Intramuscular Myxoma

Lily Zou, MD
Department of Radiology, Robert Wood Johnson Medical School
University of Medicine and Dentistry of New Jersey
New Brunswick, NJ

Clinical History

48-year-old female presented with left thigh pain due to recent fall. Palpable mass was detected on physical exam.

Figures

Figure 1. Grey scale ultrasound image of the palpable mass in the left thigh demonstrates a well-circumscribed intramuscular lesion displacing and distorting muscle fibers with heterogeneously hypoechoic echotexture and a peripheral hyperechoic rim. There is slight posterior acoustic enhancement (arrow).
Figure 2. Transverse grey scale ultrasound image demonstrates echogenic triangles at the poles, compatible with a “bright cap sign” (arrow), consistent with a peripheral rim of fat.
Figure 3. Power Doppler ultrasound image reveals the mass to be essentially avascular.
Figure 4. MRI of the Left femur with and without contrast. A) T1-weighted pre-contrast coronal MRI shows a well-circumscribed lesion in the vastus medialis muscle (arrow) with a rim of fat at the superior and inferior poles (arrowheads). B) T1-weighted post-contrast coronal MRI showing homogeneous enhancement with hypointense center (arrow). C, D) Axial and sagittal STIR MRI demonstrate a well-circumscribed mass lesion (arrow).

Diagnosis

Final pathology revealed intramuscular myxoma.
Discussion

Intramuscular myxomas are rare, benign, slow-growing tumors of connective tissue origin with a longitudinal length ranging from 2-15cm [1, 2] and an estimated incidence of 0.11 per 100,000 [3]. These lesions are more commonly found in large muscles such as the gluteal muscles and the muscles of the shoulder girdle and upper arm, but they are most often observed in the intramuscular compartment of the thigh [4, 5]. The typical age at presentation is 40-70 years and there is a female preponderance of approximately 66%. The lesions are usually solitary except in the case of Mazabraud syndrome in which they occur in conjunction with skeletal fibrous dysplasia-and are often found near the affected bones [1, 6-8]. It is important to recognize these lesions and differentiate them from malignant intramuscular lesions with similar radiographic appearance as diagnosis clearly affects the prognosis and patient management.

The ultrasound images demonstrate a well-circumscribed complex intramuscular lesion that is heterogeneously hypoechoic with mildly increased through transmission and echogenic triangles at the poles of the lesion. Internal vascularity is absent and there is a central hypoechoic area within the mass. MRI shows a heterogeneous intramuscular lesion with low signal intensity on pre-contrast T1WI MR and a “bright rim” of fat as well as “bright caps” prominent at the superior and inferior poles of the lesion. The lesion is avascular as demonstrated by mild post-contrast enhancement of the lesion with central sparing. There is high signal intensity on STIR sequence with a surrounding fat rim.

Histopathologically, myxomas possess an abundance of noncollagenous mucinous stroma with a paucity of cells, low vascularity and angiogenesis and minimal mitotic figures. Of diagnostic significance, they typically are devoid of a capsule, possessing only an incomplete pseudocapsule. It is thought that this architecture allows mucoid material from the tumor to infiltrate into the adjacent tissue leading to muscular atrophy, fatty deposition and edema [2, 4]. The characteristic “bright rim” and “bright cap” signs observed on sonography and MRI are thought to result from the aforementioned tissue deposition.

On sonography, the “bright rim sign” seen on MR corresponds to a peripheral rim of increased echogenicity while the “bright cap sign” is visualized as a triangular hyperechoic area found at least at one of the poles of the lesion [2]. The tumors typically appear as complex oval hypoechoic lesions with a heterogeneous echotexture and frequent posterior enhancement. Furthermore, the lesions often contain cysts. Nonetheless, intramuscular myxomas can be differentiated from simple cysts based on their intramuscular location in comparison to the juxtaposition of joints as seen on ultrasound evaluation of simple cystic lesions such as ganglia and bursas [2, 4, 9].
MRI is the modality of choice for the radiologic diagnosis of intramuscular myxoma, although ultrasound can be very suggestive. The most distinctive features include the previously discussed bright rim and bright cap signs, a complete or partial capsule, intratumoral cysts and the presence of edema in adjacent muscles [4, 10]. MRI of the lesions often shows a cystic appearance that is mostly homogeneous. Specificity for the diagnosis of the intramuscular myxomas can be improved by the use of intravenous contrast which will demonstrate both peripheral and internal enhancement secondary to the pseudocapsular pathology characteristic of these lesions [10].

Intramuscular myxomas must be distinguished from myxoid liposarcomas—much more ominous diagnosis. The latter tend to demonstrate more vascularity as well as echogenic fat within the lesion proper [2]. On the other hand, intramuscular hemangiomas are vascular intramuscular lesions typified by hypoechoic flow channels, hyperechoic fat and posterior acoustic shadowing when there are intralesional phleboliths [2].

Intramuscular myxomas are thought to possess no risk of malignancy and minimal risk of recurrence which is highest in the cellular myxoma subtype. However, this remains controversial in the literature. Patients usually present with a palpable, often painless mass but as the size of the lesion increases, compression of underlying structures may precipitate a plethora of clinical symptoms including pain and weakness. Despite its benign nature and indolent course, current recommendations are for surgical removal of intramuscular myxomas with wide surgical margins since there is characteristic microscopic peripheral infiltration into the surrounding muscle (despite the well-delineated gross appearance) which leaves open the small possibility for recurrence [10]. Perhaps more importantly is the fact that there is a distinct possibility for immunochemical misdiagnosis with malignant lesions.

References


Authors
Lily Y. Zou, MD
Desmond A. Brown, PhD
Albert C. Li, MD (Mentor)

Institution
Robert Wood Johnson Medical School
University of Medicine and Dentistry of New Jersey
New Brunswick, NJ