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A case of juxta-articular myxoma of the knee

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Clinical information

The patient is a 9-year-old girl who complained of progressive pain in the right knee over the course of several weeks. The pain was located posteriorly and worsened with extreme knee flexion. Plain radiographs were normal. She had no history of trauma and denied locking or popping of her knee. After completing a 1-month course of physical therapy, the patient underwent magnetic resonance imaging (MRI) of the affected knee (Fig. 1). This study showed a well-defined mass measuring 2.5×2.0×2.0 cm in the posterolateral knee joint. The mass directly abutted and displaced the posterior horn of the lateral meniscus, but the meniscus was otherwise normal. The lesion was isointense to muscle on T1-weighted images and of inhomogeneous intermediate signal intensity on T2-weighted images.

Despite the fact that the patient's pain markedly decreased over the ensuing 5 months, further consultation was sought for evaluation of the mass. At this time, examination of the right knee revealed a posterolateral palpable mass and mildly reduced flexion. The physical examination was otherwise unremarkable. There was no associated edema, erythema, or joint effusion, and there

were no signs of neurovascular compromise distal to the lesion. There was no significant family history or past medical or surgical history. The patient underwent open arthrotomy and excision of the mass.

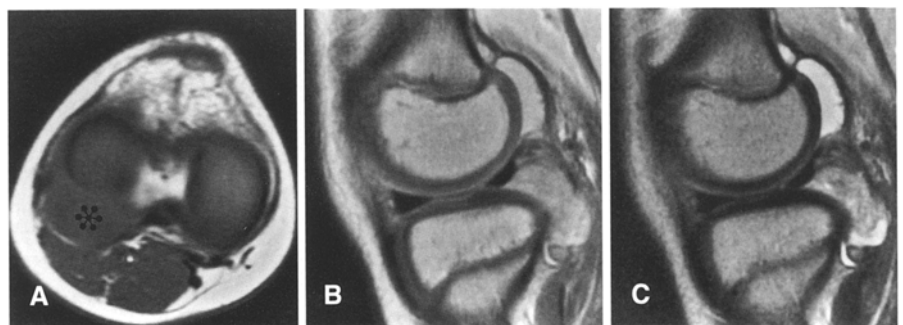
The mass appeared to be intra-articular on the MR images, and the signal characteristics suggested that it contained a high content of either hemosiderin or fibrous tissue. The lack of mineralization on plain radiographs excluded a heavily calcified mass. The differential diagnosis therefore included pigmented villonodular synovitis (PVNS) and a benign tumor or low grade malignancy containing fibrous tissue. Extra-articular masses which may be in close association with a joint were also considered in the differential diagnosis, including complex meniscal, parameniscal, or ganglion cyst. The MR appearance was not typical for synovial hemangioma, synovial lipoma, or the rare intra-articular synovial cell sarcoma.

At surgery, an intra-articular mass measuring 3.0×2.0×2.0 cm was

found attached to the posterior fibers of the lateral meniscus by a small pedicle. The mass was excised at its origin without damage to the meniscus. No meniscal pathology was noted at the time of surgery.

On gross inspection, the mass was firm and nodular with a tan-white color. Histologic examination revealed abundant myxoid stroma with scattered bipolar spindle cells (Fig. 2). Multiple areas of secondary fibrosis, hyalinization, focal necrosis, vascular proliferation, and chronic inflammation were seen throughout the specimen.

Fig. 1. **A** Axial T1-weighted MR image. The mass (*) is isointense to muscle. **B** Sagittal spin density MR image (SE 3400/19). The mass increases in signal intensity. The posterior horn of the lateral meniscus is lifted by the mass, but the meniscus appears normal. **C** Sagittal T2-weighted MR image (SE 3500/95). The mass appears more inhomogeneous, and remains intermediate signal intensity. High-signal-intensity fluid can be seen both above and below the mass



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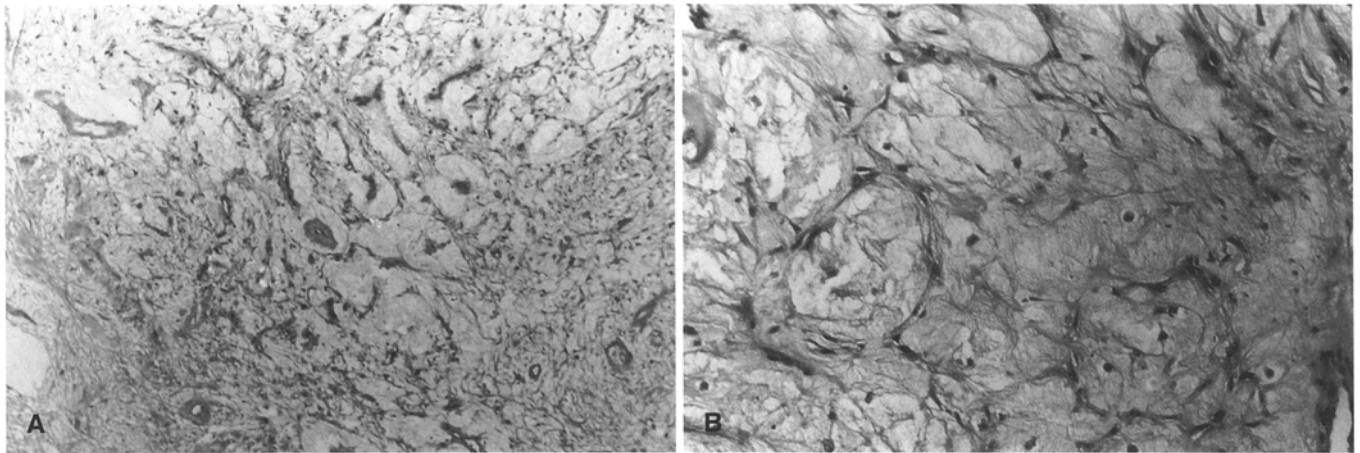


Fig. 2. Large areas of the lesion show abundant myxoid stroma with scattered bipolar cells. **A** H&E $\times 40$; **B** H&E $\times 100$

Juxta-articular myxoma was diagnosed.

Discussion

Juxta-articular myxoma (JAM) is relatively rare benign lesion that occurs most frequently in men in the third to fifth decades of life. Prior to this case, the youngest patient reported was 16 years of age [1]. These masses occur in the vicinity of large joints, most commonly the knee. While most occur in the subcutaneous adipose tissue, tendinous, meniscal, bursal, and capsular origins have been reported [1]. The masses typically range from 2.0 to 7.0 cm in size and may enlarge very rapidly. Pain and a palpable growing mass are often the chief complaints. A recurrence rate of 34% has been reported, and in several cases multiple recurrences have been reported [1]. Treatment consists of excision for both primary and recurrent disease.

Grossly, JAM may appear mucoid, myxoid, gelatinous, slimy, cystic, or multicystic [1]. Histologically, it contains a loose arrangement of primitive fibroblasts in a hypovascular myxoid matrix. Regions of hypercellularity can mimic malignancy, but mitotic figures or pleomorphic cells indicative of a true soft tissue sarcoma are absent. Although myxomas are exclusively benign, nearly 25% of initial pathologic diagnoses are of malignant myxoid variants in-

cluding myxoid liposarcoma, myxosarcoma, and extraskeletal myxoid chondrosarcoma [1]. Cystic degeneration is common, and thought to be related to motion from the adjacent joint. JAM can be either intra- or extracapsular in origin, and in approximately 22% of cases is associated with the menisci of the knee. One-third of JAMs occurring about the meniscus are associated with meniscal tears, and similar lesions have been described within menisci removed for other reasons [2, 3].

Although JAM and the more common intramuscular myxoma may appear similar histologically, intramuscular myxomas are associated with little or no cystic change such as that usually seen in JAM [1]. Intramuscular myxomas have been reported to show homogenous low signal intensity in T1-weighted images and homogeneous very high signal intensity in T2-weighted images [4–6]. In contrast, the JAM of this report was inhomogenous, and showed a pattern of low signal intensity in T1-weighted sequences and increasing signal intensity which did not approach that of fluid with progressive T2-weighting. This feature is difficult to explain, as the histology was quite similar to that of myxomatous lesions previously described.

There is much confusion in the literature regarding benign lesions occurring around the knee. Numerous reports exist describing lesions which may contain myxoid tissue and thus histologically appear similar to JAM, including meniscal cysts [2, 7–17], parameniscal cysts [8, 9, 18], and ganglion cysts [19, 20].

Meniscal cysts typically occur in men aged 30–40 years with a history of previous knee trauma. The cysts are associated with a horizontal cleavage tear of the meniscus [7, 16] and can dissect through the base of the meniscus to ultimately present as a mass at the joint line. The lateral meniscus is affected more often than the medial, possibly a reflection of its increased exposure to trauma [21].

Parameniscal cysts are much less common than meniscal cysts. They are often included as “cyst variants” in large series [9, 10, 18]; however, a distinction between parameniscal and meniscal cysts has been made by several authors [9, 18]. Parameniscal cysts, both intra- and extracapsular, are lined by true epithelium, a feature not seen in meniscal cysts [18, 22].

Ganglion cysts differ from JAM primarily with regard to their myxoid component. In ganglion cysts, the myxoid tissue is much less developed than in JAM. Ganglia tend to occur more frequently in women on the dorsum of the hand or wrist, although they have been reported about the knee [19]. Ganglia average 1.5–2.5 cm in diameter, and are thus usually much smaller than JAM. Ganglia generally contain multiple thick septations, and do not possess a true epithelial lining such as that seen with parameniscal cysts. JAM may contain cystic spaces which resemble ganglion cysts in various stages of development [1].

Pigmented villonodular synovitis (PVNS) occurs within the same age range as JAM, but with a slightly

greater frequency in females. Cortical erosion and localized osteoporosis accompanying a soft tissue mass are the classic radiographic findings. MR imaging classically shows a mass which is of relatively low signal intensity in both T1- and T2-weighted images, a reflection of the high content of hemosiderin within the lesion. The knee is most often affected. PVNS may be found either in a diffuse form which involves the entire joint or in a focal nodular form. In the focal form affecting the knee, the anterior aspect of the joint is most commonly involved. Extra-articular lesions of this type, known as tenosynovial giant cell tumor of the diffuse type or giant cell tumor of tendon sheath, are rare.

Desmoid tumors represent deep fibromatosis. They arise from striated muscle fascia, and are therefore extra-articular lesions. Desmoid tumors may demonstrate either low or high signal intensity in T2-weighted images [23].

In summary, this 9-year-old girl presented with a painful mass in the posterolateral aspect of her right knee. Excisional biopsy of the mass revealed a juxta-articular myxoma. The differential diagnosis for such an intra- or periarticular mass should include meniscal cyst, parameniscal cyst, ganglion cyst, juxta-articular myxoma, focal nodular PVNS, and desmoid tumor. Current imaging techniques do not allow differentiation between these benign lesions

and low grade myxoid-containing sarcoma.

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