

Rare forearm intramuscular myxoma: a case report

Hosein Pirmohamadi¹, Mahmood Jafar Begloo¹, Mohsen Rahimi¹, and mohsen Akbaribazm²

¹Baqiyatallah University of Medical Sciences

²Torbat Heydarieh University of Medical Sciences

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1. INTRODUCTION

Intramuscular myxoma (IMM) are benign soft tissue that account for 0.1 to 0.13 per 100,000 populations (1). Various theories have been described regarding the mechanism of IMM occurrence. Some researchers suggest that the reason is fibroblasts (unable to synthesize collagen fibers) that are not well differentiated from mesenchymal stem cells, which cause the synthesis of myxoid stroma without reticular fibers. Others consider the etiology of IMM to be caused by traumatic mechanisms or the growth of polysaccharide-producing cells in the neoplastic process (2). IMM is rare and can occur in the buttocks, thigh, upper extremities and shoulder muscles. Epidemiologically, its occurrence rate is higher in women (70%), increases with age (6th and 7th decade of life) and the most common sites of IMM is upper extremities muscles (50%-60%) (3). According to the location of the masses, soft-tissue myxomas are classified into superficial angiomyxoma, intramuscular myxoma, nerve sheath myxoma and aggressive angiomyxoma. From the clinical point of view, IMM is a palpable mass, painless, without inflammatory secretions and symptoms, which has no contractile properties and no stretch-contraction changes during flexion-extension of the adjacent muscles (4).

From a diagnostic point of view, IMM is observed as a non-calcified mass in plain radiograph, which is seen in the supplementary findings with the help of ultrasonography as echogenic cystic lesions among the muscle tissue. The most important diagnostic method of IMM from other soft tissue lesions is magnetic resonance imaging (MRI), which can be seen as hypointense homogeneous mass in T1-weighted sections and hyperintense in T2-weighted sections (5). In case of edema with IMM in MRI sections (T1-weighted sections), it should be differentiated from other fluid-containing lesions (such as cystic teratoma, hematoma, myxoid sarcoma, cystic hygroma and even normal lymph nodes). Also, IMM should be differentiated from proliferative lesions, other myxoid neoplasms, myxochondroma, myxochondroma and myxoid liposarcoma (6). Cytology-histopathology findings with the help of intraoperative frozen section and needle biopsy help the information of MRI sections in the diagnosis of IMM. Density and ratio of cells/ collagen fibers, mucoid material secretion, nodular-vesicular pattern and fat density in histopathological sections contribute to IMM (7). In the present case, forearm intramuscular myxoma was observed inter-supinator muscle.

2. CASE REPORT

A 60-year-old female presented to Najmiyeh hospital (a hospital in Tehran), with edema, progressive pain in the anterior-proximal side of the right forearm, palpation of well-defined masses with a high growing rate and movement limitation during forearm flexion. During physical examination, it was found that the mobile tumor mass with specific boundaries was inside the supinator muscle (anterior aspect of elbow joint) and did not have any inflammatory or secretion symptoms. Evaluation of the lesion with sagittal and coronal MRI T2-weighted sections showed that the $3.5 \times 2.5 \times 2$ cm well-defined high signal mass was observed in the antero-proximal elbow joint and inside the supinator muscle (Figure1 and 2).

Differential diagnosis

Differential diagnosis of forearm mass in MRI evaluation with high water content (mimicking a cyst) include synovial cyst, ganglion, bursa, neurogenic tumor, myxoid liposarcoma, and malignant fibrous histiocytoma. The majority of synovial cysts, bursae, and ganglia occur at typical locations such as the popliteal fossa and dorsal aspect of wrist. These lesions typically are seen in intermuscular planes, whereas most soft tissue myxomas are intramuscular in location. Also, as compared to intramuscular myxoma, these lesions show a cystic nature with peripheral rim enhancement and delicate septae only seen on postcontrast computed tomography and MRI images. Ultrasonography, these lesions show an anechoic component as compared to IMM, which are hypoechoic in echotexture, with posterior acoustic enhancement and no anechoic component. Neurogenic tumors are also typically located in the intermuscular compartment. A nerve entering and exiting the lesion is usually appreciated at the margins of the lesion. These imaging findings are not seen in IMMs. Similarly, myxoid liposarcomas are usually intermuscular lesions developing in the subcutaneous fatty tissue, unlike soft tissue myxomas. In addition, myxoid liposarcomas usually contain small amounts of intrinsic fat. In this respect, the IMM may have some imaging features similar to myxoid liposarcoma. Myxoid malignant fibrous histiocytomae are intramuscular lesions similar to myxomas, however these lesions show far more heterogenous appearance on imaging, with areas of hemorrhage. These lesions also show some solid nodular component with prominent contrast enhancement, whereas the myxomas show moderate, relatively homogenous enhancement.

During surgery, a longitudinal incision was made in the anterior area of the elbow joint on the supinator muscle at the site of the tumor lesion and the encapsulated mucoid-gelatinous gray myxoid mass $3.5 \times 2.5 \times 2$ cm was removed from the muscle.

Histopathological evaluation

Histopathological evaluation showed that the structural tumor mass is capsule, hypocellular, without infiltration, containing scattered fibroblasts (spindle or stellate cells) in the mucoid background. Further evaluation showed the presence of scattered thin collagen fibers, small cells with hyperchromatic nuclei with scanty cytoplasm. The lesion did not have cystic degeneration, mitosis and necrosis (Figure 3).

Post-operation and follow-up

There were no complications post-operatively, the surgical site was no signs of hematoma, infection, pain and inflammatory symptoms, and in the movement test after 2 weeks, the patient easily performed flexion-extension of the elbow joint (Figure 4).

3. DISCUSSION

IMM is a rare benign soft tissue tumor that probably occurs due to incomplete differentiation of mesenchymal cells into fibroblasts. Of course, IMM can be a part of McCune-Albright syndrome and Mazabraud's syndrome, which in this case is caused by mutations in the GNAS gene (8). For the first time, the term myxoma was applied by Virchow to a soft tissue tumor of mesenchymal cell origin in 1863. Also, Stout in 1948 described the histopathological criteria for the diagnosis of myxoma and its differentiation from other lesions (9). Capsular mass with hypocellular tissue in a mucoid background with scattered reticular fibers in the skeletal muscles, which, if not diagnosed/treated in time, can cause movement limitation in the joint, interfere with the action of the involved muscle, and compress the nerves in the area. IMM has a good prognosis and usually the involved muscle/joint follows its anatomical function following supportive measures-physiotherapy (3).

This lesion is seen in T1-weighted images as an ovoid-shaped mass with low signal intensity and in T2-weighted images as high signal intensity. In the present case, a hyperintense egg-shaped mass was observed in T2-weighted sections inside the supinator muscle (10). Adamonis et al. (2019) also reported the presence of IMM in the deltoid muscle in a 34-year-old man, whose tumor lesion was observed in T2-weighted images with hyperintense appearance. In histopathological evaluation, after observing spindle-stellate fibroblast cells in mucoid background with scattered collagen fibers of deltoid IMM was confirmed (5). In the present case, the mass has a thin capsule around the homogenous hypocellular mucoid tissue, in which the spindle-shaped

cells (fibroblasts) are located in scattered collagen fibers. The reports of the upper extremity IMM cases show that the lesions in the muscles are removed subcutaneously after surgery and cause few complications (11). Movement limitations caused by IMM are also quickly resolved with physiotherapy if the nerves of the brachial plexus are not compressed.

4. CONCLUSION

IMM is a rare benign soft tissue tumor that is caused by incomplete differentiation of fibroblasts from mesenchymal cells in skeletal muscles. MRI allows the diagnosis of IMM and its differentiation from other skeletal muscle lesions, which is confirmed by the histopathological evaluation of the lesion after biopsy. This benign lesion has a fast growth and has a good prognosis after surgery and removal.

AUTHOR CONTRIBUTIONS

Mohsen Akbaribazm : Conceptualization; writing – original draft; writing – review and editing. **Hosein Pirmohamadi** : Conceptualization, supervision, writing – review and editing. **Mohsen Rahimi** and **Mahmood Jafar Begloo** : Supervision, writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that there is no conflict of interest.

CONSENT

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DATA AVAILABILITY STATEMENT

All data associated with the article is available if required.

KEY CLINICAL MESSAGE

Intramuscular myxoma (IMM) is a benign soft tissue tumor of mesenchymal origin that occurs mainly in skeletal muscles which can be removed through surgery after diagnosis.

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Figure legends:

Figure 1: (A) Sagittal and (B) Axial MRI section showing high signal intensity on T2-weighted images in soft tissue lesions. The red arrow indicates IMM inside the supinator muscle.

Figure 2: Coronal MRI section showing high signal intensity on T2-weighted images in soft tissue lesions. The red arrow indicates intramuscular myxoma inside the supinator muscle.

Figure 3: Histopathology of IMM masses. IMM with thin capsule, fibroblasts (spindle or stellate cells) and hypocellular mucoid background containing thin scattered collagen fibers. C: Capsule, F: Fibroblasts and Co: Collagen fibers [(A1 and A2) H&E staining x 100, Scale bar = 200 μ m and (B1 and B2) H&E staining x 400, Scale bar = 50 μ m].

Figure 4: The surgical site of the right forearm of a 60-year-old female patient with IMM after healing of the incision site and follow-up physical examinations of supinator muscle and elbow joint functions.







