J Musculoskelet Neuronal Interact 2015; 15(4):372-374

Hylonome

Clinical Quiz

A rare intramuscular myxoma of the hypothenar

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Keywords: Myxoma, Intramuscular, Tumor, Hypothenar

Case

A 72 year old man was admitted to our department with a nine month history of gradually enlarging mass in the volar aspect of the hypothenar area of his left hand. The patient reported that the size of the mass had been increasing during the past 3 months. As far as the rest medical record is concerned he was under medication for diabetes mellitus type 2 with metformin and he had undergone an operation for colorectal malignancy 6 years before presentation. He had no previous hand injury. The patient experienced persistent pain and altered sensation on the ulnar side of the hand, accompanied with progressively diminishing grip strength. Physical examination revealed a small, mobile, and painless palpable mass in the hypothenar area. The skin overlying the mass was normal. The range of motion of the finger joints was not restricted and there was no vascular impairment.

Laboratory exams were within normal limits and radiographs of the hand demonstrated no osseous involvement or radiopaque lesion. Ultrasound (U/S) examination revealed a solid, hypoechoic and homogeneous tumor with capsule in the ulnovolar aspect of the left hand (Figure 1A). Magnetic Resonance Imaging (MRI) indicated a multi-lobulated mass, located in the volar aspect of the fourth web space of the hand, not infiltrating adjacent structures or having bony involvement and bone edema (Figure 1B). No biopsy was performed preoperatively, since all radiological studies suggested a benign tumor.

The patient was operated under axillary nerve block. Using a Brunner incision volarly the skin and the palmar fascia were divided carefully. The neurovascular bundles were identified and protected. The mass was removed en block with clear margins. The gross specimen consisted of a lobulated, well-circumscribed, gelatinous mass (Figure 2A).

The histopathology of the specimen revealed (Figure 2B) uniform, cytologically bland spindle-shaped cells with eosinophilic cytoplasm separated by abundant extracellular myxoid stroma containing very sparse capillary – sized blood vessels.

After the last follow up, 12 months following the excision, the ring and the little finger of the patient had normal sensation and range of motion (Figure 2C). No signs of tumor recurrence were noticed during follow up.

Commentary

Stout[1] in 1948 was the first to present a large series of patients and to establish the diagnostic criteria for myxoma. According to his findings, main characteristics are star-shaped or fusiform cells in a loose matrix with collagen fibers, poor vascularization and absence of mesenchymal elements. It may derive from muscular, nerve, bone or other tissue[1]. Heart is the most common affected area, followed by the jawbones and the lower limbs.

Hand is rarely affected by myxomas. Al Qattan[2] categorized hand myxomas, according to whether they originate from bone, periosteum or soft tissues, into 3 groups. Intramuscular myxoma is considered as the rarest type. It is a benign mesenchymal tumor consisting of bland splinted cells surrounded by an avascular myxoid stroma. The etiology of this entity is unknown, though non-differentiated fibroblasts and mesenchymal pluripotent cells are considered as the origin of the tumor. Those fibroblasts are capable of producing hyaluronic acid rather than collagen. It occurs mainly in adults between their fourth to sixth decade of life, having a female predominance. It is usually a painless, small mass of less than 2 cm diameter, movable at rest and fixed during muscular contraction. Larger myxomas and lesions causing neurologic symptoms have also been reported. Benign intramuscular tumors such as lipoma, gaglion cyst, giant cell tumor of tendon sheath, hemangioma, neurofibroma or malignant tumors like myxoid liposarcoma, rhabdomyosarcoma, clear cell sarcoma, epilthiloid sarcoma, fibrosarcoma, should be differentiated from intramuscular hand myxomas.

Typical radiographs may reveal a soft tissue mass, with or without calcifications. In ultrasound examination, an intramuscular myxoma appears as hypoechoic lesion, with well-defined margins, which is similar to cystic areas, surrounded by normal tissue. On Computer Tomography (CT) scan it appears also as a well defined, ovoid, homogenous, low density

The authors have no conflict of interest.

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Edited by: P. Makras
Accepted 6 September 2015

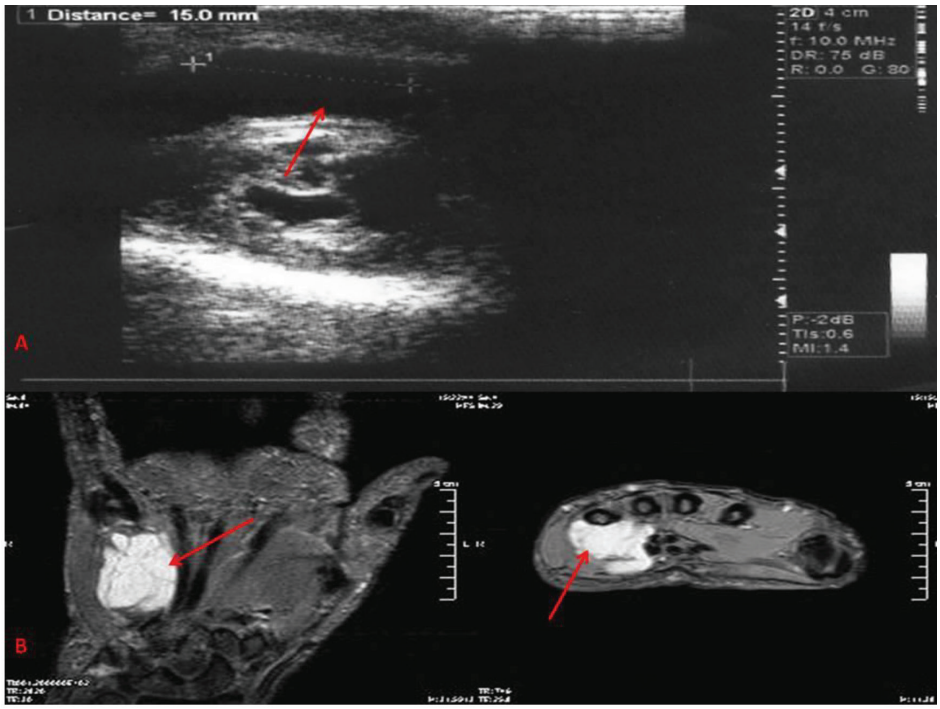

Figure 1. A) Ultra sound (U/S) imaging indicating an hypoechoic, homogeneous mass. **B)** T2-weighted MRI images reveal a multilobulated mass in the hypothenar area measured about 3,5 cm x 3 cm on coronal plane, and 2,5 cm x 2,5 cm on transverse plane.

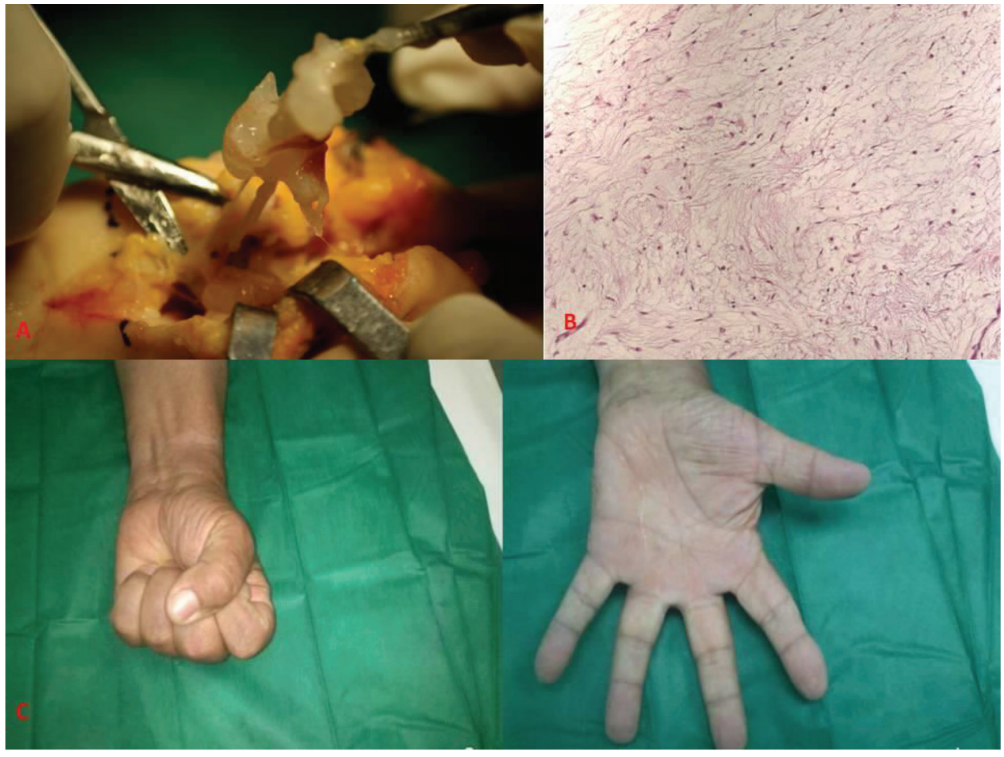

Figure 2. A) Surgical specimen with characteristic myxomatous appearance. **B)** Microscopic appearance of the tumor showing uniform, cytologically bland spindle-shaped cells with eosinophilic cytoplasm separated by abundant extracellular myxoid stroma. (Original magnification x 20). **C)** Normal range of motion of the involved fingers in the left hand at 12 months follow-up. No contractures were detected and the postoperative scar was minimal.

mass not abutting muscular tissue. A pseudocapsule may be present. Examination with MRI[3] can be helpful for the diagnosis by revealing this pseudocapsule, the surrounding perilesional fat and edema. On T1-weighted images intramuscular myxomas appear as homogenous low intensity mass, while on T2-weighted images they appear as homogenous high intensity signal mass. Heterogeneous signal can be depicted, when there is distraction from fibrous septa.

Histological findings put the definite diagnosis of intramuscular myxomas. Macroscopically they are grayish, gelatinous "invading" tumors, extending between the muscle fibers and aponeurotic layers. Microscopically mucoid matrix is poorly vascularized, containing sparse cellular elements, fusiform and stellate cells. Fibroblasts can rarely be found at the periphery, as parts of the pseudocapsule, or diffused, as part of fibrous septa[4]. No signs of mitosis should be observed.

Definite treatment is surgical excision with clear margins. Local recurrence of intramuscular myxomas is rare[5] and is associated with syndromes, such as Mazabraud syndrome, incomplete excision or in cases of intramuscular low-grade myxoid neoplasms. Radiotherapy is not indicated as a treatment option. Conservative treatment is considered in cases of small lesions, or when vital structures are in danger. If a new lesion occurs, further wide surgical resection should be performed.

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Questions

1. Intramuscular myxoma is:
A. A benign tumor of adipose tissue.
B. A malignant mesechymal tumor.
C. A malignant small, round, blue cell tumor.
D. A benign mesechymal tumor.

Critique

Intramuscular myxoma is a rare benign mesechymal tumor consisting of bland splinted cells surrounded by an avascular myxoid stroma. Non-differentiated fibroblasts and mesenchymal pluripotent cells are considered the origin of the tumor. Those fibroblasts are capable of producing hyaluronic acid rather than collagen.
The correct answer is D.

2. Which statement is correct about intramuscular myxoma?
A. It metastasizes to lungs.
B. Radiotherapy is the definite treatment.
C. It may recur locally, depending on clear surgical margins.
D. Mucoid matrix is highly vascularized.

Critique

Intramuscular myxoma is a benign soft tissue tumor. Histopathological findings microscopically show a poorly vascularized mucoid matrix, containing sparse cellular elements, fusiform and stellate cells. Fibroblasts are rare and no signs of mitosis should be observed. Surgical excision with clear margin is the suggested treatment of choice. Recurrence is local and depends on tumor-free surgical margins, while associated with syndromes and low grade myxoid neoplasms. Radiotherapy is not indicated due to side effects.
The correct answer is C.