Pathology Feature

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Section Editor

Massive intramuscular myxoma associated with fibrous dysplasia of bone[1]

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A massive intramuscular myxoma associated with fibrous dysplasia of the bone in a 70-year-old man was successfully resected. Only 15 such cases of this rare combination of lesions have been reported. Careful differential diagnosis is necessary because of the similarity of this lesion to sarcoma.

Index terms: Bones, fibrous dysplasia • Muscles, neoplasms • Myxoma

Cleve Clin Q 50:469–472, Winter 1983

Myxomas of somatic soft tissues are uncommon benign mesenchymal tumors composed of sparse stellate mesenchymal cells embedded in abundant hyaluronic acid-rich ground substance. They often have an intramuscular location and occasionally attain large size. When found in the muscles of the thigh, they are easily mistaken clinically for aggressive sarcomas, especially liposarcomas. If an associated radiographically visible lesion in an underlying or nearby bone is present, the suspicion of a malignant neoplasm is heightened.

The first description of an association between soft-tissue myxomas and fibrous dysplasia of bone has been attributed to Henschen's report in 1926.[1] Currently, only 15 such cases have been reported in the American and European literature.[2–5] Recently, we encountered another patient with this rare combination of lesions. His massive 29-cm tumor is the largest myxoma recorded. This case report is presented to underscore the importance of recognizing this rare syndrome when evaluating patients with tumors of the extremities.

Case report

Clinical history: A 70-year-old white man was referred to the Cleveland Clinic in January 1983, with a 15–20 year history of a left anterior thigh mass. The lesion had been asymptomatic until the previous two years when it began to expand and extend proximally. This change was associated with a feeling of abdominal fullness and a dull, aching pain over the left thigh. In 1946 a "soft spot" in the left femur had been removed, and a "lipoma" was excised from the left anterior groin in 1976.

Physical examination revealed a large, soft-tissue mass in the left anterior thigh, which extended proximally to the groin and into the pelvis. The lesion was tense and tender but did not restrict the range of motion of the left hip or knee. A ^{99m}Tc bone scan *(Fig. 1)* demonstrated increased uptake in the left femur, from the proximal epiphysis to the distal diaphysis, and in the proximal epiphysis of the left tibia. In addition, there was diffuse increased uptake in the soft tissues surrounding the left femur. These findings were felt to be consistent with either a primary bone tumor or a soft-tissue neoplasm with secondary bony involvement. A computed tomography (CT) scan of the femurs *(Fig. 2)*, pelvis, and abdomen revealed a 29-cm lobulated soft-tissue mass of low attenuation, which extended from the left knee

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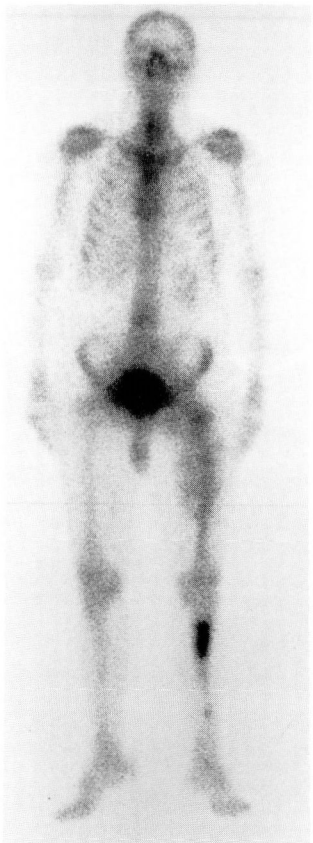

Fig. 1. ^{99m}Tc scan demonstrates increased uptake in the left femur from the proximal metaphysis to the distal diaphysis and in the proximal left tibial metaphysis. Diffuse soft-tissue uptake is seen in the proximal left thigh.

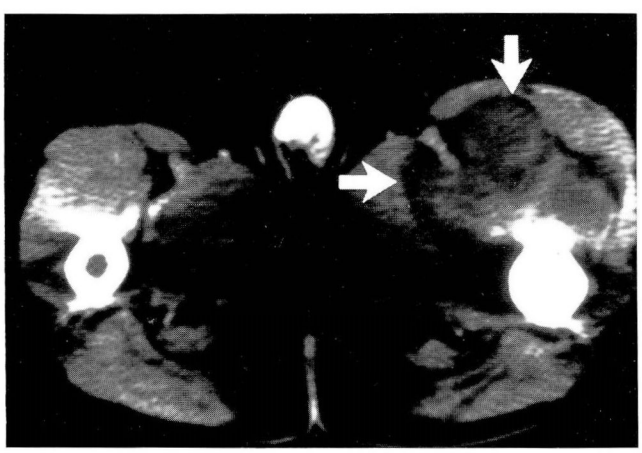

Fig. 2. CT scan at the level of the proximal shaft of the femur demonstrates a lobulated soft-tissue mass in the anterior left thigh (arrows). The diffuse density within the medullary cavity of the left femur probably represents fibrous dysplasia.

into the pelvis. It appeared to follow muscle planes, was of variable density, and was believed to be most consistent with a liposarcoma. There were no lesions in the abdomen or chest.

On 13 January 1983, an open biopsy specimen of the mass was diagnosed histologically as a myxoma. Because of the accompanying osseous changes and the rarity of massive soft-tissue myxomas, it was decided to obtain additional tissues for histologic examination. On 19 January 1983, a second biopsy of the left thigh mass and a biopsy of the left tibial lesion were performed; specimens were interpreted as intramuscular myxoma and fibrous dysplasia of bone, respectively. On 25 January 1983, the patient underwent a marginal excision of the mass. At operation, the soft-tissue tumor was found to be nonencapsulated and multilobulated, with extensive involvement of the vastus intermedius and iliacus muscles. It extended along the femoral nerve from the pelvis into Hunter's canal beneath the sartorius muscle. After dissection from the femoral nerve and the motor nerves to the rectus femoris, vastus lateralis, and vastus medialis muscles, the neoplasm was resected. The vastus intermedius muscle and its corresponding nerve were completely excised because of extensive involvement by tumor.

The patient was seen on 28 April 1983, three months after the resection. He was walking without a limp and had excellent strength in the quadriceps muscles. He was able to flex his knee and hip 120 degrees, with full internal and external rotation and no flexure contracture. The patient was without pain, and there were no signs of local recurrence of the myxoma.

Pathological findings: The 1.24-kg resected specimen consisted of multiple nonencapsulated tumor nodules containing blue-white translucent gelatinous material demarcated by fibrous septa *(Fig. 3)*. A small amount of skeletal muscle and aponeurosis were attached. Histologically, the tumor was a typical intramuscular myxoma and was identical to that of the previous biopsy specimens. Most of the neoplasm was composed of pancicellular myxoid ground substance rich in hyaluronic acid. Embedded within the myxoid material were small numbers of stellate and spindle-shaped mesenchymal cells with small bland nuclei *(Fig. 4)*. No nucleoli or mitotic figures were present. A few cells contained small intracytoplasmic vacuoles, but no lipoblasts were seen. Some areas of the tumor were characterized by increased amounts of collagen. There were no areas of hemorrhage, necrosis, or plexiform capillary network. The neoplasm interdigitated with residual atrophic muscle fibers *(Fig. 5)*.

The biopsy specimen of the left tibial lesion performed on 19 January 1983, revealed the characteristic histopathologic features of fibrous dysplasia of bone. It consisted of irregular small islands of woven bone with a few osteoblasts surrounded by abundant fibrous connective tissue *(Fig. 6)*.

Discussion

Soft-tissue myxomas associated with fibrous dysplasia of bone are rare. This is only the 16th patient to be described with this combination of lesions.[2–5] The myxoma in the current case was principally intramuscular, involving the vastus intermedius and iliacus muscles. Intramuscular myxomas are a distinct subset of soft-tissue myxomas, accounting for approximately 17% of all

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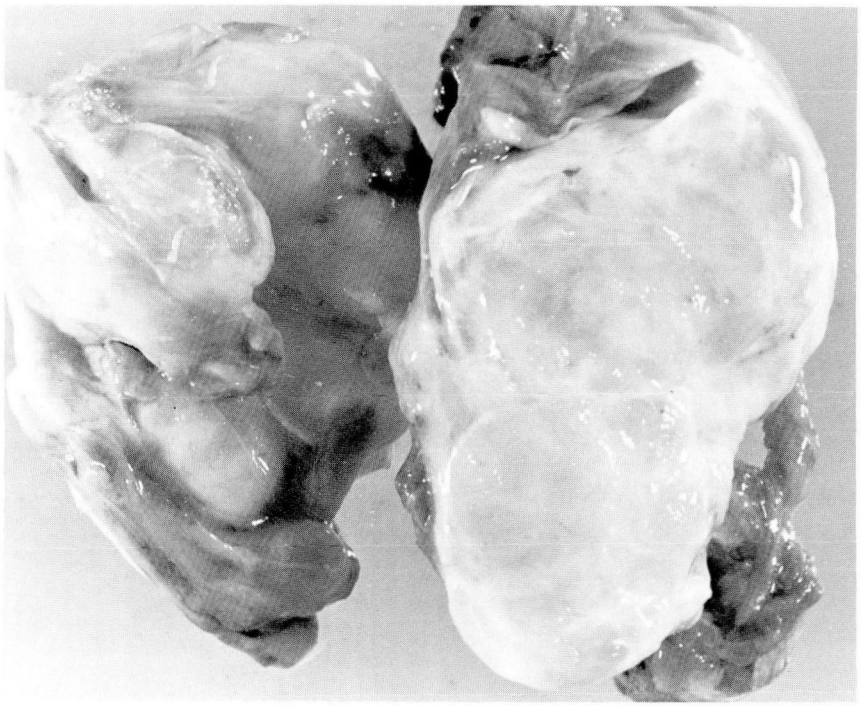

Fig. 3. Portion of the excised large myxoma. Note lobulated, glistening, translucent surface.

such tumors[6]; the other-soft tissue myxomas are located in fascial planes, subcutaneous tissues, neurovascular sheaths, and periosteum.[2] Of the 18 cases of intramuscular myxoma reported by Kindblom et al,[7] all but one were completely enclosed in muscle. The bony lesions of fibrous dysplasia have been associated solely with the intramuscular variant of soft-tissue myxoma.[2,3,5]

Although most intramuscular myxomas are solitary tumors, almost all of the myxomas in patients with fibrous dysplasia have been multiple.[5] Usually multiple myxomas are situated in the same general topographic region, but they have also occurred in different anatomic locations, such as the arm and thigh.[5] The huge multinodular tumor in our patient may actually have been several smaller myxomas coalesced after years of slow growth. His previously removed "lipoma" may also have been a myxoma.

About two-thirds of reported patients with fibrous dysplasia and myxoma were aware of the bone lesions years before the myxomas were noticed.[3,5] Although fibrous dysplasia commonly is monostotic, the polyostotic form has occurred in 86% of patients who had an associated myxoma. Furthermore, approximately 47% of patients with both fibrous dysplasia and myxomas have also had Albright's syndrome of polyostotic fibrous dysplasia, pigmented cutaneous patches, and endocrine abnormalities including precocious puberty.[3,5] Our patient had polyostotic disease with involvement of the left femur and tibia, but did not have the other features of Albright's syndrome.

The clinical diagnosis of myxoma may be especially difficult when the tumor is large, especially when accompanied by radiographically visible changes of an underlying or nearby bone lesion. Often a sarcoma of soft tissue or bone is suspected, as occurred in this case. Soft-tissue myxomas have ranged in size from 1 to 18 cm, with most about 4–6 cm in greatest dimension.[2,5–7] Our patient's 29-cm myxoma is the largest such tumor to be reported to our knowledge. The lesion most likely to be mistaken for a myxoma is a myxoid liposarcoma since both tumors occur in adults, are often located in the thigh muscles where they present as a painless mass, and histologically contain mesenchymal cells surrounded by abundant hyaluronic acid-rich myxoid ground substance. Misdiagnosis may lead to unnecessary amputation.[6]

The prognosis with intramuscular myxomas is excellent, since they are benign tumors that do not metastasize.[2,6,7] Among the 15 reported cases associated with fibrous dysplasia, there was only

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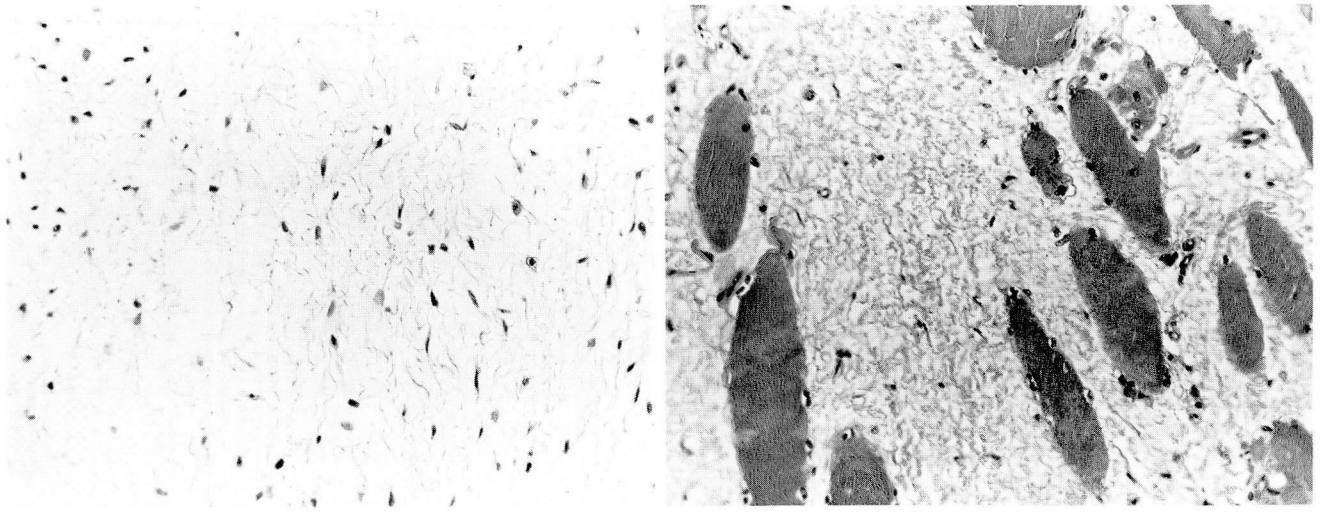

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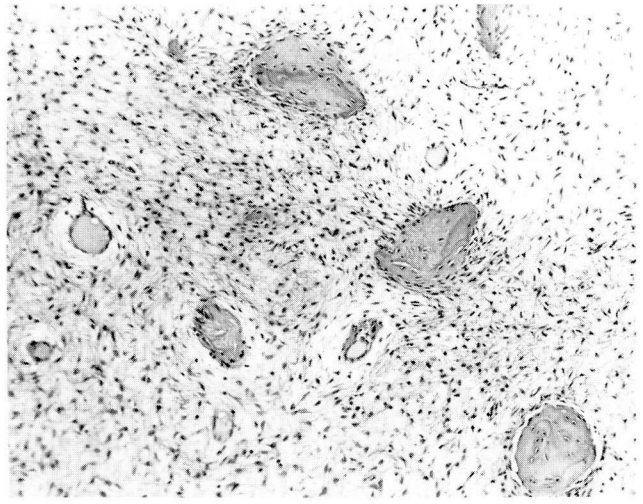

Fig. 4. Microscopic appearance of the myxoma. Scattered, stellate-shaped mesenchymal cells are visible in abundant myxoid ground substance. Few capillaries are seen, and there are no mitotic figures (hematoxylin-eosin stain, × 200).

Fig. 5. Microscopic appearance of the myxoma at interface with skeletal muscle. Individual muscle fibers are surrounded by the myxoid neoplasm (hematoxylin-eosin stain × 200).

Fig. 6. Microscopic appearance of fibrous dysplasia of bone. Islands of woven bone are interspersed throughout abundant fibrous tissue (hematoxylin-eosin stain × 200).

one recurrence of the myxoma.[3] However, an osteosarcoma arising in an area of fibrous dysplasia in a patient with Albright's syndrome and multiple myxomas has been reported.[4] Awareness of the association between benign soft-tissue myxomas, usually intramuscular and multiple, and fibrous dysplasia of bone is valuable in the differential diagnosis of tumors of the extremities.

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