Case Report

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Intramuscular Myxomas: A Potential Diagnostic Dilemma

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Abstract

Intramuscular myxomas are rare benign soft tissue tumors of mesenchymal origin. They show areas of increased cellularity and vascularity that can lead to a mistaken diagnosis of sarcoma, especially myxofibrosarcoma, low-grade fibromyxoid sarcoma, and myxoid liposarcoma. FNAC of such a tumor is inconclusive as it may show just myxoid stroma or hypercellularity depending on whether the hypercellular portion or myxoid portion is sampled which further impedes diagnosis. Hence, total surgical exision followed by histopathological examination of the tumor is the gold standard. Their recognition is important to avoid an erroneous diagnosis of sarcoma.[2] Surgery is always curative and recurrence is rare. This is a case report of 55 years old male presenting with such a tumor.

Keywords: Intramuscular myxoma; Myxomas.

Introduction

Intramuscular myxomas are a rare and benign type of soft tissue myxoma, considered to be of mesenchymal origin. They usually present as a long standing, painless mass arising from large muscles around the thighs, buttocks, or shoulder girdle regions. Commonly seen in adults between the 4th and 6th decades with a female predilection (70%). Their incidence ranges from 0.1 to 0.13 per 100,000 population.[1,2] They are pathologically and classically described as hypocellular and hypovascular tumor, composed of cytologically bland stellate cells and bipolar fibroblasts separated by abundant extracellular myxoid matrix. They are often seen invading the surrounding muscle from which they arise from. They are also seen as a part of Mazabraud Syndrome, which is a rare syndrome commonly seen in elderly women characterized by polyostotic fibrous dysplasia and multiple intramuscular myxomas in large muscle groups, which is closely associated with McCune Albright syndrome.[3–5] Surgery is curative, recurrence is rare. Although if recurrence does occur, it is a local recurrence.[6]

Case Report

A 55 years old male came with complaints of swelling over the right thigh upper medial aspect since 5 months duration. The swelling was initially small and gradually progressed to an ovoid swelling of size of approximately 8 × 6 cms. There was no associated pain, weight loss, loss of appetite and no restriction of movements. Examination of the swelling revealed a smooth, ovoid, nontender swelling of 8 × 6 cms size noted over the right upper medial aspect of the thigh. The consistency was firm to hard with ill-defined margins and was not mobile. However, on adduction of the thigh, the swelling was seen to become less prominent.

Keeping in mind the possibility of a sarcomatous growth, the patient was thoroughly investigated. An ultrasound imaging of the thigh was suggestive of a heterogeneously, hypoechoic lesion of size 6 × 5.2 × 5 cms, encapsulated with no vascularity

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within the tumour in the superficial muscle plane suggestive of soft tissue neoplasm. An MRI of the thigh was done which is suggestive of a 6.2 × 5. 8 × 5.3 cm well defined, smooth swelling in the intramuscular plane without invasion of surrounding structures in right upper thigh seen to be in close promixity to femoral vessels, suggestive of benign soft tissue lesion (Fig. 1).

An HRCT of the thorax was done to rule out any concurrent lung metastasis and revealed a_normal study. An oncosurgical opinion was sought, and a complete surgical excision was planned. A lazy "S" incision was taken over the medial aspect of the right upper thigh. Intraoperatively a 12 × 10 cm swelling was noted arising from the right Adductor Magnus muscle, which was adherent to the femoral vessels and seen to encroach into the muscle. The swelling was excised in toto with a 2 cm margin and a few fibers of the muscle where the tumor was seen to be encroaching were also shaved off. The cut section of the tumor revealed gelatinous material. The tumor was sent for histopathological examination (Fig. 2).

Histopathological examination revealed a well-circumscribed tumor intramuscular tumor on gross examination, composed of bland spindle shaped cells with round to oval an intramuscular myxoma with all margins free of tumor and was adviced close follow up as a low grade fibromyxoid sarcoma can mimic this tumor.

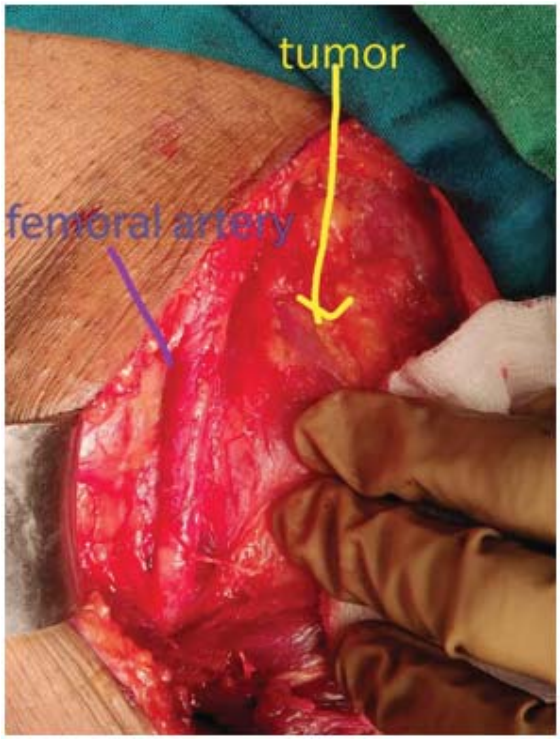

Fig. 1: Tumor Association with femoral artery

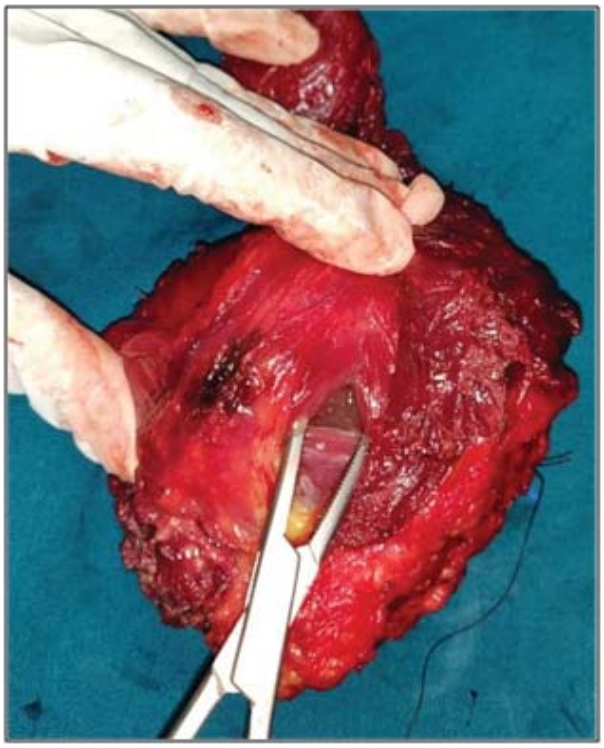

Fig. 2: Cut open specimen revealing gelatinous material

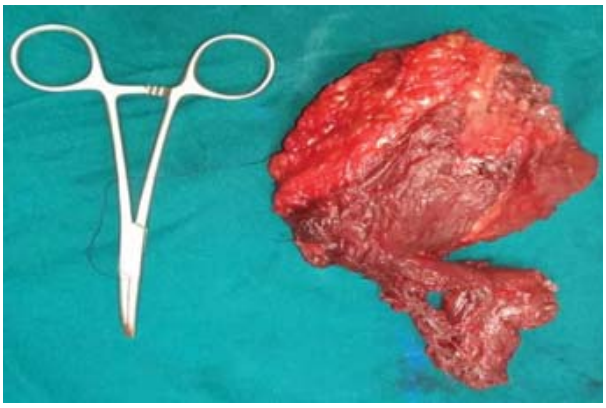

Fig. 3: Excised tumor with shaved off muscle fibers

Treatment and follow up

Surgery is the definitive cure for all intramuscular myxomas. A wide local excision with a 2 cm margin was done and a few of the superficial fibers of the adductor magnus muscle were shaved off. The patient followed up with the Oncologist, and was reassured it to be a benign lesion with no further management. Patient is currently on a 6 monthly follow up.

Discussion

Myxomas are soft tissue tumors of mesenchymal origin. First described by Virchow et al. in 1863 as

a benign tumor resembling a similar substance to that of the umbilical cord.[7]

Intramuscular myxomas are a rarer type of myxoma, which were first reported by Enzinger et al. in 1965[8] till this period, the existence of myxomatous tumors were very well described in literature but this rare entity which showed similar histopathological features to that of a myxoma, but seen to arise intramuscularly was unheard of. These tumors are seen to occur in the muscles of the thigh, buttocks, shoulder and upper extremities[9] (Fig. 3). Intramuscular myxomas primarily affect patients between 40 and 70 years of age, with female predominance.[1–5]. They present as a painless, palpable mass, or varying size either from the shoulder, hip girdle musculature or gluteal musculature, rare cases of them arising distally from the hypothenar muscles also have been reported.[10] These tumors may occur as a solitary lesion or in association with fibrous dysplasia (McCune Albright syndrome) wherein they are multiple in number.[4,5,10] Histopathologically described as a tumor with abundant myxoid matrix with areas of hypocellularity with hypercellularity, composed of bland undifferentiated stellate cells. However, they do not show any signs of mitotic figures or nuclear atypia, hence benign. They are hypovascular tumors. Immunohistochemical studies have shown staining for Vimentin and no reaction for S 100. Due to the coexistence of hypercellular and hypocellular areas, FNAC is usually inconclusive in diagnosis.[12,13] Radiologically they appear as a well defined hypo echoic lesion of soft tissue origin on ultrasound. On CT intramuscular myxoma are seen as a well-defined homogenous low density lesion. MRI characteristics of the tumor are seen to have low signal intensity on T1 weighted images and high signal intensity on T2 weighted, gradient echo or STIR images.[14] Treatment of choice is complete surgical exicision with a tumor free margin. This is because the capsule is usually incomplete and the tumor is often seen invading the surrounding musculature.[12] Recurrences are rare, and even if they occur are local. There have been reports of recurrent intramuscular myxomas, where treatment is surgical resection followed by radiotherapy.[11]

Conclusion

Intramuscular myxomas are a rare, benign soft tissue tumor of mesenchymal origin. They show areas of increased cellularity and vascularity that can lead to a mistaken diagnosis of sarcoma, especially myxofibrosarcoma, low-grade fibromyxoid sarcoma, and myxoid liposarcoma. FNAC of such a tumor is inconclusive as it may show just myxoid stroma or hypercellularity depending on whether the hypercellular portion or myxoid portion is sampled which further impedes diagnosis. Hence, total surgical exision followed by histopathological examination of the tumor is the gold standard. Their recognition is important to avoid an erroneous diagnosis of sarcoma. Surgery is always curative and recurrence is rare.

References

1. Heymans O, Gebhart M, Alexiou J. Intramuscular myxoma. Acta Chir Belg 1998;98:120–22.
2. Ozbek N, Danaci M, Okumus B, et al. Recurrent intramuscular myxoma: Review of the literature, diagnosis and treatment options. Turk J Cancer 2006;36(2):75.
3. Nielsen GP, O'Connell JX, Rosenberg AE. Intramuscular myxoma: A clinicopathologic study of 51 cases with emphasis on hypercellular and hypervascular variants. Am J Surg Pathol 1998;22(10):1222–227. Pubmed citation – surgical resection curative.
4. Kindblom LG, Stener B, Angervall L. Intramuscular myxoma. Cancer. 1974;34:1737–1744. doi: 10.1002/1097-0142(197411)34:5<1737::AID-CNCR2820340523>3.0.CO;2-U [PubMed] [CrossRef] [Google Scholar].
5. Miettinen M, Hockerstedt K, Reitamo J, et al. Intramuscular myxoma: A clinicopathological study of twenty-three cases. Am J Clin Pathol. 1985;84:265–272 [PubMed] [Google Scholar] - fibrous dysplasia association.
6. PN Chan, SF Lee, TC Chow, et al. A rare benign tumor: Intramuscular Myxoma. J HK Coll Radiol 2004;7:40–3.
7. Virchow R. Cellular pathology as based upon physiological and pathological histology. Philadelphia: JB Lippincott; 1863. pp. 525–26 [PubMed] [Google Scholar].
8. Enzinger FM. Intramuscular myxoma; A review and follow-up study of 34 cases. Am J Clin Pathol. 1965;43:104–113 [PubMed] [Google Scholar].
9. Allen PW. Myxoma is not a single entity: A review of the concept of myxoma. Ann Diagn Pathol. 2000;4:99–123. doi: 10.1016/S1092-9134(00)90019-4 [PubMed] [CrossRef] [Google Scholar].
10. Al-Qattan MM, El-Shayeb A, Rasool MN. An intramuscular myxoma of the hand. Hand Surg 2004;9:97–99 [PubMed] [Google Scholar].
11. Robert J Logel. Recurrent Intramuscular myxoma associated with Albright's Syndrome: Journal of Bone and Joint Surgery. 1976 Jun;58(4):565–8.

12. Yaligod V, Ajoy SM. Intramuscular myxoma: A rare tumor. Journal of Orthopedic Case Reports. 2014;3(4):41–44. doi: 10.13107/jocr.2250-0685.130 [PMC free article] [PubMed] [CrossRef].
13. Kabukcuoglu F, Kabukcuoglu Y. Mazabraud's syndrome: Intramuscular myxoma associated with fibrous dysplasia. Orphanet Encyclopedia. 2005 Jan [Google Scholar].
14. Lal B, Gandhi V. IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) 2018 Sep;17(9),Ver 3:36–38.