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Intramuscular Myxoma of the Gluteus Maximus: Diagnosis and Management of a Rare Benign Tumor: A Case Report

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Abstract: **Case Report**

Background: Intramuscular myxoma (IM) is a rare benign soft-tissue tumor, frequently underrecognized due to its subtle clinical presentation and radiological/histopathological overlap with cystic lesions or low-grade myxoid sarcomas. Accurate diagnosis requires multidisciplinary correlation of imaging, histopathology, and clinical context, particularly to exclude associated syndromes such as Mazabraud syndrome. ***Case presentation***: We report the case of a 56-year-old Moroccan woman with no significant family history, who presented with a painless, slowly progressive mass in the right gluteal region evolving over four years. Clinical examination revealed a mobile, non-tender swelling measuring 4 × 5 cm. Ultrasound and MRI identified a 12.7 cm multiloculated solid-cystic lesion within the right gluteus maximus, extending beyond the aponeurosis. Initial surgical biopsy suggested a synovial cyst. Subsequently, a wide monobloc surgical excision was performed. Histopathological examination confirmed benign IM, characterized by a hypocellular myxoid stroma with sparse spindle cells, absence of mitoses or nuclear atypia, and clear resection margins. Systemic skeletal survey excluded fibrous dysplasia. At one-year follow-up, the patient remained asymptomatic with no evidence of recurrence. ***Conclusion***: This case underscores the diagnostic challenges posed by IM, often misinterpreted as a benign cystic lesion. Complete surgical excision, coupled with thorough histopathological analysis, remains the cornerstone of management and ensures an excellent long-term prognosis.

Keywords: Intramuscular myxoma, Gluteus maximus, Benign soft tissue tumor, Magnetic resonance imaging (MRI).

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INTRODUCTION

Intramuscular myxoma (IM) is a rare benign soft-tissue tumor of mesenchymal origin, characterized by an abundant hypocellular myxoid matrix. First described by Stout in 1948, it is the most common subtype of benign myxoma arising within skeletal muscle, with a predilection for the lower limbs, especially the thigh and gluteal regions. Although strictly benign and devoid of metastatic potential, IM poses a significant diagnostic challenge due to its clinical, radiological, and histopathological similarities with benign cystic lesions (e.g., synovial cyst) or, more critically, with low-grade myxoid sarcomas. Magnetic resonance imaging (MRI) is a cornerstone of the preoperative workup; however, definitive diagnosis relies on histopathological examination, ideally performed on a complete excision specimen. Furthermore, the occurrence of multiple lesions warrants systematic investigation for Mazabraud syndrome, a rare association of intramuscular myxomas and osseous fibrous

dysplasia. We herein report a case of IM of the gluteus maximus, initially misdiagnosed, to illustrate its diagnostic pitfalls, therapeutic strategy, and favorable outcome following complete surgical excision.

Case Presentation

We report the case of a 56-year-old Moroccan woman, mother of two, with no significant personal or family medical history, except for a total thyroidectomy performed four years earlier for benign disease, under levothyroxine replacement therapy. Her symptoms began approximately four years prior with the gradual onset of a painless, mobile mass in the right gluteal region, slowly increasing in size, in an afebrile context with preserved general health.

General examination revealed a conscious, hemodynamically and respiratory stable patient. Local examination identified a mobile, non-tender mass measuring 4 × 5 cm, with no overlying skin changes or functional impairment. No palpable lymphadenopathy was detected, and the remainder of the physical examination was unremarkable.

Soft-tissue ultrasound demonstrated a well-defined, lobulated, predominantly anechoic lesion deep to the fascia of the right gluteal region, containing a solid isoechoic component. The lesion was avascular on color Doppler, exhibited thin internal septations, and extended into the subcutaneous fat.

Complementary MRI revealed a 127.2 × 66.6 × 96.6 mm multiloculated solid-cystic intramuscular mass involving the right gluteus maximus, crossing the aponeurosis and extending into the subcutaneous tissue. It comprised a multiseptated cystic component and a solid component showing T1 hypointensity and T2 hyperintensity, with post-contrast enhancement. No morphological or signal abnormalities were observed in the ipsilateral femur.

An initial surgical biopsy was performed; histopathological analysis favored a diagnosis of synovial cyst, with no features of malignancy.

Management included hospitalization, a complete biological workup, including thyroid function tests, and pre-anesthetic assessment. Surgical resection was performed under general anesthesia via a Kocher–Langenbeck approach. A wide monobloc excision of the tumor was carried out, ensuring macroscopically clear margins. The mass, located within the muscular body of the gluteus maximus, was oval-shaped, firm in consistency, surrounded by a hard, whitish pseudocapsule, and measured approximately 7 cm along its greatest axis (Figures 3 and 4). The postoperative course was uneventful, with drain removal on postoperative day 2 and discharge on day 3. Postoperative functional rehabilitation was prescribed.

Macroscopic examination of the excised specimen revealed a 310 g mass measuring 11.5 × 9.5 × 2 cm. Deeply, it contained a posteriorly located, encapsulated myxoid tumor, multilocular, filled with mucoïd material, and featuring a central blackish hemorrhagic area. Resection margins were precisely measured: synovial margin at 3 mm, anterior margin at 70 mm, inferior margin at 80 mm, and superior margin at 50 mm. Additionally, a poorly formed, thrombosed, tortuous angiomatous and fibrous zone was identified in the subcutaneous tissue.

Microscopically, the tumor consisted of thin spindle cells dispersed within a transparent, mucoïd, hypocellular myxoid stroma. Overall, cellularity was low. Peripherally, a fibrous fascial layer and atrophic muscle fibers were observed. No mitotic figures or nuclear atypia were identified. All resection margins were tumor-free. Taken together, these features were consistent with a diagnosis of benign intramuscular myxoma.

Regular clinical follow-up was performed at 3, 6, and 12 months postoperatively. At one year, no local recurrence was observed.

Figures 1: A preoperative clinical photograph showing the right gluteal mass

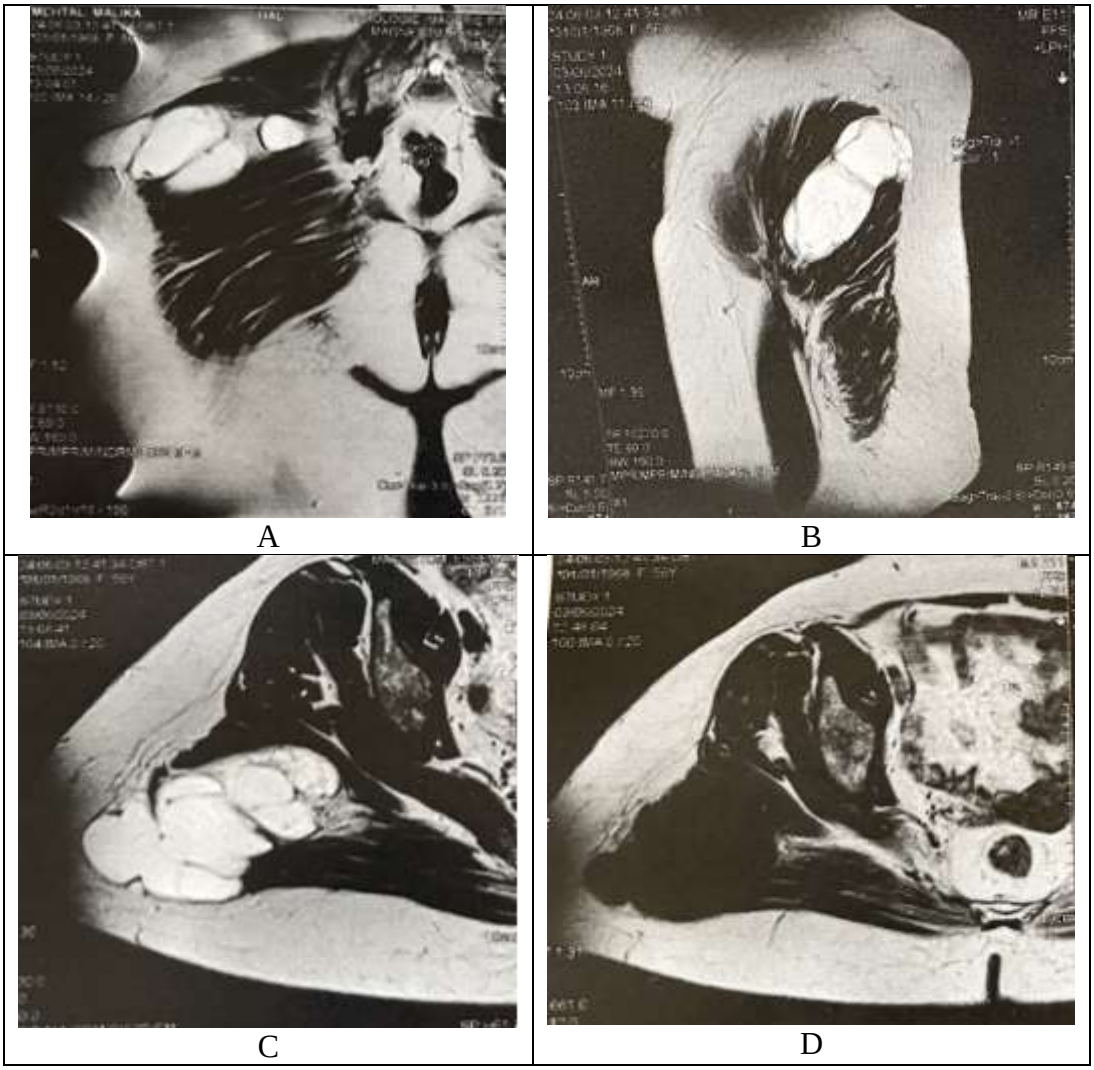

Figure 2: MRI images showing the right gluteal intramuscular myxoma as a multiloculated solid-cystic mass, exhibiting T1 hypointensity (D) and T2 hyperintensity (A, B, C) relative to the surrounding right gluteus maximus muscle: (A) Coronal MRI section; (B) Sagittal MRI section; (C, D) Axial MRI section

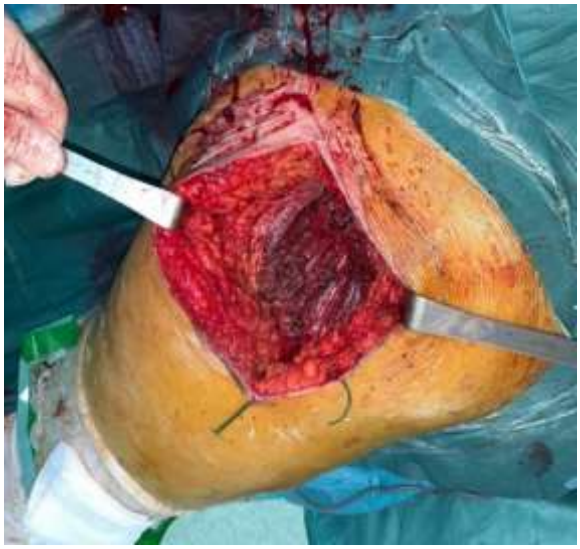

Figure 3: Intraoperative view following tumor excision

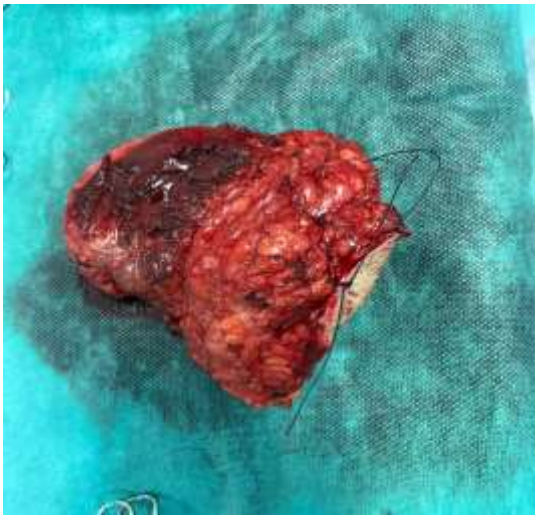

Figure 4: Intraoperative view showing the solid-cystic appearance of the tumor, extending into the subcutaneous fat

Figure 5: Immediate postoperative photograph

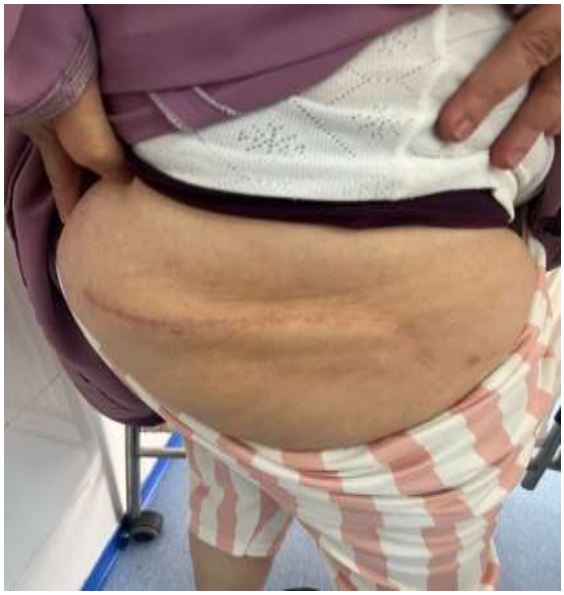

Figure 6: One-year follow-up clinical photograph

DISCUSSION

Intramuscular myxoma (IM) represents a rare nosological entity among benign soft-tissue tumors, characterized by a mesenchymal proliferation localized within skeletal muscle. Although initially described by Rudolf Virchow in 1871 as a "mucoïd tumor" in reference to the umbilical cord's gelatinous substance, it was Stout's work in 1948 that defined its hallmark histological features: an abundant extracellular matrix, poor in cellular elements, rich in mucopolysaccharides (notably hyaluronic acid), and exhibiting scant vascularity [1, 5]. IM is now recognized as the most common benign soft-tissue myxoma, within a heterogeneous spectrum that includes superficial myxoma, aggressive angiomyxoma, myxolipoma, and acral fibromyxoma [2, 3, 4].

IM predominantly affects adults aged 40–70 years, with a reported female predominance of 57–80% [1, 2, 8]. Although exceptionally rare in children, cases in patients as young as 2–5 years have been documented [9, 10], indicating that age alone should not exclude IM from consideration [8]. Its estimated incidence is 0.1–0.13 per 100,000 individuals [2].

The cellular origin remains debated: hypotheses include derivation from poorly differentiated fibroblasts unable to synthesize collagen yet capable of mucin production, or from primitive mesenchymal stem cells [1, 4, 5]. Recent evidence implicates post-zygotic GNAS mutations; however, molecular testing sensitivity varies (29 to 90%), and access remains limited, making histopathology the diagnostic gold standard.

IM typically presents as a painless, slowly progressive mass over months to years [1, 2, 4, 6]. Pain occurs in <30% of cases, usually late and secondary to mass effect [1, 7], and is notably absent in our patient. Tumor size ranges from 2 to 17 cm, with no clear symptom correlation [1, 8].

The thigh is the most common site (51%), followed by the arm (9%), the calf (7%), and the gluteal muscles (7%), as in our case. Rare locations include the neck or hand; thenar involvement is exceptional [1, 2, 4, 6].

Critically, multiple IMs warrant evaluation for Mazabraud syndrome (IM + fibrous dysplasia) or McCune–Albright syndrome (polyostotic fibrous dysplasia, café-au-lait spots, endocrine hyperfunction), both linked to GNAS mutations [2, 5, 6, 8]. Our

patient's skeletal survey was negative, excluding these syndromes.

Radiographs are often normal or show non-specific soft-tissue opacity [1, 5, 6]. Ultrasound reveals a hypoechoic, well-defined, minimally vascular mass [1, 4, 5, 6]. CT shows a homogeneous, low-density lesion [1, 5, 6].

MRI is diagnostic: IM appears as a T1-hypointense, T2-hyperintense intramuscular mass, often with perilesional edema. Secondary signs include the fat rim sign (peripheral fat from muscular atrophy), bright rim sign (T2 peripheral hyperintensity), and bright cap sign (proximal triangular fat on sagittal views) [1, 2, 4, 5, 6]. Despite this, IM is frequently misdiagnosed as a synovial cyst, as occurred initially in our patient, or, more seriously, as myxoid liposarcoma, myxofibrosarcoma, extra skeletal myxoid chondrosarcoma, or low-grade fibromyxoid sarcoma (LGFMS) [1, 2, 4, 5, 6, 8].

Definitive diagnosis requires histopathological evaluation. Fine needle aspiration is often inadequate due to hypocellularity; surgical excision is preferred [4, 5, 6]. Macroscopically, IM appears gelatinous, grayish, and multilobulated [4, 5, 6].

Microscopically, it shows a hypocellular, hypovascular myxoid matrix with bland spindle/stellate cells, no atypia, and no mitoses [1, 2, 4, 5, 6]. Immunohistochemically, it is positive for vimentin and CD34, negative for S100, and, critically, negative for MUC4, distinguishing it from MUC4-positive LGFMs [2, 5].

Complete surgical excision is standard. Although IM appears well-circumscribed macroscopically, it microscopically infiltrates the surrounding muscle [1, 8]. Recurrence is rare (<10%) and linked to incomplete resection or hypercellular variants [1, 4, 5, 6, 8].

Two approaches exist:

Wide resection (as performed in our patient), which minimizes recurrence risk but may cause functional morbidity [8].

Marginal resection is sufficient when the diagnosis is confirmed multidisciplinarily [8]. Recent data suggest comparable recurrence rates [8].

No metastases have ever been reported, confirming IM's benign nature [1, 5]. Nevertheless, in cases of diagnostic uncertainty, wider margins or reoperation if malignancy is histologically confirmed remain warranted [8].

CONCLUSION

Although rare and benign, intramuscular myxoma poses a significant diagnostic challenge due to its radiological and histopathological similarities with malignant myxoid tumors. An integrated, multidisciplinary approach combining meticulous clinical evaluation, high-quality MRI, rigorous histopathological analysis (ideally on a complete excision specimen), and systematic screening for syndromic associations is essential to avoid misdiagnosis and inappropriate management.

This case underscores the critical importance of such collaboration: the initial misinterpretation as a synovial cyst highlights how easily IM can be overlooked without comprehensive correlation of findings.

Surgical excision remains the treatment of choice. Current trends favor more conservative resection strategies, provided the diagnosis of benignity is firmly established. Moreover, advancing insights into the molecular underpinnings of IM, particularly GNAS mutations, holds promising potential for the future development of more precise, less invasive diagnostic tools.

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