Myxoma… An Enigma!

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Abstract

Although not rare, myxomas are benign tumours of uncommon incidence. Intramuscular myxomas are of very rare occurrence, incidence ranging from 0.1 to 0.13 per 100,000. Less than 200 cases of Intramuscular myxoma have been reported in literature by now and to the best of our knowledge, none of them in Peroneus longus. We report a case of intramuscular myxoma found in Peroneus Longus of a 32 year old male. A 32 year old male presented with a painless lump on upper part of his right leg. The swelling had gradually increased in size since its appearance 8 months prior to his visit. On MRI location into peroneus longus confirmed. Surgical excision done and histopathology was suggestive of myxoma. We report probably the first case of an intra muscular myxoma of Peroneus longus muscle. From an orthopaedic surgeons point of view, possibility of Mazabraud's syndrome should be kept in mind and skeletal survey should be carried out if needed to rule out fibrous dysplasia.

Keywords: Myxoma, peroneus longus, Surgical excision, intramuscular, Mazabraud's syndrome

Introduction

Although not rare, myxomas are benign tumours of uncommon incidence. The most usual locations of occurrence are various, including the heart, subcutaneous and aponeurotic tissues, genitourinary tract, skin, retroperito¬neum, intestine, pharynx, and skeleton. They are mesenchymal tumours of fibroblastic origin which produce excess mucopolysaccharide, are incapable of producing mature collagen and histologically resemble the umbilical cord.[1,2] But, amongst these, Intramuscular myxomas are of very rare occurrence, incidence ranging from 0.1 to 0.13 per 100,000. [3] The intramuscular variety is most commonly found in large muscles of thigh, and then in less common locations like Shoulder muscles, Gluteal muscles, Paraspinal muscles, trunk and leg muscles.[4]Less than 200 cases of Intramuscular myxoma have been reported in literature by now and to the best of our knowledge, none of them in Peroneus longus. We report a case of intramuscular myxoma found in Peroneus Longus of a 32 year old male.

Case report

A 32 year old male presented with a painless lump on upper part of his right leg. Patient didn't have any history of trauma at the site, nor a history of similar lumps elsewhere in the body. The swelling had gradually increased in size since its appearance 8 months prior to his visit. On examination, the mass was just below the head of the fibula, measured about 5 x 3 centimetres, was firm in consistency and was mobile. Absence of foot drop was confirmed keeping the location of the mass in mind. Medical history, plain radiograms and routine blood investigations of the patient were unremarkable.

On ultrasonography, the shape was regular ovoid with clear boundary echo and heterogenic internal echo. To evaluate further, MRI of the lesion was done, which showed on T1-weighted imaging a well-defined hypointense intramuscular lesion in the peroneus longus muscle [Figure 1a], The lesion was hyperintense on T2-weighted imaging without engulfing the peroneal nerve [Figure 1b and Figure 1c].

Fine needle aspiration cytology was not performed because a malignant lesion was not suspected. Surgical excision was done with an incision on the lateral aspect of upper leg directly overlying the lesion. Peroneal nerve was carefully separated, and a well capsulated, ovoid mass of 6x4x3 cm was excised [Figure 2a and Figure 2b].

No post-operative complications were encountered. The histopathology confirmed the diagnosis of myxoma based on small spindle cells, stellate cells, fibres in abundant myxoid stroma and absent mitotic activity [Figure 3].

After 1 year of follow up, no recurrence was seen, patient was symptom free.

Discussion

Intramuscular myxoma is a mucopolysacchride rich benign tumour arising from skeletal muscles in adults. Till date, a definitive aetiology for intramuscular myxomas has not yet been described; however, it is commonly thought that malfunctioning fibroblasts are responsible for both the presence of immature collagen fibres and the abundance of glycosaminoglycans.[5]

In 1863, it was Virchow who coined the term "myxoma" to

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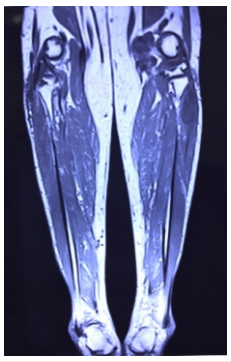

Figure 1a: Hypointense signal in proximal peroneus longus on T1 coronal sequence of MRI

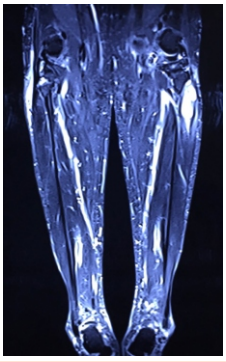

Figure 1b: Hyperintense signal in proximal peroneus longus on T2 coronal sequence of MRI

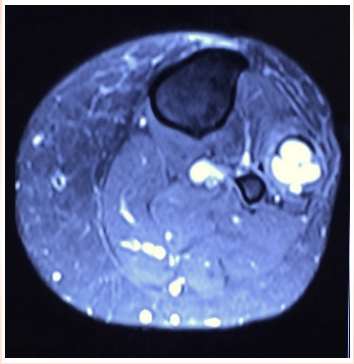

Figure 1c: Lesion not involving the peroneal nerve

describe a tumour histologically resembling the umbilical cord [6]. In 1948, Stout outlined the diagnostic criteria which had to be fulfilled before a tumour could be labelled as a myxoma in a study of 142 cases. [7]. He found only 3% of all the myxomas were in¬tramuscular. In 1965, Enzinger reported the number to be 17% of the 200 cases he collected.[8] They are most commonly found in the age group of 4-70 years, occur in females more than males, and have an adult predomi¬nance, with only two cases reported in infants.[9]. Almost half of the reported cases hail from large muscles of thigh and then other parts but to the best of our knowledge, no case of an intramuscular myxoma in Peroneus longus has been reported. Intramuscular myxoma may be associated with fibrous dysplasia, and they may be multiple, an association that is termed Mazabraud's syndrome. It may also be associated with McCune-Albright syndrome, where polyostotic fibrous dysplasia, endocrine dysfunction and café au lait spots are also be present.(10)

Considering the radiology, the ultrasound images of intra muscular myxomas are a well-circumscribed mass with complete or partial capsule, a complex intramuscular lesion that is heterogeneously hypoechoic with mildly increased through transmission and echogenic triangles at the poles of the lesion. Typically, the internal vascularity is absent and there is a central hypoechoic area within the mass. Even though Ultrasound is very suggestive, MRI is the most definitive and conclusive investigation for diagnosing myxomas. It shows a heterogeneous intramuscular lesion with low signal intensity on precontrast T1WI MR and a "bright rim" of fat as well as "bright caps" prominent at the superior and inferior poles of the lesion.[11]. Specificity for the diagnosis of the intramuscular myxomas can be improved by the use of intravenous contrast for demonstration of both peripheral and internal enhancement secondary to the pseudocapsular pathology characteristic of these lesions.[12]

Macroscopically, the tumour is gray-white, shiny, and is circular or ovoid in shape. On close examination of the borders, most intramuscular myxomas are found to infiltrate surrounding muscle. On cutting open, it has a white or grey-white mucoid gelatinous surface. [13] It is difficult to diagnose this tumour before biopsy. [14]Due to abundant substance of the myxomatous tissue and poor cellularity, it is difficult to make a diagnosis on FNAC. Intramuscular myxoma is histologically composed of sparse spindle and stellate cells in abundant mucoid material. An irregular meshwork of reticulin is observed, and blood vessels are infrequent. The basic histology of intramuscular myxoma is indistinguishable from myxoma that occurs elsewhere in the body. The tumour appears well circumscribed or encapsulated; however, on close inspection, the delicate fibrous capsule is incomplete, and most of all lesions infiltrate adjacent muscle.

The diagnosis of intramuscular myxomas is difficult owing to the lack of specific symptoms and common labora¬tory tests. It has been reported that carbohydrate antigen (CA) 19 9, a tumour marker, may be correlated with Intramuscular myxoma.. In a previous study, the serum level of CA 19 9 increased preoperatively and returned to a normal level six months following surgery. The conclusion couldn't be solidified as the levels also increased in a variety of other malignant and benign conditions. (15)In differential diagnosis, the benign tumours confused with intramuscular myxomas are lipoma, hemangioma, angiomyx¬oma, dermoid cyst, epidermoid cyst, myxoid neurofibroma. Intramuscular myxomas must be distinguished from myxoid liposarcomas–a much more ominous diagnosis. Enzinger et al reported a case of intramuscular myxoma of Soleus who went on to have an amputation for incorrect diagnosis of the lesion [8].

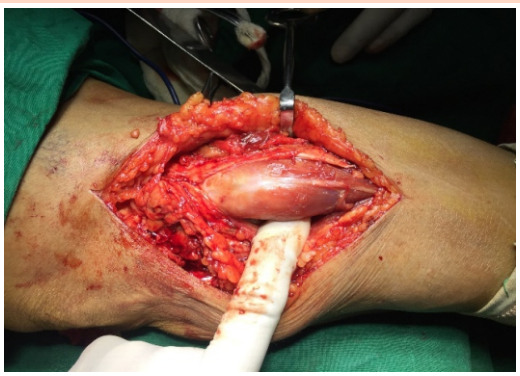

Figure 2a: Lesion not involving the peroneal nerve

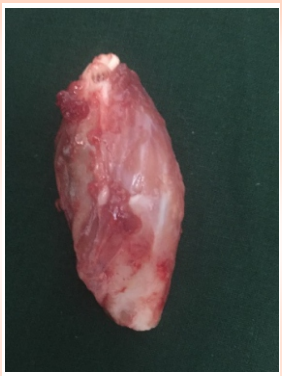

Figure 2b: Hyperintense signal in proximal peroneus longus on T2 coronal sequence of MRI

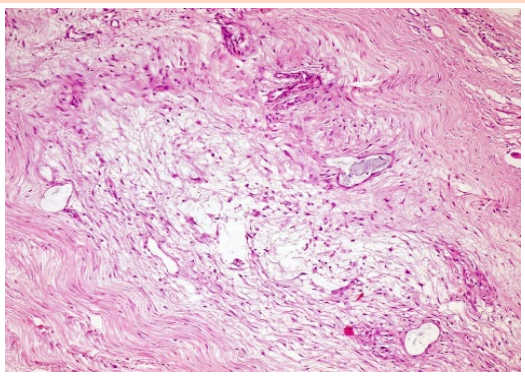

Figure 2a: Lesion not involving the peroneal nerve

The definitive treatment for intramuscular myxoma is surgical excision. It has also been suggested in literature to excise a of a small amount of surrounding musculature to prevent local recurrence, as intramuscular myxomas tend to be infiltrative and its borders may extend beyond visible margins. Though some cases of recurrence due to inadequate excision have been reported, no case of metastasis has ever been reported. (16). While the role of radiation therapy is not clear, odontogenic myxomas have been reported as having poor responses to radiotherapy (17)

To summarize, we report probably the first case of an intra muscular myxoma of Peroneus longus muscle. Due to lack of specific pathognomonic features, it is possible to make a confirmed diagnosis only after post-operative histopathology. From an orthopaedic surgeon's point of view, possibility of Mazabraud's syndrome should be kept in mind and skeletal survey should be carried out if needed to rule out fibrous dysplasia.

Disclosure

i) Study was approved by the Institutional Review Board/Ethical Committee.

ii) Participant provided written informed consent for the participation in the study.

iii) All procedures performed in the study were conducted in accordance with the ethics standards given in 1964 declaration of Helsinki, as revised in 2013.

iv) Patient consent was obtained for purpose of the study with due care to maintain his privacy.

v) The Author(s) declare(s) that there is no conflict of interest.

Declaration of patient consent : The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest : Nil **Source of support :** None

References

1. Shugar JM, Som PM, Meyers RJ, et al. Intramuscular head and neck myxoma: report of a case and review of the literature. Laryngoscope 1987;97:105–7.
2. Nishijima W, Tokita N, Watanabe I, et al. Intramuscular myxoma of the neck. Arch Otolaryngol 1985;111:699–701.
3. Ozbek N, Danaci M, Okumus B, Gursel B, Cakir S, Dabak N, Karagoz F. Recurrent intramuscular myxoma: review of the literature, diagnosis and treatment options. Turk J Cancer. 2006;36(2):75.
4. Allen PW. Myxoma is not a single entity: a review of the concept of myxoma. Ann DiagnPathol 2000;4:99–123.
5. Zac Tataryn et al, Intramuscular myxoma of the cervical paraspinal musculature: case report and review of the literature, American journal of Otorhinolaryngology,2014, volume 36, issue 2,273-276.
6. Murphey M, McRae G, Fanburg-Smith J et al: Imaging of soft-tissue myxo¬ma with emphasis on CT and MR and comparison of radiologic and patho¬logic findings. Radiology, 2002; 225: 215–24
7. Stout AP: Myxoma the tumor of primitive mesenchyme. Ann Surg, 1948; 127: 706–19
8. Enzinger FM: Intramuscular myxoma a review and follow-up study of 34 cases. AM J ClinPathol, 1965; 43: 104–13
9. Ishoo E: Intramuscular myxoma presenting as a rare posterior neck mass in a young child: case report and literature review. Arch Otolaryngol Head Neck Surg 133: 398 401, 2007.
10. Shuitingfu, Zhuowei tian, Chenpingzhang,yue he, Monosotic fibrous dysplasia and solitary intramuscular myxoma of the head and neck: A unique presentation of Mazabraud's syndrome and a literature review, Oncol Lett. 2015 November; 10(5): 3087–3094.
11. Zou, Lily Y.; Brown, Desmond A.; Li, Albert C. Intramuscuar Myxoma, Ultrasound Quarterly: September 2013 - Volume 29 - Issue 3 - p 255–256doi: 10.1097/RUQ.0b013e3182a25746
12. Luna, A., S. Martinez, and E. Bossen, Magnetic resonance imaging of intramuscular myxoma with histological comparison and a review of the literature. Skeletal Radiol, 2005. 34(1): p. 19-28.
13. Ozawa H, Fujii M, Tomita T, et al. Intramuscular myxoma of scalene muscle: a case report. AurisNasus Larynx 2004;31: 319–22.
14. Aind R A, Dwivedi M K, Pal R, Devangan L, Shekhar P V. Intramuscular myxoma of gluteal region. Indian J Radiol Imaging 2004;14:177-8
15. Larentzakis A, Toutouzas K and Katsaragakis S: Intramuscular myxoma associated with an increased carbohydrate antigen 19.9 level in a woman: a case report. J Med Case Rep 5: 184, 2011.
16. Orlandi A, Bianchi L, Marino B, Spangnoli LG, Nini G. Intramuscular myxoma of the face an unusual localization. DermatolSurg 1995;21:251–4.
17. Large ND, Niebel HH, Feredricks WH. Myxoma of the jaws. Oral Surg Med PatholRadiol 1960;13:1462–8.

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