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Imaging manifestations and pathological analysis of intramuscular myxoma: A case report and literature review ☆,☆☆

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ABSTRACT

Intramuscular Myxoma(IM) is a rare benign soft tissue tumor, and its etiology and histology source is still unclear. It is important to understand the pathological components of IM and its corresponding imaging features, as well as performing accurate and careful imaging assessments of IM before surgery. We present a case of a 43-year-old male who presented a lump in his left thigh and gradually enlarged during the past 8 years. The patient underwent CT, MRI, and CTA examined and was later pathologically confirmed as IM. This article will combine the literature,to explore the imaging manifestations and its pathological basis of intramuscular myxoma.

Introduction

Intramuscular myxoma (IM) is a rare benign soft tissue tumor, and its etiology and histology source is still unclear. Most myxomas occur in the heart and are rare in the musculoskeletal system, with an incidence rate ranging from 0.1 to 0.13 per 100,000 [1]. The imaging characteristics of IM are similar to other soft tissue tumors containing mucus, which can easily lead to misdiagnosis. Hence, it is important to understand the pathological components of IM and its corresponding imaging features, as well as performing accurate and careful imaging assessments of IM before surgery. Herein, we report the clinical, imaging, and pathological data of a case of IM of the left thigh, combining with related literature, to explore the imaging manifestations and its pathological basis of IM.

Case report

A 43-year-old male patient found a lump in his left thigh unintentionally 8 years ago. At that time, the lump was the size of a walnut with no tenderness and the patient had not gone to the hospital for examination. The lump was painless but gradually enlarged during the past 8 years. Now, the patient felt his left thigh sore and discomfort and was admitted to our hospital for treatment. The patient was in good physical condition with no special personal or family history. Specialist physical examination showed swelling in the inner side of the middle-lower segment of the left thigh, which measured about 12 × 14 cm with hard and clear boundary, a certain activity, normal skin temperature, no tenderness, no radial numbness, and no pulse. The left foot dorsal artery pulsation was present and the pathological reflex was not elicited of the

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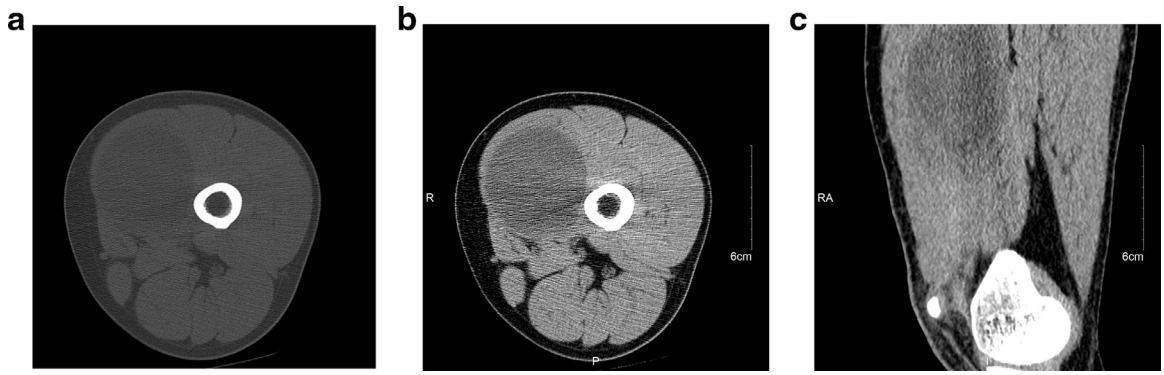

Fig. 1 – (A, B, C). CT plain scan transverse position (bone window, soft tissue window), sagittal position (soft tissue window) showed a type of oval low-density focus in the soft tissue of the medial segment of the left femur, with clear boundaries, uniform density, and plain scan The CT value was about 16HU.

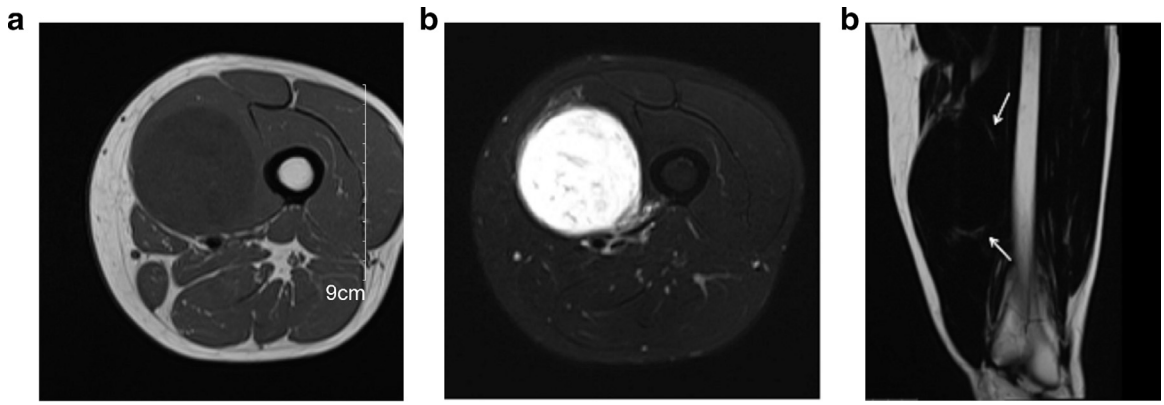

Fig. 2 – Transverse T1WI lesions showed a uniformly low signal, which was lower than the surrounding muscle tissue (A). The T2WI fat-suppressing sequence at the transversal position showed the high signal, the internal signal was not uniform, and no fat component was seen in the lesion, but a short low-signal shadow could be seen (B). Coronary PDWI-FS-Dixon fat image showed a circular high signal above and below the lesion, suggesting that there was a fat band around the lesion (white arrow) (C).

left leg. No obvious abnormality was found in the laboratory indicators.

Computed tomography (CT), magnetic resonance imaging (MRI), and computed tomography angiography (CTA) were performed after the patient was admitted to the hospital. A plain CT scan (Fig. 1) showed that an oval-shaped lower density lesion was seen in the anterior compartment of the thigh, beneath the vastus medialis muscle, with a uniform density of about 16HU. MRI (Fig. 2) showed that the mass had clear boundaries. On T1WI sequences the mass demonstrated uniform hypointensity and arc-shaped fat signals could be seen on the edge of the upper and lower mass on T1WI and PDWI fat images. On T2WI and fat suppression sequences the mass demonstrated hyperintensity and internal short hypointensity septations, signs of edema of the soft tissue around the mass were seen. Diethylenetriaminepentaacetic acid (Gd-DTPA) enhanced scanning showed uneven mild-moderate enhancement of the mass with the enhanced surrounding capsule. On sensitivity-weighted imaging (SWI) sequences, there were no obvious signs of bleeding or vascular malformations in the mass, and Pha-Images and SWI-Images (Siemens Avanto 1.5T) showed a few venous blood vessels in the mass (Fig. 3). The patient's CTA examination showed no significant abnormality of the left thigh artery blood vessels and branch vessels.

The patient underwent a left thigh tumor resection under general anesthesia. During the operation, the mass was found deep in the medial femoral muscle, adhering to the medial femoral muscle, large adductor muscle, and long adductor muscle with edema and mucus infiltration. The texture of the mass was hard and the capsule was complete with mucus. The entire mass was completely removed and sent for pathological examination.

The histopathological examination revealed a spindle cell tumor with extensive myxosis, considering IM (Fig. 4). No cell heterogeneity and mitotic images. No malignant

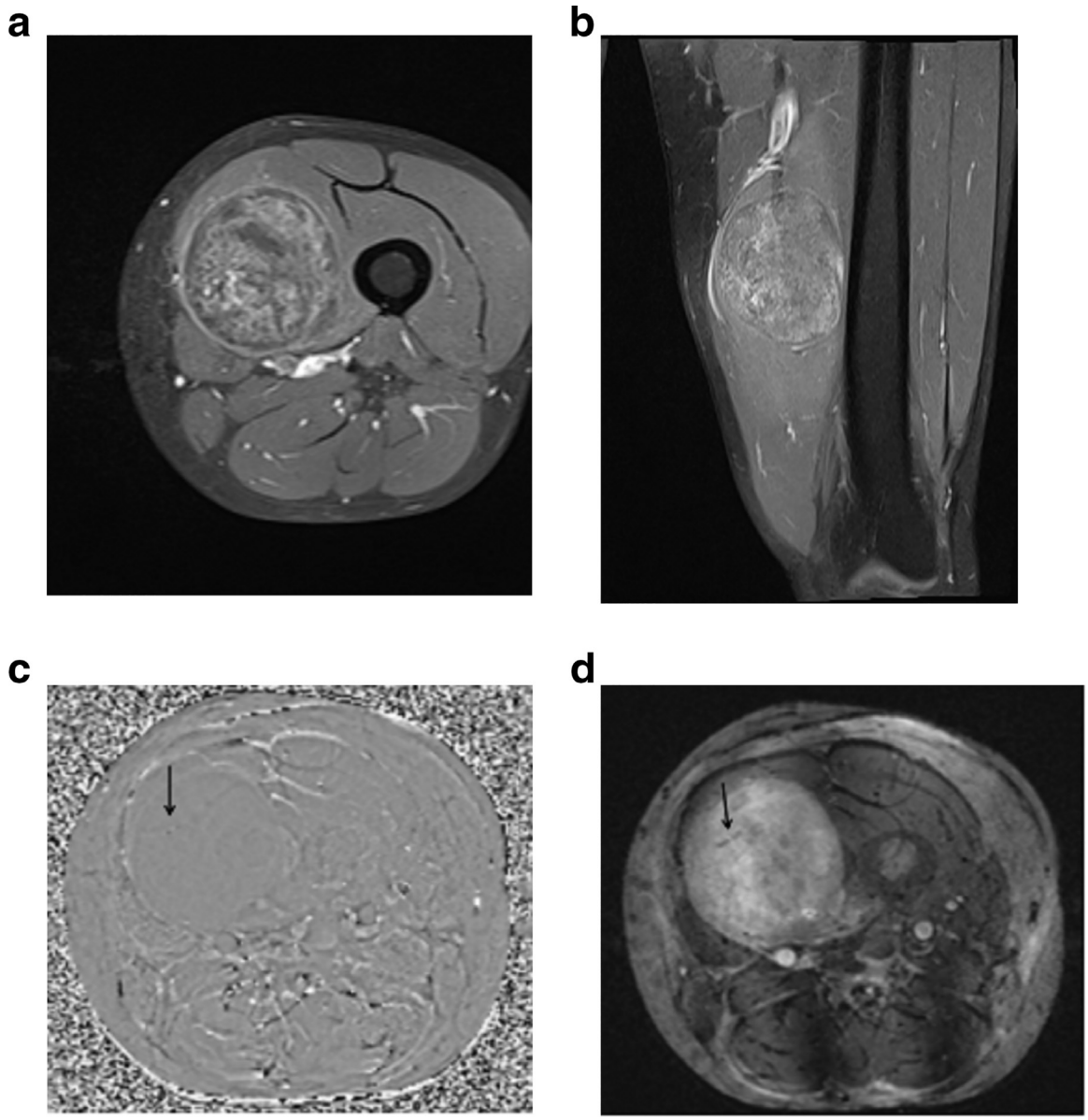

Fig. 3 – T1WI-enhanced scans of the transverse and sagittal positions showed circular discontinuous enhancement around the lesion, and flocculent, uneven, and mild enhancement within the lesion (A, B). Pha-Images and SWI-Images (Siemens Avanto 1.5T) showed a small amount of vascular structure in the lesion (black arrow) (C, D).

tendency. Immunohistochemical prompts: Vimentin (+), SMA (–), Desmin (–), S-100 (–), Ki67 (–), Syn (–), GFAP (–), and EMA (–).

Discussion

In 1871, Vichow first proposed the concept of myxoma, and in 1948 Stout clearly defined myxoma and proposed diagnostic criteria. In 1965, Enzinger first defined IM as a definite clinical-pathological lesion [2]. IM is currently indeterminate in etiology and is more common in adults over 40 years, rarely seen in children or adolescents. The incidence of females is higher than that of males. IM is usually single and generally larger in diameter. It's also known as "Mazabraud syndrome" when lesions are multiple and combined with bone Fibroproliferative disease. IMs often occur in large skeletal muscle groups, such as the thigh, upper arm, calf and buttock muscle groups. The locations in the muscle differ [3]. The common clinical manifestations of IM are the slow-growing, noncharacteristic masses in local muscles, generally without obvious clinical symptoms and with no special laboratory tests. Sometimes, patients can feel localized pain if the mass caused some oppression.

In the latest version of the 2013 WHO classification of soft tissue tumors, IM is defined as a benign "unidentifiable tumor." At present, many studies consider that IM may be de-

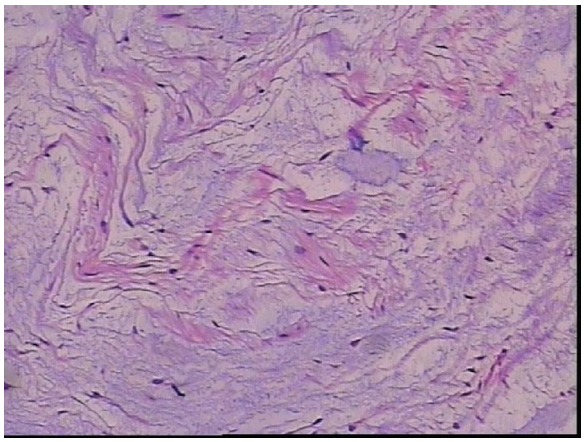

Fig. 4 – Pathological diagnosis (mass of left thigh) spindle cell tumor with extensive myxosis, considering intramuscular myxoma.

rived from primitive mesenchymal tissue, which has lost its ability to produce collagen but large amounts of hyaluronic acid and immature collagen fibers [4]. Therefore, IM may have fibrous septations internal but sparse vascular structure. The pathological characteristics of IM could be concluded as few cells, few blood vessels, and large amounts of mucus stroma. On gross examination, IM is jelly-like and well circumscribed. The cut surface of IM is sticky, slippery, and may have pseudocapsule [5]. Because of the pseudocapsule, the lesion may tend to infiltrate adjacent tissues, making surgery difficult to completely remove and tend to recur, but generally, it doesn't metastasize. Under the microscope, the tumor is mainly composed of a rich mucinous stroma, a small amount of undifferentiated stellate cells, spindle cells [6]. The tumor cell nucleus is small, consistent size, and deep staining with no atypia and mitosis. In recent years, some scholars have found that IMs contain areas with increased local cells, ranging from about 10% to 80%. The number of cells, vascular structure, and collagen in this area are increased, which may be pathologically misdiagnosed as sarcoma [7].

In our case, CT showed a round-like cystic density mass with clear boundaries and uniform density. This is because IM contains a large amount of mucus stroma and the mucopolysaccharide in it has good hydrophilicity, leading water content increased in tumor tissue [8]. Therefore, the density value of CT of IM is close to the cyst. Meanwhile, the density of most IMs is lower than the surrounding normal muscle tissue, which could easily be misdiagnosed as cystic lesions. Internal septations and pseudocapsule sometimes could be seen in the tumor, are mainly composed of immature collagen fibers and tumor cells [6].

In MRI, our lesion appeared as a soft tissue mass with a clear boundary, showing a uniform low-equal signal on T1WI sequences and high signal on T2WI and lipid-suppressing sequences. This is in line with the pathological basis of the tumor's rich mucus stroma. Line-shaped low-signal internal septations could also be seen on T2WI sequences. Edema signals could obviously be seen around the lesion on T2WI-FS and PDWI-FS sequences. This is the manifestation of edema presented by adjacent muscle tissue which stimulated by the invading mucus for a long time because most tumors have no capsule or only pseudocapsule [6]. About 79%-100% of the IM shows a high signal on the DWI sequences, which is also the manifestation of this pathological basis [9]. Meanwhile, the extravasated mucus stroma could also cause the atrophy of adjacent muscles and reactive fat deposits. On MRI, it appears as a thin layer of fat signal ("fat band") located around the tumor and a cap-shaped fat signal ("fat cap") located above and below the tumor's long axis [5]. In our case, short arc-shaped ultra-thin fat signals could be seen at the upper and lower of the lesion on T1WI sequences, which was consistent with the above MRI imaging characteristic. Magnetic SWI is highly sensitive to vein structure, bleeding, and iron deposition, which can be used to reveal the microstructure and composition of tumors. At present, there are no reports of IM's SWI-related research at home and abroad. In our case, the patient performed an SWI scanning after the MRI enhanced scanning. According to Pha-Images and SWI-Images display, a small amount of vascular structure could be seen in the lesion, according with IM's oligovascular pathological feature.

After Gd-DTPA enhanced scanning, the IM lesion in our case showed a marginal ring-shaped enhancement and internal uneven mild enhancement. short strip-like septation enhancements and no obvious enhancement areas were also visible in the lesion. Combined with pathology, the marginal ring-shaped enhancement and internal septation enhancements are mainly the performance of pseudocapsule and internal fibrous separation after enhancement, showing mild-moderate enhancement. Because IM is a solid lesion instead of a true cystic lesion, the solid components and areas rich in vascular structure will be relatively obviously enhanced after scanning. While the areas with rich mucus stroma and sparse vascular structure will be relatively insignificantly enhanced. This is the pathological basis of IM's uneven enhancement [10]. Luna et al. [6] summarized IM's enhancement into 3 types: (1) marginal annular-enhancement with internal septation-like enhancement, which is more common. (2) marginal annular-enhancement with an internal uneven or progressive enhancement: Part of the IMs can be shown as a gradual enhancement way, with mildly enhanced in the arterial phase, further enhancing in the venous phase and continuous enhancement in the delay phase. This progressive enhancement way may be related to a large amount of mucus stroma in the tumor. Since the vascular structure in the area rich in mucous stroma is scarce, and the mucus can block the perfusion of the contrast agent, the arterial phase could only show the enhancing of a few vascular structures in the tumor, which shows slight enhancement. In the venous phase, the degree of enhancement will strengthen with the continuous accumulation of contrast agent in the extracellular space. In the delay phase, because the mucus stroma delays the clearance of the contrast agent, the enhancement will be persistent [11–13]. (3) Simple marginal annular-enhancement: this type is rare.

In addition to CT and MRI, ultrasound also has a certain diagnostic value for IM. IM often appears like a substantial hypoechoic mass in ultrasound. Doppler blood flow imaging could sometimes detect point or rod-like blood flow signals. Adjacent muscles may be mild atrophy, edema, and fatty infil-

tration, showing intermittent linear hyperechoes around the tumor—"bright ring signs" in ultrasound. Fatty infiltration and edema can also be seen in the muscle space at both ends of the tumor's long axis, which called "bright hat sign" in ultrasound. "bright ring sign" and "bright hat sign" are most typical ultrasound manifestations of IM [14]. It was a pity that our patient didn't take an ultrasound test before surgery.

Fluorine-18 fluorodeoxyglucose positron emission tomography-computed tomography(F-18 FDG PET-CT) is not often used to diagnose IM. There are a few case reports demonstrating F-18-FDG uptake in IM showing low-grade F-18 FDG uptake with standard uptake value (SUV) values between 1.3 and 2.6 [15]. At present, the mechanism of uptake is uncertain but may reflect a varying proportion of metabolically active cells [16]. One case demonstrated the possible role of F-18 FDG PET-CT scan to detect postoperative residual IM [15].

Preoperative diagnosis of IM is relatively difficult, and it is easy to be misdiagnosed as a cystic lesion when only performing CT or MR plain scanning. In addition, to be distinguished from neurogenic tumors such as schwannomas and neurofibromas is also necessary. Sometimes, it's difficult to distinguish IM from myxoid tumors such as myxoid liposarcoma and myxoid fibrosarcoma in imagings. If myxoid liposarcoma is well-differentiated and flocculent or linear fat signal could be seen in the lesion [13] like a lipoma, the decreased signal on lipid-suppressing sequences will be helpful for identification. However, when myxoid liposarcoma is poorly differentiated or has no obvious fat signals, it is more difficult to distinguish from IM, but the lesion is generally more malignant, often accompanied by bleeding, necrosis, and cystic changes, and shows obvious uneven enhancement after the MR enhancement scan. As to myxoid fibrosarcoma, most lesions are originated from the myofascial fascia. Due to the compression of the adjacent muscles and the limitation of the sarcolemma, "tail signs" could be seen at the edge of the lesion [17]. The lesions often have unclear boundaries, uneven density or signal accompanied by hemorrhage and necrosis, and show obviously uneven enhancement. Before surgical resection, performing an imaging-guided biopsy will provide certain help for the diagnosis of the lesion. As the boundary of our case's lesion was relatively clear, the clinical diagnosis tended to be benign. So direct surgical resection was taken to reduce the injury of multiple operations.

Summary

In conclusion, IM is a rare benign soft tissue tumor. Preoperative CT and MRI examinations have good diagnostic value for IM, especially MRI has characteristic features such as "fat band" and "fat cap". Therefore, when a mass with a clear boundary and insignificant clinical symptoms is found in the skeletal muscle (especially in the large muscle group of the extremities), the possibility of IM should be considered when the abovementioned imaging manifestations appear, but it still needs to be compared with other lesions containing rich mucus to make a differentiation. A surgical excision as soon as possible to confirm the lesion's pathological diagnosis is recommended.

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