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Intramuscular myxoma of the soleus muscle: A rare tumor in an unusual location

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Female, 52
Final Diagnosis: Myxoma
Symptoms: —
Medication: —
Clinical Procedure: —
Specialty: Oncology

Objective: Challenging differential diagnosis

Background: Intramuscular myxoma is a benign intramuscular neoplasm. However, sometimes it is difficult to distinguish it from soft-tissue sarcomas that underwent myxomatous degeneration. To the best of our knowledge, only 2 cases of intramuscular myxoma in the soleus muscle have been previously reported.

Case Report: We present the case of a 52-year-old Caucasian woman who was referred to our tertiary anticancer hospital for magnetic resonance imaging (MRI) diagnosis for suspicion of mesenchymal tumor. Percutaneous core biopsy revealed an intramuscular myxoma (IM). Despite the benign nature of the lesion, the patient desired that the tumor be removed. She underwent wide local excision. The final histopathologic diagnosis was IM. She is doing well 3 years after the operation.

Conclusions: Percutaneous core biopsy is the procedure of choice for providing preoperative tissue diagnosis. The treatment of choice is wide local excision. IM does not usually recur after surgical removal.

MeSH Keywords: myxoid tumours • soft tissue tumour • Myxoma

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Background

IM is a rare benign neoplasm confined to skeletal muscles. Classically, it is described by bland spindle-shaped and stellate cells embedded in hypovascular myxoid stroma [1].

Case Report

A 52-year-old woman consulted her family physician about a slow-growing, firm, mildly tender lump on the posterior aspect of her left leg, detected by herself 6 months before. Her past medical history was unremarkable. For further characterization of the tumor, an MRI was ordered, which showed on T1-weighted imaging a well-defined hypointense intramuscular lesion in the soleus muscle, and hyperintense on T2-weighted imaging (Figure 1). The patient was referred to our department for further investigation with the suspected diagnosis of mesenchymal tumor. Percutaneous core biopsy revealed IM. She underwent wide local excision of the lesion and recovered well. The final histopathologic diagnosis was intramuscular myxoma (Figures 2 and 3). She is free of recurrence 3 years after the intervention.

Discussion

In 1863, Virchow coined the term “myxoma” to describe a tumor histologically resembling the umbilical cord [1].

In 1948, Stout outlined the diagnostic criteria and defined myxoma [2]. He studied 142 cases (exclusive of those lesions within the heart) and found only 3% of the tumors to be intramuscular. In 1965, Enzinger reported that from 200 studied myxomas, IM cases were approximately 17% [3].

The incidence rate of IM varies from 0.1 to 0.3 per 100,000 population [4]. In descending order of appearance, the tumor is most frequently located in the large muscles of the thigh (51%), shoulder girdle, buttocks, and upper extremity [1]. Soleus muscle is an uncommon site of appearance, accounting for only 2 reported cases (Table 1). Therefore, our case is the third.

The age at presentation is 40–70 years and the female prevalence is about 66% [1]. IM usually appears as an isolated mass except in the case of Mazabraud syndrome, in which they are multiple and in conjunction with fibrous dysplasia of the bones; in these cases myxomas tend to occur near areas of the affected bones [5]. The tumor is usually detected by the patient as a round-shaped, painless, slowly growing mass. Patients may report histories from a few months to a few years in duration.

Histologically, IM is characterized as a hypocellular hypovascular tumor composed of spindle-shaped and stellate cells embedded

Figure 1. Well-defined homogeneous hypodense intramuscular lesion on T1 weighted imaging and hyperintense on T2 weighted imaging.

Figure 2. Intramuscular myxoma (hematoxylin & eosin ×40).

Figure 3. Intramuscular myxoma (desmin ×40).

in an abundant myxoid stroma [1]. There is absence of a true capsule; it possesses only an incomplete pseudocapsule [6].

Focal areas of hypervascularity and hypercellularity make IM difficult to differentiate from other myxoid soft-tissue tumors

Table 1. Published cases of Intramuscular Myxomas in the soleus muscle.

1. Enzinger FM. Am J Clin Pathol, 1965;43: 104–113
2. Travers E et al. Surg Chronicles, 2011; 16: 123–124

such as myxoid liposarcoma, low-grade myxofibrosarcoma and low-grade fibromyxoid sarcoma [6,7]. In immunohistochemical stains, IM cells stain positively for vimentin and show variable staining for CD34 and actin; immunostain for S-100 protein is characteristically negative, unlike myxoid liposarcoma [7].

The recent discovery of GNAS 1 mutations has helped to increase the specificity in the diagnosis of IM [8,9]. GNAS 1 mutations have been identified in IM with and without fibrous dysplasia [8,9].

On computed tomography (CT), IM is shown as a homogenous low-attenuated mass [1]. Magnetic resonance imaging (MRI) is the imaging modality of choice for the preoperative diagnosis of IM. It most commonly appears hypointense on T1-weighted MRI and hyperintense on T2-weighted MRI [6]. The most distinctive features of IM are the presence of a peritumoral fat rind visible on T1 and an increased signal in the adjacent muscle on T2-weighted or fluid-sensitive MR sequences [6]. In the majority of cases, the presence of peritumoral fat is more prominent at the superior and inferior poles of the tumor. This characteristic appearance is called the “Bright caps sign” [10] (Figure 4).

The differential diagnosis of IM includes aggressive angiomyxoma, myxoid neurofibroma, myxoid liposarcoma, low-grade myxofibrosarcoma, cellular myxoma, juxta-articular myxoma, and nodular fasciitis [1,6,7].

The worst-case scenario is a possibility misdiagnosis with malignant neoplasm. Enzinger et al reported the case of a 48-year-old man with soleal IM who went on to have an amputation for the lesion [3].

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Figure 4. Characteristic “bright caps sign” on the poles of the tumor.

Percutaneous core biopsy is the most useful procedure for establishing preoperative tissue diagnosis.

To the best of our knowledge, based on PubMed and Scholar Google searches, this is the third case of an IM of the soleus muscle reported in the literature.

Conclusions

Despite the benign nature of IM, the treatment of choice is wide local excision. Histopathologic diagnosis should be reviewed in case of recurrence.

Conflict of interest

None.