

Myxofibrosarcoma, in the calf of a middle aged female: a case report

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Abstract

Myxofibrosarcoma belongs to the group of sarcoma tumours, which represent only 1% of the adult tumours worldwide. It is one of the rare, aggressive connective tissue neoplasm of malignant fibrocytes in a myxoid matrix, and mostly occurs in people in their 60s to 80s. Like many other tumours of connective tissue, it reveals high recurrence rates, but rarely metastasise. We present a case of a 50-year-old female who had a large, soft tissue sarcoma over the left leg. Wide surgical excision was done due to its increased size and aggressive clinical behaviour, so as to increase the patient's comfort. On histopathology, the tumour was diagnosed as high-grade myxofibrosarcoma with no evidence of metastasis.

Keywords: Myxofibrosarcoma, local recurrence, metastasis, soft tissue sarcoma.

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Introduction

Myxofibrosarcoma is an aggressive tumour of connective tissue that is fibroblastic in origin and set in a myxoid matrix and has been categorised as a myxoid variant of malignant fibrous histiocytoma.^{1,2} It is commonly found in the extremities of elderly people^{3,4} with highest rate of occurrence in the seventh decade² of life with a slight male predominance.⁵ There is no known ethnic or racial predilection.⁶ They are mostly encountered in the lower extremities (77%), trunk (12%), and retroperitoneum or mediastinum (8%) and head (3%).^{1,2,7} Morphologically, these tumours may be categorised as high-grade, intermediate-grade, or low-grade. The cause of myxofibrosarcoma is unidentified and no known risk factors have been described till now.⁶

Complete surgical resection is the standard treatment of choice.^{2,4,6,8} Adjuvant or neoadjuvant radiotherapy and/or chemotherapy can be used in patients with limited disease³ but the absolute role of both are not



Figure-1.

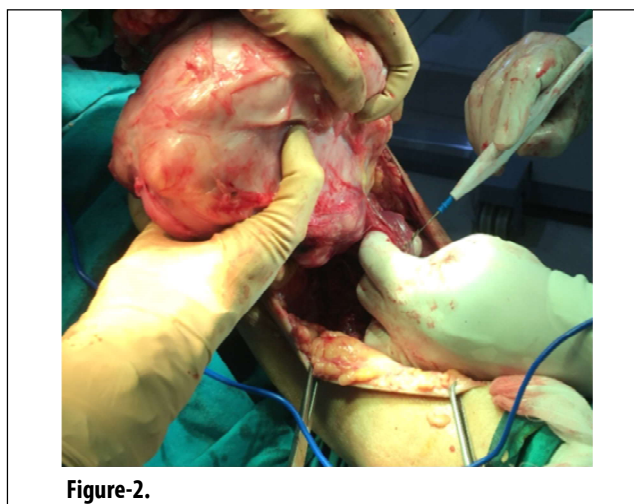


Figure-2.

fully clear.⁴ As it has an erratic infiltrative growth pattern along facial planes, during surgical resection negative margins are difficult to obtain.^{4,8} Although Myxofibrosarcoma has a better prognosis than other sarcoma histotypes, local recurrence rate is around 50-60%,³ leading to poor overall survival.⁸

Case Report

A 50-year-old woman, known case of diabetes and hypertension, presented to the surgical OPD of Abbasi Shaheed hospital on October 15, 2018, with the chief complaint of swelling on the medial aspect of left leg

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for the last 1.5 years. According to the woman she was in her usual state of health 1.5 years back when she noticed swelling for the first time on the medial aspect of her left leg just below the knee joint. Initially it was about tennis ball size which then increased gradually and continuously, and had grown to, according to her, football size. There was no prior history of trauma or any similar swellings. It was sudden in onset. Initially it was not associated with any pain, discharge, fever or any other symptoms but over the past six months its size had increased very quickly and pain became an associated feature. Pain was gradual in onset, dull aching in nature, continuous, increased on walking or exertion, and relieved on rest. She was pain-free when sitting or lying, she went to other hospitals for the same complaint one year back, FNAC was advised to her but was not performed due to unknown reasons.

On examination, the swelling was about 20x20x25 cm in size at the medial aspect of her left leg, extending just below the knee joint up to the mid leg at the posterior-medial aspect of the calf. It was round with a bosselated appearance, and the skin over it was erythematous, tense and glossy with venous prominence. No pigmentation, scar, ulcer or colour changes were noticed. On palpation it was hot, tender and firm with distinct, smooth margins. There were no signs of acute inflammation, and it was non-fluctuant, non-compressible, non-pulsatile, fixed to the underlying skin, immobile. No audible pulsations were noticed and distal pulses were palpable. There was no wasting or paresis.

On MRI of the left leg there was a large, lobulated abnormal signal intensity lesion in the soft tissues of the left leg posterior-medially measuring approximately 17x16x20 cm, involving subcutaneous tissues medially. It was laterally, abutting and infiltrating the medial aspect of gastrocnemius and soleus, and displacing the neurovascular bundle and the rest of the muscles. Superiorly, it extended up to the knee joint articulation and inferiorly lay approximately 15 cm above the distal articular surface of the tibia. Overall findings were suggestive of soft tissue neoplastic lesion of the left leg.

On colour Doppler scan of the left lower limb vessels, it showed high vascularity with low resistance. Arterial waveform suggested likelihood of neoplastic lesion. Doppler study of the left lower limb vessels was normal. There was no evidence of stenosis or occlusion.

Bone scan showed soft tissue swelling over the proximal half of the left leg posterior-medially with no underlying bony involvement.

The patient was prepared for surgery, with the suspicion of sarcoma in the left leg.

Excision and biopsy were done under anaesthesia. After giving elliptical incision over the swelling, the tumour mass was separated from the fat and fascial planes of the muscles. It was about 20x20x20 cm in dimension arising from the gastrocnemius muscle, compressing the vessels. It was excised completely. Haemostasis was secured and the wound was closed in layers.

The patient's recovery was unremarkable. She was discharged on third post-operative day.

Histopathology revealed a neoplastic lesion with multinodular architecture separated by incomplete fibrous septae. The neoplastic cells were plump spindle shaped and present in a myxoid background. Intervening areas showed curvilinear vessels with perivascular condensation of lymphocytes. Cells showed marked to moderate degree of pleomorphism. Areas of necrosis and haemorrhage were seen. Immunohistochemical stains showed ASMA & CD34 positive. These features favour high-grade myxofibrosarcoma. The closest peripheral and deep margin was 0.1 cm away. She was advised radiotherapy from oncology centre. Currently, the patient is on radiotherapy.

Discussion

Myxofibrosarcoma is a rare tumour, and accounts for about 5% of all soft tissue sarcomas.⁹ It is considered the most common malignant mesenchymal tumour in elderly patients.^{3,4,6,10} It was first described in 1977 as a myxoid variant of malignant fibrous histiocytoma and was reanalysed from the histological and immunohistochemical perspective as myxofibrosarcoma by WHO in 2013. It generally develops in the extremities of elderly people with a mean age of 65 years, commonly with a predilection for lower extremity and male gender.⁹ The exact pathology is unknown.⁶

Myxofibrosarcoma clinically presents as a slow growing painless,^{4,9,10} skin-coloured or erythematous swelling¹⁰ which can occur subcutaneously, as a multinodular mass, or deeply as a single nodule between the musculo-facial planes.⁹ Around 70% of the cases grow in the dermal and subcutaneous tissues, and 30% in the fascia

and skeletal muscle.^{1,6,10} Growths just beneath the skin are commonly multinodular in appearance, while deeper ones appear as a single infiltrating mass. Sometimes, it is very hard to determine the extent of cancer growth due to severe tumour penetration.⁶

A diagnosis of myxofibrosarcoma is based upon complete proper history and thorough physical examination, MRI or CT scan of the involved region and tissue biopsy of the tumour specimen before the surgery.⁶ The diagnosis is made histopathologically and tumours are categorised as low, intermediate and high grade. When the skin biopsy is taken superficially, it may appear to be a benign condition, since the shallow portions of the tumour shows benign characteristics, whereas deeper sections show histological features of malignancy. Therefore, it is essential to check the lesion effectively for histopathological examination.¹⁰

In Myxofibrosarcoma the rate of local recurrence is high, between 16-54%,^{1,5,9} linked with a high grade of tumour and tumour size of >5cm,⁵ but the incidence of metastasis is low, between 20% and 25%, which is usually linked to histomorphologically high-grade tumours.^{7,9} Tumour grade is the most important prognostic factor, patients with high grade tumours are three to four times more at risk of metastasis as compared to patients with low-grade tumours.⁵ The most common site is the lung, followed by the pleura, lymph nodes, skin and soft tissue and bones.^{1,8,9}

The treatment of choice for soft tissue sarcomas, including myxofibrosarcomas, is wide surgical excision, in combination with radiotherapy and/or chemotherapy^{5,6} but due to local aggressive relapses in some cases amputation or radical resection may be required.⁹ Due to a lack of randomised trials, the role of adjuvant and neoadjuvant radio-chemotherapy in improving local or distant recurrences is not well defined.⁹ Pre- and postoperative radiotherapy has been considered and plays an important role as limb-saving therapy and also decrease the risk of local recurrence. Chemotherapy is usually advised to those patients who have high-grade tumours, as it decreases the risk of distant metastases and improve overall survival.⁵ Post-operative care is important and essential; minimum activity is advised until the surgical wound heals. Follow-up care with regular screening is important and a long-

term follow-up is advised. In general, the five-year survival rate is about 60% to 70%.⁶

Conclusion

The management of myxofibrosarcoma varies according to the grading defined by radiological and histopathological confirmation. In our case, we observed that even though the role of chemotherapy and radiotherapy in combination is recommended for high grade myxofibrosarcoma, our oncological management was limited to radiotherapy only. At present, six months post-operatively patient is doing well. Therefore, prompt timely diagnosis with wide surgical excision with safety margins in the absence of metastasis may play a role in a better prognosis with merely radiotherapeutic intervention.

Disclaimer: Informed consent taken by patient.

Conflict of Interest: None.

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