

A RARE CASE OF INTERMUSCULAR MYXOMA

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ABSTRACT

Introduction: Intermuscular myxoma is a rare benign soft tissue tumor with excess myxoid matrix and has an incidence of 0.1 to 0.3 per 100,000. The frequent locations include mostly lower extremities but can also occur in the upper limbs and neck. **Case report:** Here I illustrate the case of a 40 year old man presented with a slow growing mass in left popliteal region. MRI and then FNAC concluded that it is intermuscular myxoma. He underwent a surgery for excision of the tumor with an uneventful post operative period. **Conclusion:** Intermuscular myxoma is an uncommon benign neoplasm most commonly occurring in the lower limbs. It is most common in adults. Literature suggests that they originate from primary mesenchymal cells. Methods such as CT, Ultrasound and MRI are used for diagnosis with MRI being suggested most often. Biopsy is gold standard for the diagnosis and is extremely useful for diagnostic differentiation. En-bloc excision is the treatment of choice.

KEYWORDS : Myxoma, En-Bloc, FNAC, Excision, Myxoid Matrix, Stellate cells.

INTRODUCTION:

Intermuscular myxoma is a rare benign soft tissue tumor that presents as a slowly growing deeply seated mass. It has an incidence of 0.1 to 0.3 per 100,000[1]. The common sites including thigh, upper arm, calf, and the gluteal region. These are with a hallmark of abundant extracellular myxoid matrix. Although the lesion appears to have clear borders and a contained appearance, some times it can be found in surrounding tissues. MRI is the radiological modality of choice in diagnosing these cases, while tissue biopsy is the gold standard.

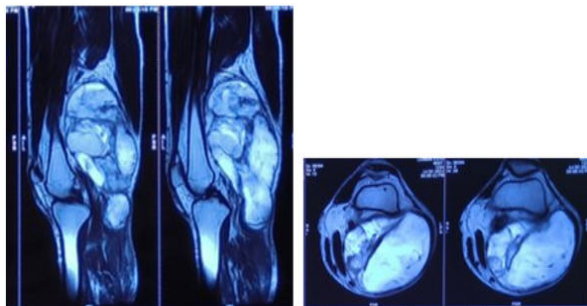
Case report/Summary:

A 40Y old man was admitted with slow-growing relatively painless swelling in the left popliteal region for 8 years. The recent onset of pain during walking for a few weeks causing difficulty in his activities brought him to the hospital.

On clinical examination, a smooth, lobulated painless and firm swelling covering popliteal region extending into back of thigh posteriorly. MRI was suggestive of myxoma/ chondromyxoid tumour.

FNAC- myxomatous soft tissue lesion with a borderline malignancy pattern. He underwent complete resection of the lesion by single S shaped incision over posterior thigh, curving from superomedial to inferolateral direction. En-bloc excision without any spillage measuring 18x10x10 cm.

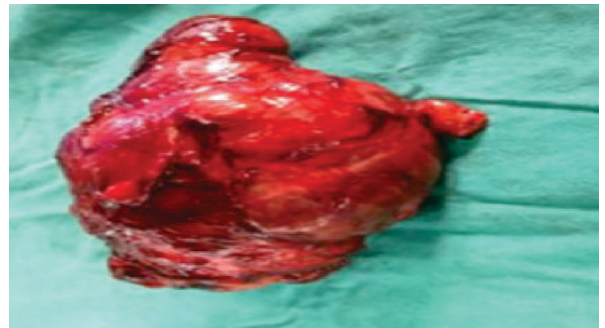
The tumour excision was done carefully under spinal anesthesia avoiding the tissue and vessels surrounding it. Histopathology report showed encapsulated tumor tissue comprised of stellate cells and few spindle cells with abundant of myxoid areas. Histopathological diagnosis was consistent with MYXOMA. The patient made an uneventful recovery and there were no signs of recurrence after surgery.



Lesion on coronal section of MRI and axial section of MRI



HPE: Stellate cells and few spindle cells with abundant myxoid stroma



Excised specimen en bloc

DISCUSSION:

Intermuscular myxoma is a rare benign soft tissue tumor with incidence of 0.1 to 0.3 per 100,000[1]. Term myxoma was first used by Virchow (1871)[2]. Literature suggests that they originate from primary mesenchymal cells, lost capacity to produce collagen instead produces copious amount of hyaluronic acid and immature collagen. Although they are most commonly intramuscular in location(82%), they can rarely be intermuscular or subcutaneous[1]. They can be located in any skeletal muscle group however most commonly affect the lower limbs compared to the upper limbs. Tumor either manifests as either a solitary lesion or in association with fibrous dysplasia[3] such as Mazabraud's syndrome[4] and McCune-Albright syndrome.

On using MRI, the mass commonly displays hypodense in T1-weighted images and hyperdense in T2-weighted, gradient echo or STIR images[5]. The tumor was examined using a fine needle aspiration(FNA). The examination showed that the tumor cells were stellate shaped, hypocellular and hypovascular along with some areas of cellularity and vascularity increased[6]. These findings were consistent with myxoma characterized by a myxoid background, which was

confirmed through histopathological examination after surgical excision of the lesion.

The differential diagnosis includes myxofibrosarcoma, myxoid liposarcoma, myxoid chondrosarcoma, myxoid fibrous histiocytoma, Histopathological investigation should be performed when there is an uncertainty of diagnosis to exclude other differentials including primary malignancy or metastatic disease. Myxoid fibrous histiocytoma differs from myxoma as it has heterogeneous appearance. Even though MRI and other imaging procedures are key for diagnosis, histopathological understanding is necessary to differentiate the diagnosis from other tumours. Low grade fibromyxoid sarcoma (LGFMS) frequently resembles IM, as they share similar histopathological characteristics, including a myxoid background, unassuming oval spindle cells. Recent literature has revealed that Mucin 4 (MUC4), a gene commonly found in different carcinomas, exhibits high sensitivity and specificity in LGFMS. Therefore, the absence of MUC4 expression on immunohistochemistry staining proves valuable in differentiating between IM and LGFMS[7].

The recommended treatment is surgical excision with complete resection of the tumor clearing the tumor margins in surrounding tissues. Incomplete resection might cause recurrence. A recent study concluded that the use of zoledronic acid therapy decreased the bone pain and reduced the size of the myxoma[8].

CONCLUSION:

Intermuscular myxomas are benign soft tissue lesions in which surgical excision is curative. So, patients presenting with a solitary soft tissue tumor should, therefore, be thoroughly investigated to rule out the soft tissue sarcomatous pathology. En-bloc excision without spillage is the treatment of choice which ensures long and lasting recovery.

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