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CME ARTICLE

Soft Tissue Mass in the Proximal Forearm of a 17-Year-Old Woman

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HISTORY AND PHYSICAL EXAMINATION

A 17-year-old girl developed a palpable, mildly painful mass, which had been slowly enlarging for a period of 6 months before the initial examination, in the volar aspect of the proximal ⅓ of her dominant forearm. The clinical presentation of the lesion coincided with a period of overuse of the extremity by the patient, who was a student taking final exams. The patient could not recall a history of trauma to the extremity and had no constitutional symptoms. Her medical history was unremarkable.

On physical examination, a firm, well-circumscribed, nut-sized lesion was palpated in the musculature of the volar surface of the proximal forearm. The lesion was not tender to palpation, the overlying skin had normal appearance, and there was no regional lymphadenopathy. The range of motion (ROM) of the elbow and the forearm were not restricted and there was no neurovascular impairment, confirmed by electromyographic (EMG) and Doppler studies. Laboratory findings were within normal limits. Radiographs and magnetic resonance images were taken. Based on the history, physical examination, and imaging studies, what is the differential diagnosis?

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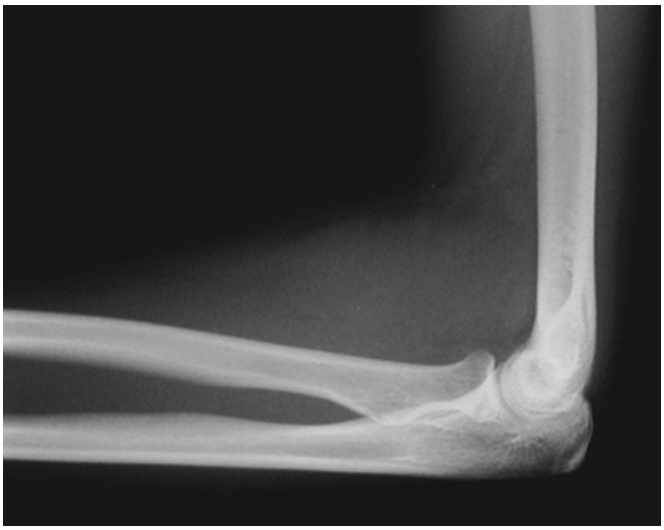

Fig 1. A lateral plain radiograph of the patient's affected elbow shows a soft tissue mass in the volar surface of the proximal ⅓ of the forearm.

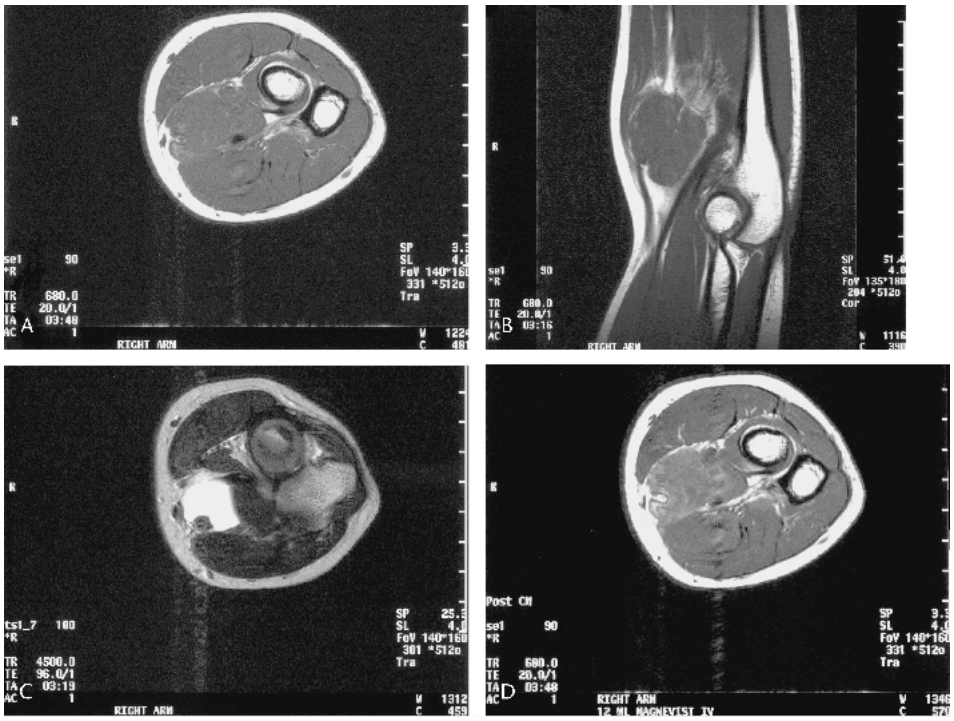

Fig 2A–D. Magnetic resonance images of the proximal forearm of the patient in T1-weighted sequences in the (A) axial and (B) sagittal planes show that the mass is isointense to muscle signals. (C) In the T2-weighted axial sequence, the signal homogeneously increased. (D) A postcontrast T1-weighted sequence shows faint inhomogeneous enhancement. The lesion seemed to invade the muscle mass focally.

IMAGING INTERPRETATION

The plain lateral radiograph of the elbow showed a soft tissue mass in the volar proximal forearm with no obvious F1 osseous involvement (Fig 1). Multiplanar magnetic resonance imaging (MRI) of the forearm showed a well-circumscribed mass, measuring approximately 5 × 4 cm in F2 the proximal forearm (Fig 2). The mass was situated volar to the distal brachialis muscle and the supinator and between the pronator and brachioradialis muscles. It could not be distinguished from the distal brachialis muscle, and it abutted the radial artery. The mass was isointense to muscle signal on the T1-weighted MRI sequences and showed a homogeneously increased intensity signal on the T2-weighted MRI sequences. Postcontrast images revealed faint, inhomogeneous enhancement.

DIFFERENTIAL DIAGNOSIS

Schwannoma
Rhabdomyosarcoma
Synovial sarcoma
Soft tissue myxoma
Myxoid liposarcoma
Myxoid nodular fasciitis

Fine-needle aspiration of the lesion was inconclusive. Therefore, the mass was biopsied through a volar approach. A lobulated, well-circumscribed, ovoid tumor measuring 5.5 × 4.5 × 3.5 cm was found overlying the supinator and flexor digitorum superficialis muscles between the brachioradialis and pronator teres. An intraoperative frozen section was obtained and was consistent with a myxomatous, benign lesion. An excision biopsy was done. Based on the history, physical findings, radiographic studies, and histologic picture, what is the diag- F3 nosis and how should this patient be treated?

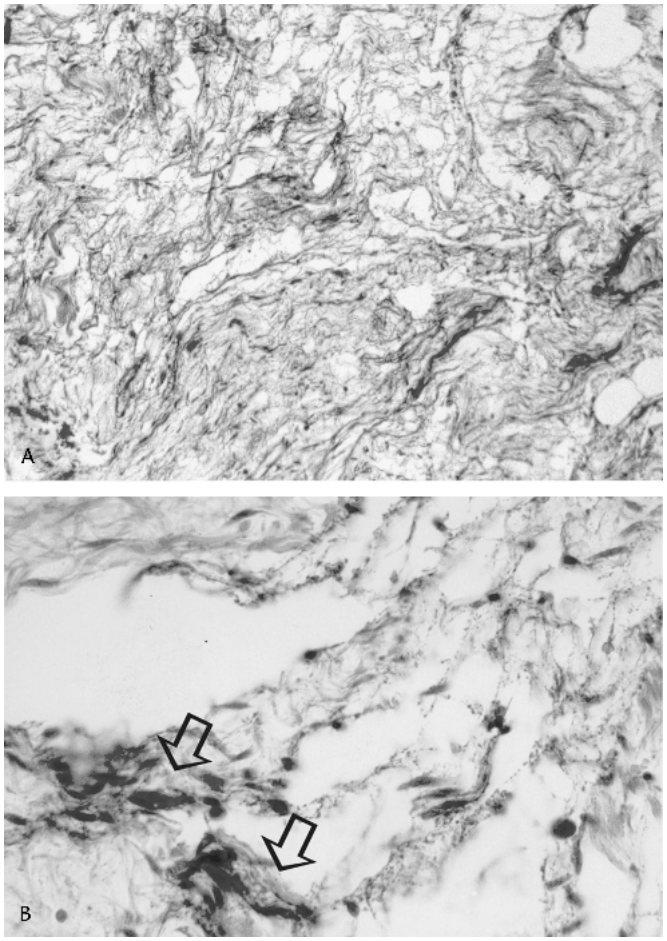

Fig 3A–B. (A) Photomicrographs of the resection specimen show the tumor is composed of abundant extracellular myxoid material. Characteristic features shown are the paucity of stromal cells and vascular structures (Stain, hematoxylin and eosin; original magnification, ×230). (B) At the periphery of the tumor, residual degenerating voluntary muscle fibers surrounded by myxoma tissue are clearly shown (arrows); a cystic space with dropped-out mucin also clearly is shown (Stain, hematoxylin and eosin; original magnification, ×430).

See page XXX for diagnosis and treatment.

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HISTOLOGY INTERPRETATION

Gross examination of the specimen showed a well-circumscribed tumor covered partly by bundles of fascial tissue and skeletal muscle. The cut surface of the tumor was gray-white and vaguely lobulated, with a gelatinous and myxoid consistency. Foci of small cystlike spaces filled with mucoid material were unevenly distributed in the mass.

On thorough histologic examination, the tumor was paucicellular, composed of sparse uniform cells with small oval, spindle-shaped, and/or stellate, darkly stained nuclei, and separated by abundant extracellular myxoid material (Fig 3A). The stromal cells were vimentin positive but negative for desmin, S100 protein, and endothelial markers; the myxoid interstitial material stained positively with alcian blue at pH 2.6. The vasculature consisted of few thin-walled vessels. Fine fibrillar reticulin resembled fibers of frayed string (Fig 3). Sections taken from the interface of the tumor and focally surrounding muscle (corresponding to the MRI findings of invasion) showed minimal infiltration of the adjacent skeletal muscle (Fig 3B). This infiltration consisted of short, fine strands of myxoid AU5 tissue extending between shrunken individual, degenerating skeletal muscle fibers.

DIAGNOSIS

Intramuscular myxoma

DISCUSSION AND TREATMENT

The term myxoma was first used by Virchow[25] in 1863 to describe tumors with structures similar to that of the umbilical cord without any other kind of differentiation. Stout[22] defined myxoma as a true neoplasm composed of stellate cells in a loose myxoid stroma through which course delicate reticulin fibers. Stout excluded myxoid tumors with recognizable differentiation, such as liposarcomas, fibrosarcomas, and chondrosarcomas, and ganglion cysts.

Intramuscular myxoma was first delineated as a definite clinicopathologic lesion by Franz Enzinger in 1965.[7] It is a rare, benign soft tissue tumor, composed of fibroblasts and abundant myxoid stroma,[2,7,18,22] with an incidence of approximately one case per 1 million people per year.[2] Two thirds of the patients are women, and it usually occurs between the ages of 40 and 70 years.[2,18,22]

The tumor usually is located in the thigh and, in descending order of frequency, in the buttock, shoulder, lower leg, and arm.[2,7,18,22] Its exact location in the musculature varies; some tumors are completely surrounded by muscle tissue, whereas others are attached firmly on one side to muscle fascia. Intramuscular myxoma occurs as a solitary entity, less frequently in association with fibrous dysplasia of the bone (Mazabraud's syndrome), or as a part of the McCune-Albright syndrome (polyostotic fibrous dysplasia, café-au-lait spots and endocrine hyperfunction). In these syndromes, multiple myxomas are encountered more often.[3,9,12,16,19,23]

The only report on soft tissue myxomas in childhood is that by Dutz and Stout in 1961.[6] In that report, the contemporary division of soft tissue myxomas (mainly into intramuscular myxomas, juxta-articular myxomas, superficial angiomyxomas, aggressive angiomyxomas, and myxomas of the nerve sheath)[2] was not used. From that report only, two cases could be considered intramuscular myxomas. In one case (Case 3), a 5-month-old girl was presented with multiple recurrent lesions in the extensor carpi ulnaris muscle. The multiplicity of the lesions in the absence of concomitant fibrous dysplasia of the bone and their recurrent nature are not typical characteristics of intramuscular myxoma. In the other case (Case 27), a 15-year-old girl presented with a mass that involved the tendinous insertion of the pectoralis major muscle to the humerus. The intratendinous growth and the site of the lesion are characteristic of juxta-articular myxoma, rather than intramuscular myxoma.

Typical MR imaging characteristics of intramuscular myxomas include low T1-weighted and increased T2-weighted signal characteristics of the mass.[1,15] Variable degrees of enhancement, most commonly in an inhomogeneous pattern, is usually seen after administration of gadolinium contrast.[14,17] In recent, retrospective reviews, the authors proposed the presence of two signs, the peritumoral fat rind and T2-weighted hyperintensity of adjacent muscles, as strongly suggestive of intramuscular myxoma, as opposed to myxoid liposarcoma.[4,17] The lesion may have poorly defined margins that blend with the surrounding soft tissues[4,17]; this is referred to as infiltrating growth by some authors.[2,4] This infiltrating growth effect of intermuscular myxomas on MRIs could be attributed to the gradual transition from the tumor tissue to muscle[11] and could correspond to areas of "residual degenerating voluntary muscle fibers surrounded by myxoma tissue"[2] often seen histologically at the periphery of the tumor. The typical MRI signal characteristics, along with heterogeneous enhancement, were present in our patient. The mass was partially surrounded by fat rind, but the latter could be normal, rather than related or reactive to the tumor fatty tissue. Although, for the most part, the mass was well defined, there was evidence of blending with surrounding tissues. As with all soft tissue masses, the imaging findings are suggestive rather than specific for

intramuscular myxoma because almost identical findings can be encountered in many other tumors.[14]

Some of the tumors discussed in the differential diagnosis show characteristic features on MRI. Schwannomas often present a central target pattern expressed as low signal area in the center of the tumor with increased intensity signal in the periphery on T2-weighted sequences.[24] Rhabdomyosarcomas (alveolar type) are common in the extremities and trunk in this age group (10–25 years) but usually the signal is intermediate-to-high on T2-weighted sequences with lobular enhancement.[21] Synovial sarcomas most commonly are seen in lower extremities, and MRI usually shows intermediate T1-weighted and high T2-weighted signal with heterogeneous internal architecture and ill-defined margins.[5] In general, sarcomas are indistinguishable from each other by MRI; location and age are used to include or exclude from the differential diagnosis. In nodular fasciitis, the signal is usually decreased on T1-weighted and decreased or mixed on T2-weighted sequences, but can vary according to the exact histologic type.[20,26]

In our patient, the histologic differential diagnosis in this age group included a reactive myofibroblastic lesion, such as myxoid nodular fasciitis, and the group of myxoid hypocellular soft tissue tumors that should be distinguished from each other, such as juxta-articular myxoma, low-grade fibromyxoid sarcoma, and myxoid liposarcoma. Myxoid nodular fasciitis was excluded from the differential diagnosis because of the absence of a rapidly enlarging and painful or tender swelling, a history of recent onset, and MRI findings. Histologically, myxoid nodular fasciitis is composed of plump fibroblasts and myofibroblasts arranged in short fascicles and untidy bundles. The myofibroblasts possess uniform, plump, but tapering, vesicular nuclei with occasional prominent nucleoli and frequent mitoses. The stroma contains delicate thin-walled capillaries, a mild predominantly lymphocytic infiltrate, and extravasation of erythrocytes. Juxta-articular myxoma was excluded by considering the anatomical location of the tumor. Also, histologically, juxta-articular myxomas tend to be less homogeneous and more fibroblastic and cystic than intramuscular myxomas, and mitotically active, atypical reactive cells may be apparent.[2] Additionally, a low-grade fibromyxoid sarcoma is an uncommon neoplasm that occurs in the deep soft tissues of adults in their 40s, 50s, and 60s.[18] The tumor characteristically has alternating fibrous and myxoid areas with bland spindle-shaped tumor cells arranged in swirling or whorled formations around thin-walled capillaries.[8,10] These typical features were not noted in our patient. Myxoid liposarcomas, which affect younger adults, are commonly deep-seated tumors with the vast majority occurring in the thigh, although they infrequently may be located subcutaneously or even in the skin.[10] Histologically, they are more cellular than intramuscular myxoma, the cells are plumper, and some may differentiate as signet-ring lipoblasts. In addition, the plexiform capillary pattern and mucinous pools or lakes of myxoid liposarcoma allow for identification at low power.[10]

Intramuscular myxoma is a benign tumor and local surgical excision with tumor-free margins usually suffices.[2,7,11,19] Awareness of this entity is of particular importance because this lesion, which has no tendency toward recurrence and is treated by local excision, easily is mistaken for a sarcoma, especially myxoid liposarcoma and botryoides rhabdomyosarcoma.[7] Nevertheless, there have been a few recurrences after excision of intramuscular myxomas described in the literature,[9,12,13,16] but even in these patients the pathology was doubtful or the possibility of confusing multiple myxomas with recurrences could not be ruled out.

Our patient was treated with local surgical excision with tumor-free margins. Radiographic images of the axial and appendicular skeleton were negative for fibrous dysplasia, and endocrine studies were in normal limits. The 29-month followup period has been uneventful, with no complications. The patient is currently recurrence free, with normal wrist and elbow function.

Intramuscular myxoma is a benign, nonrecurrent tumor. Its presentation in adolescence, although extremely rare, cannot be ruled out. Although MRI offers a helpful description, a differential diagnosis of intramuscular myxoma from other aggressive tumors can only be made histologically.

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