However, they found no difference in the clinical characteristics or outcomes associated with the FUS gene. Neither was it associated with change in the local recurrence nor metastases. We did not conduct the test on our patient due to cost constraints.

Imaging studies particularly MRI aid in corroborating the diagnosis and the extent of the tumour. Hwang *et al.* studied imaging modalities such as conventional radiography, ultrasound (US), computed tomography (CT) and MRI in 29 patients.^[12] They evaluated anatomic site of lesion, size, number, US echogenicity, CT attenuation, calcifications and MRI signal intensity pattern with contrast enhancement. They concluded that LGFMS was commonly single at initial diagnosis and multiple at local recurrence. In a case report by Miyake *et al.*, a 10-year-old boy with shoulder LGFMS



Figure 5: Postoperative healed wound with latissimus dorsi flap

found MRI corroboration with the tumour pathology.^[13] The myxoid areas showed findings of high signal intensity on T2-weighted images, whereas the hypercellular areas showed low signal intensity on T2-weighted images. Our patient's MRI showed the tumour superficial to the deltoid region, and the contrast MRI showed extreme vascularity of the tumour.

The recommended treatment at the Aarhus Sarcoma Centre in Denmark is wide surgical excision. Margins were called as marginal if the resection included the pseudocapsule or wide if the resection included a cuff of normal tissue.^[14] Local recurrences were treated with surgery without adjuvant therapy. However, distant metastases were treated with chemotherapy agents including ifosfamide, doxorubicin, imatinib, trabectedin, gemcitabine, docetaxel, as well as palliative radiotherapy. Trabectedin yielded the best response.

Differential diagnosis of LGFMS includes lesions showing spindle cell proliferations with myxoid pattern with or without fibrous component.^[15] The entities with predominantly myxoid pattern without significant fibrous component include myxomas, low-grade myxofibrosarcoma, angiomyxomas, myxoid liposarcoma, and a myxoid neurofibroma. Tumours with mixed myxoid and fibrous morphologies include neurofibroma, fibromatosis, perineurioma, malignant peripheral sheath tumour, and fibrous histiocytoma. Additional entities that should be encountered are desmoid tumour, desmoplastic fibrosarcoma, and low-grade differentiated liposarcoma.

As these tumours are known to metastasise after a long interval, sometimes after as many as 45 years, a thorough clinical follow-up is recommended. However,

no study has till date recommended any protocol for the follow-up. Once a diagnosis of LGFMS has been made a thorough oncological evaluation including observing the chest carefully is advisable as chest metastases are the most common.

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How to cite this article: Indap S, Dasgupta M, Chakrabarti N, Agarwal A. Low grade fibromyxoid sarcoma (Evans tumour) of the arm. *Indian J Plast Surg* 2014;47:259-62.

Source of Support: Nil, **Conflict of Interest:** None declared.

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