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Rare intramuscular myxoma of the flexor hallucis longus: A case report

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Abstract
Intramuscular myxoma is a rare benign soft tissue tumor with unclear etiology and histology. Typically occurring in the heart and rarely in the musculoskeletal system, it presents as a relatively clear lesion with a clinical diagnosis tending towards benignity. We report a case of a 74-year-old male with swelling and pain in the left leg, diagnosed via MRI with a characteristic cystic mass in the flexor hallucis longus muscle. Histopathological examination revealed typical features of myxoma, including low signal intensity on T1-weighted imaging and high signal intensity on T2-weighted imaging, consistent with a mucus-rich tumor stroma.

Keywords: Intramuscular Myxoma-Flexor Hallucis Longus

Introduction
Intramuscular myxoma can be found in any skeletal muscle group but most commonly occurs in the quadriceps (65%), hip adductor muscles (35%), gluteus muscles (20%), gastrocnemius, and upper arms [1, 2]. The size of myxoma has been reported to range from 1-17 cm. Intramuscular myxoma can be solitary or multiple, and when multiple myxomas appear, they are often associated with monostotic or polyostotic fibrous dysplasia, known as Mazabraud syndrome [3].
The incidence of this condition is reported as 1 in 1,000,000 and is most frequently diagnosed in female patients (57%) between 40-70 years old [1]. It is suspected to be related to postzygotic mutations of the GNAS1 gene located on chromosome 20q13.2 – q13.3, especially when associated with Mazabraud syndrome. GNAS1 encodes the G-protein alpha subunit (GSa) involved in cell proliferation. These mutations affect mesenchymal precursor cells in the early stages [4, 5].
Typical presentations include a slowly enlarging mass in 64% of patients. The lesions appear as deep-seated masses confined to skeletal muscles without malignant potential. Painful masses occur in 55% of patients [1, 3]. Lesions typically appear as well-defined intramuscular masses. Because myxomas are benign, conservative treatment is recommended. Myxomas should be excised if they cause pain, pressure symptoms, neurological symptoms, or impair function [6].
Histopathological investigation should be performed when there is diagnostic uncertainty to exclude other differentials, including primary malignancy or metastatic disease. The most common complication of surgical excision of myxomatous tissue is recurrence, occurring in 30% of cases with a median of 8.5 years (range: 1.9-16 years).

Case
A 74-year-old male presented to RS Bethesda Yogyakarta with complaints of swelling in the left leg accompanied by pain persisting for 3 days. The patient has a history of controlled hypertension and cholesterol, as well as a cardiac ring implantation in 2015. He was referred to the surgical department for further evaluation. The patient's chief complaint is severe pain with a scale of 5. On physical examination, the patient had a Glasgow Coma Scale (GCS) of E4V5M6, blood pressure of 96/66 mmHg, strong radial pulse with a frequency of 70 beats per minute, respiratory rate of 20 breaths per minute, body temperature of 36 °C, warm

extremities, and oxygen saturation of 100%. An electrocardiogram showed slight ST elevation inferiorly. Diagnostic imaging was performed with MRI of the ankle joint without contrast application using a multipurpose coil and STIR technique, with axial, sagittal, and coronal sections. Findings revealed normal bone structures of the ankle joint and foot (as far as visualized) with good signal intensity, no bone destruction, fractures, or dislocation of bone fragments. A cystic mass with septations measuring 1.9 cm x 2.7 cm x 4.8 cm was observed in the intramuscular flexor hallucis longus, well-defined and without apparent rupture of the Achilles tendon. Structures of the distal calf and foot muscles, especially the abductor and flexor digiti, showed no abnormalities.

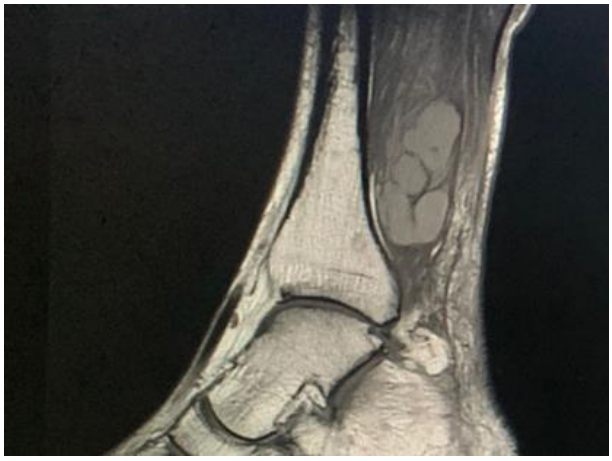

Fig 1: Sagittal T1-weighted imaging of the left ankle joint, showing normal bone structures of the ankle joint and foot, with no evidence of bone destruction, fractures, or dislocation of bone fragments

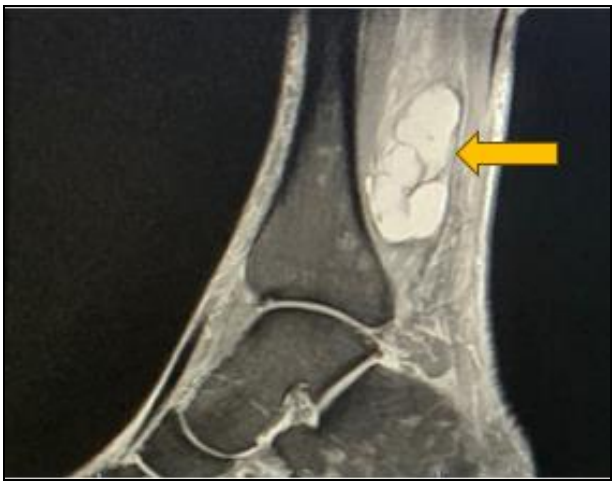

Fig 2: Sagittal T2 fat-saturated imaging of the left ankle joint, showing a well-defined cystic mass with septations in the intramuscular flexor hallucis longus, located in the posterior aspect of the ankle

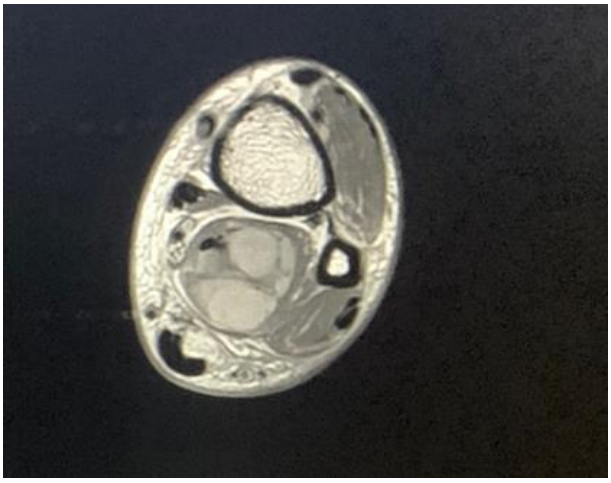

Fig 3: Axial T1-weighted imaging, showing no apparent rupture of the Achilles tendon

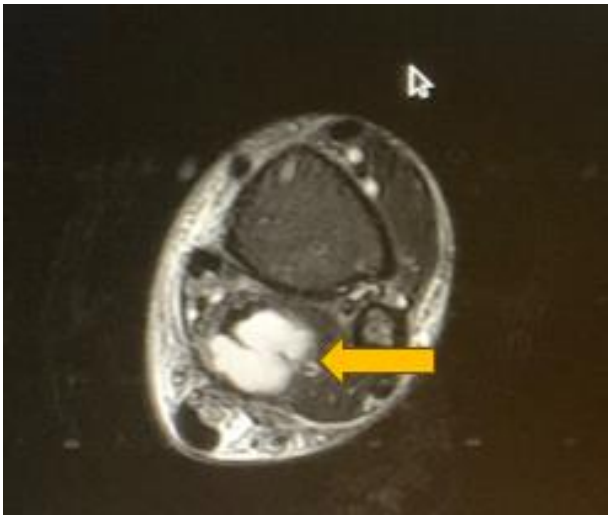

Fig 4: Axial T2-weighted imaging, showing a mass within the intramuscular flexor hallucis longus

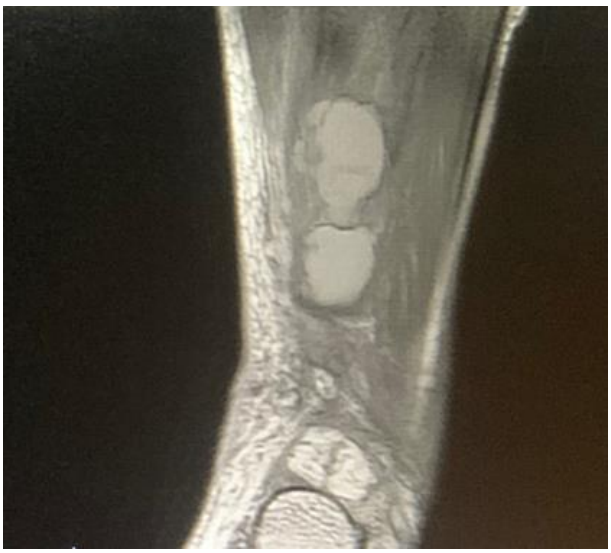

Fig 5: Coronal T1-weighted imaging, showing no involvement of surrounding structures

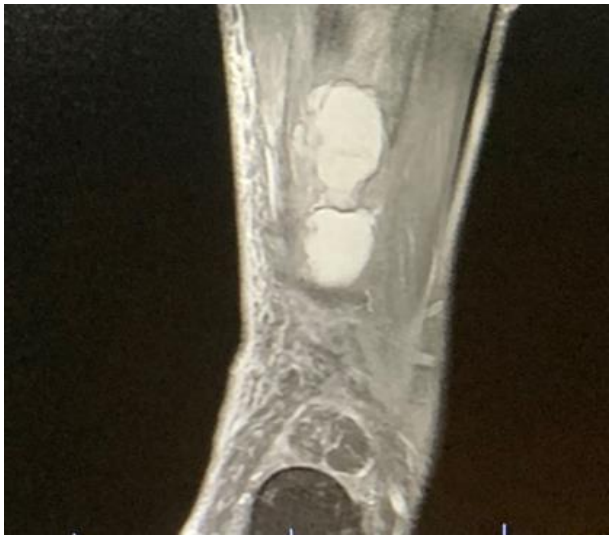

Fig 6: Coronal T2-weighted imaging, showing no intraarticular fluid collection.

Discussion

Intramuscular myxoma (IM) is a rare benign soft tissue tumor with unclear etiology and histology. It commonly occurs in the heart and rarely in the musculoskeletal system, with an incidence ranging from 0.1 to 0.13 per 100,000 [7]. IM imaging characteristics resemble those of other soft tissue tumors containing mucus, making misdiagnosis possible. Therefore, it is important to understand the pathological components and appropriate imaging features of IM, as well as to perform accurate imaging assessments for proper operative management [8].

IM often occurs in adults over 40 years old, rarely seen in children or adolescents, with a higher incidence in females than males. IM is typically solitary and generally larger in diameter. Also known as "Mazabraud syndrome," multiple lesions combined with Fibroproliferative bone disease can occur. IM often occurs in large skeletal muscle groups, such as the thigh, upper arm, calf, and buttocks [9]. Clinical manifestations of IM include a slowly growing non-specific mass in local muscles, generally without clear clinical symptoms and without specific laboratory tests. Sometimes patients may experience local pain if the mass causes pressure.

In the latest WHO 2013 soft tissue tumor classification, IM is defined as a benign "unclassified" tumor. Typically, IM has internal fibrous septations but rarely vascular structures. The pathological characteristics of IM can be summarized as few cells, few blood vessels, and a large amount of mucous stroma. Grossly, IM appears gelatinous and well-defined. The section surface of IM is sticky, smooth, and may have a pseudocapsule [10].

In the case, MRI showed a round cystic mass with clear borders. This is because IM contains a large amount of mucous stroma and mucopolysaccharides within it, which have good hydrophilicity, thus increasing water content in tumor tissue [11]. Internal septations and pseudocapsules can sometimes be seen in the tumor, mainly composed of immature collagen fibers and tumor cells [8].

The lesion appears with low signal intensity on T1-weighted imaging and high signal intensity on T2-weighted imaging with fat suppression. This is consistent with the pathological basis of a mucus-rich tumor stroma. Approximately 79%-100% of IMs show high signal intensity on DWI, which is also a manifestation of the pathological basis [12].

Sometimes, it is difficult to differentiate IM from myxoid tumors such as myxoid liposarcoma and myxoid fibrosarcoma on imaging. If well-differentiated myxoid liposarcoma and flocculent or linear fat signals can be seen in lesions like lipomas, decreased signal on fat suppression sequences will be helpful for identification [13]. However, when poorly differentiated myxoid liposarcoma lacks clear fat signals, it is more difficult to distinguish from IM. However, generally more malignant lesions are often accompanied by hemorrhage, necrosis, cystic changes, and show a clear uneven density increase after contrast-enhanced MR scanning. For myxoid fibrosarcoma, most lesions originate from the myofascial fascia. Because of compression from adjacent muscles and sarcoma constraints, a "tail sign" can be seen at the edge of the lesion [14].

Before surgical management, performing a biopsy guided by imaging can provide assistance in diagnosing the lesion. In this case, the lesion is relatively clear, with a clinical diagnosis tending towards benignity. Therefore, surgical management is recommended to be carried out directly to reduce further injury.

Conclusion

Intramuscular myxoma (IM) is a rare benign soft tissue tumor with unclear etiology and histology. It commonly occurs in the heart and rarely in the musculoskeletal system. The pathological characteristics of IM can be summarized as few cells, few blood vessels, and a large amount of mucous stroma. Lesions appear with low signal intensity on T1-weighted imaging and high signal intensity on T2-weighted imaging with fat suppression. In this case, the lesion is relatively clear, with a clinical diagnosis tending towards benignity.

Conflict of Interest

Not available

Financial Support

Not available

References

1. Petscavage-Thomas JM, Walker EA, Logie CI, Clarke LE, Duryea DM, Murphey MD. Soft-Tissue Myxomatous Lesions: Review of Salient Imaging Features with Pathologic Comparison. Radiographics. 2014;34(4):964-80. DOI:10.1148/rg.344130110
2. Majoor BCJ, van de Sande MAJ, Appelman-Dijkstra NM, Leithner A, Jutte PC, Vélez R, *et al*. Prevalence and Clinical Features of Mazabraud Syndrome: A Multicenter European Study. The Journal of bone and joint surgery. American volume. 2019;101(2):160-168. DOI:10.2106/JBJS.18.00062
3. Murphey MD, McRae GA, Fanburg-Smith JC, Temple HT, Levine AM, Aboulafia AJ. Imaging of soft-tissue myxoma with emphasis on CT and MR and comparison of radiologic and pathologic findings. Radiology. 2002;225(1):215-24. DOI:10.1148/radiol.2251011627
4. Kushchayeva YS, Kushchayev SV, Glushko TY, Tella SH, Teytelboym OM, Collins MT, *et al*. Fibrous dysplasia for radiologists: beyond ground glass bone matrix. Insights into imaging. 2018;9(6):1035-1056. DOI:10.1007/s13244-018-0666-6
5. Vescini F, Falchetti A, Tonelli V, Carpentieri M, Cipri C, Cosso R, *et al*. Mazabraud's Syndrome: A Case Report And Up-To-Date Literature Review. Endocrine, metabolic & immune disorders drug targets; c2018.

DOI:10.2174/1871530319666181226103700
6. Munksgaard PS, Salkus G, Iyer VV, Fisker RV. Mazabraud's syndrome: case report and literature review. Acta radiologica short reports. 2013;2(4):2047981613492532. DOI:10.1177/2047981613492532
7. Özbek N, Danaci M, Okumus B, *et al*. Recurrent intramuscular myxoma: review of the literature, diagnosis and treatment options. Turkish J Cancer. 2006;36:75-8.
8. Zhang Y, Huang J, Ding J. Imaging manifestations and pathological analysis of intramuscular myxoma: A case report and literature review. Radiol Case Reports [Internet]. 2021;16(2):300-4. Available from: https://www.sciencedirect.com/science/article/pii/S1930043320305690
9. Granel-Villach L, Alcalde-Sanchez M, Salvador-Marin M, *et al*. Differential diagnosis and management of intramuscular myxomas: A review of our experience. Cir Cir. 2017;85:356-60.
10. Bancroft LW, Kransdorf MJ, Menke DM, *et al*. Intramuscular myxoma: characteristic MR imaging features. AJR. Am J Roentgenol. 2002;178:1255-9.
11. Ginat DT, Meyers SP. Intracranial lesions with high signal intensity on T1-weighted MR images: differential diagnosis. Radiographics. 2012;32:499-516.
12. Petscavage-Thomas JM, Walker EA, Logie CI, *et al*. Soft-tissue myxomatous lesions: review of salient imaging features with pathologic comparison. Radiographics. 2014;34:964-80.
13. Sung M, Kang HS, Suh JS, *et al*. Myxoid liposarcoma: appearance at MR imaging with histologic correlation. Radio Graphics. 2000;20:1007-19.
14. Kikuta K, Kubota D, Yoshida A, *et al*. An analysis of factors related to the tail-like pattern of myxofibrosarcoma seen on MRI. Skelet Radiol. 2015;44:55-62.

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