Abstract: Intramuscular myxoma is a very rare benign soft tissue tumor. The lack of specific symptoms and widely used laboratory tests makes the diagnosis quite difficult. We report a case of an intramuscular myxoma in the form of a gluteal swelling which is of short duration. We cannot conclude with the diagnosis until we get the histopathological examination report.

Keyword: Interesting gluteal swelling, Intramuscular myxoma.

AN INTERESTING CASE OF A GLUTEAL SWELLING – INTRAMUSCULAR MYXOMA (IMM)

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Introduction: Intramuscular myxoma (IMM) is a rare benign soft tissue tumor that presents as a slowly growing, deeply seated mass confined to the skeletal muscle. IMM has an incidence of 0.1 to 0.3 per 100,000. According to the World Health Organization, IMM is classified as a tumor of uncertain differentiation. The majority of this tumor presents in the 4th to 6th decade and there is a slight female predominance.

Plain X-ray may be normal or show some non-specific soft tissue mass. On ultrasonography, it appears as a heterogeneous hypoechoic relative to skeletal muscle mass, with well-defined margins. On CT-scan, IMM is a well-defined, homogeneous, low density mass within the muscle. On MRI, it has homogenous low signal intensity in T1 weighted images. But these images cannot differentiate this tumour from other tumours. The definite diagnosis can be made only with the histopathological examination after surgical excision.

The differential diagnosis include sarcoma, metastasis and some benign tumour such as lipoma, hemangioma, hematoma and desmoids tumour.

Case Report:
A 32 year old male patient presented with a swelling in the left gluteal region for one month. It was insidious in onset and gradually increase in size. The patient complained of difficulty in sitting due to the swelling. Not associated with pain or discharge from the swelling. No history of trauma or I.M. injection and no history of fever with chills and rigor. No history of similar swelling in the past. He did not have any co-morbid illness and he was taking mixed diet. On general examination, the patient was moderately built and well nourished. His B.P was 110/60 mmHg and pulse rate 74/min. On abdominal examination, abdomen was soft and no organomegaly. On systemic examinations no abnormality detected. No generalized lymphadenopathy.

On local examination, a hemispherical swelling measuring 10x8 cm was seen on the left gluteal region. It extended superiorly - iliac crest, inferiorly - gluteal fold, laterally side of thigh, medially inter-gluteal canal. The borders were well defined with smooth surface. The skin over the swelling was normal. The swelling became more prominent on contracting the gluteal muscle. It was mobile, not warmth, not tender, soft, fluctuant and not transilluminant. No pulsation was felt. No inguinal lymph node palpable and no signs of distal neurological deficit. The provisional diagnosis made was soft tissue neoplasm ( ? Lipoma) or ? Lymph cyst.

Routine blood investigations were normal. Ultrasound showed multiseptated cystic swelling 9x7 cm with solid component, suggested HPE correlation ( ? Lymphocoele). FNAC showed scanty scattered macrophage on a proteinaceous background suggestive of cystic lesion ( needs HPE).
CECT images show:
Well-defined, homogenous mass at the left gluteal region (Fig.1&2).
† Hydatid cyst.

Operative Notes:

HPE report:
Section shows multiloculated cystic cavities surrounded by highly myxoid bluish fibrous stroma containing few scattered stellate cells. Benign myxoid tumour with histological feature of less cellular myxoma

Then excision of the cyst, done under spinal anaesthesia by making an elliptical incision over the swelling. The cyst was removed in to-to (Fig.3). Hemostasis achieved(Fig.4) , DT kept and wound closed in layers. Suction drain removed on 7th PoD and sutures removed on 10th POD.

Discussion:
Intramuscular myxoma is a rare entity. It is classically described as hypocellular and hypovascular, and is composed of cytologically bland stellate and bipolar fibroblasts separated by abundant extracellular myxoid matrix. The clinical manifestations are non-specific, in most of the patients the sole presenting sign is a painless, palpable mass that is firm, slightly movable and often fluctuant. Pain or tenderness is present in fewer than one-fourth of patients. The large tumour may produce numbness, paresthesia, and muscle weakness distal to the lesion. Because of the relative lack of symptoms, most of the tumours are present for several months or even years before they are excised. A history of trauma is rarely given, and the tumour is not etiologically related to thyroid dysfunction, as in myxedema. IMM usually arises from large skeletal muscles like the thigh (51%) and the gluteal area (7%) . The majority of IMMs appear as a single mass. If multiple, they are associated with fibrous dysplasia of the bones of the same extremity, known as Mazabraud syndrome . In most of the cases the fibrous dysplasia are noted during the growth period whereas the multiple IMMs become apparent only during adult life. Occasionally, multiple IMMs may appear first. So, if specifically sought radiological evidence of bone abnormalities can be detected in patients with multiple IMMs.

Pathological features: The gross appearance of IMM is characteristic and changes little from case to case. Most tumours are ovoid or globular and have a glistening gray-white or white appearance , depending on the relative amounts of collagen and myxoid material.
They consist of a mass of stringy, gelatinous material with occasional small fluid-filled, cyst-like spaces, and they are covered by bundles of skeletal muscle or fascial tissue. On histologic examination, the tumor varies little in its appearance and is composed of relatively small numbers of inconspicuous cells, abundant mucoid material, and a loose meshwork of reticulin fibres. The constituent cells have small, hyperchromatic, pyknotic-appearing nuclei and scanty cytoplasm that sometimes extends along the reticulin fibres with multiple processes, giving the cell a stellate appearance.

In IMM mitotic activity is practically absent unlike sarcoma. Immunohistology shows expression of vimentin and a myxoid material which is entirely digestible by hyaluronidase. IMM shows no reactivity for S-100 protein, unlike myxoid liposarcoma. The treatment of IMM is wide local excision, and has an excellent prognosis. Recurrence rate is less than 5%. Recurrence may be attributable to insufficient resection of the tumor. In the rare examples that do recur, re-excision is typically curative. Recent studies show that the detection of GNAS1 mutations has an increased specificity in the diagnosis of IMM, although testing for GNAS1 mutations is not commonly applicable. This makes the diagnosis of IMM difficult before surgical excision.

Regarding our patient, we diagnosed IMM only after the HPE report. But we had fully resected the tumour and the patient had no evidence of recurrence at follow-up 6 months later. We publish this paper as IMM is a rare entity but should be considered as a differential diagnosis while coming across such a prominent soft tissue swelling.

Reference: