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Case report 609

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Radiological studies

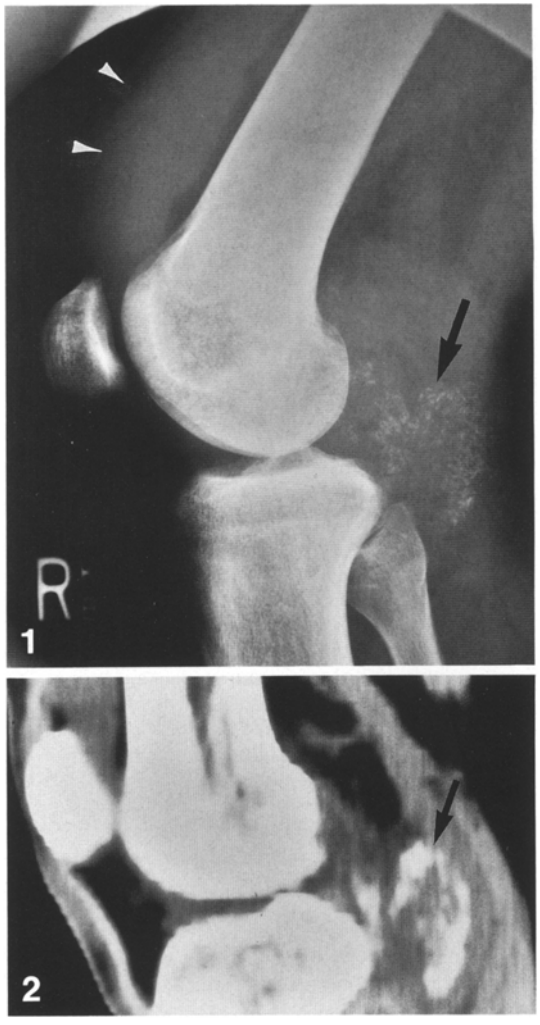

Fig. 1. A lateral radiograph of the right knee shows multiple punctate calcific densities (*arrows*) posterior to the joint. A suprapatellar effusion (*arrowheads*) is also noted

Fig. 2. Reformated sagittal CT confirms the presence of posterior calcific densities (*arrows*), but does not determine if they are in the joint space

Clinical information

This 32-year-old man presented with a 6-month history of stiffness and swelling in his right knee as well as a "crackling" motion at the joint. Initial radiographs (not available) were interpreted by the family physician to be normal. His symptoms progressed for the next 4 months at which time he was referred to an orthopedist.

Radiographs at that time revealed a discrete area of small punctate calcifications posterior to the knee thought not to involve the joint space (Fig. 1). Based on these radiographic findings a soft tissue tumor was suspected.

An open biopsy was performed, resulting in an initial histological diagnosis of "well-differentiated chondrosarcoma."

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Diagnosis: Synovial (osteo)chondromatosis

Pathological features

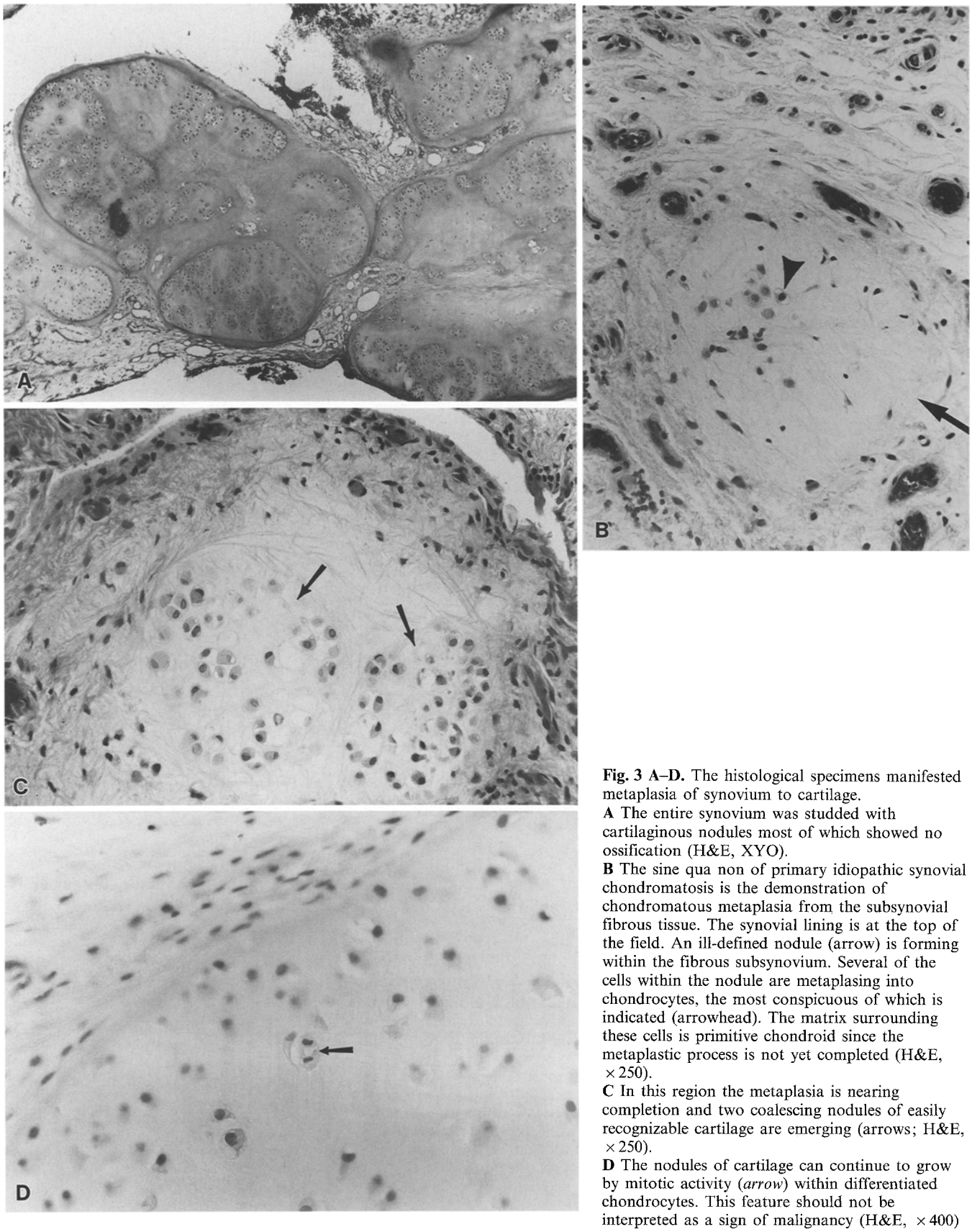

Fig. 3 A–D. The histological specimens manifested metaplasia of synovium to cartilage.
A The entire synovium was studded with cartilaginous nodules most of which showed no ossification (H&E, XYO).
B The sine qua non of primary idiopathic synovial chondromatosis is the demonstration of chondromatous metaplasia from the subsynovial fibrous tissue. The synovial lining is at the top of the field. An ill-defined nodule (arrow) is forming within the fibrous subsynovium. Several of the cells within the nodule are metaplasing into chondrocytes, the most conspicuous of which is indicated (arrowhead). The matrix surrounding these cells is primitive chondroid since the metaplastic process is not yet completed (H&E, ×250).
C In this region the metaplasia is nearing completion and two coalescing nodules of easily recognizable cartilage are emerging (arrows; H&E, ×250).
D The nodules of cartilage can continue to grow by mitotic activity (*arrow*) within differentiated chondrocytes. This feature should not be interpreted as a sign of malignancy (H&E, ×400)

The differential diagnoses include chondrosarcoma, paraarticular chondroma, post-traumatic osteochondromata within a popliteal cyst, synovial chondromatosis limited to a popliteal cyst, degenerative joint disease with loose bodies, synovial sarcoma, synovial hemangioma, villonodular synovitis, and lipoma arborescens.

Further clinical information

After the diagnosis of chondrosarcoma was made, the patient was referred to a surgical oncologist. Upon reviewing the radiographs, the oncologist and the radiologist believed that the more likely diagnosis was synovial osteochondromatosis. Three skeletal pathologists independently diagnosed "extra-articular chondromatosis" (see pathological illustrations).

An arthrogram was performed. The arthrogram confirmed joint involvement of the knee by revealing suprapatellar filling defects characteristic of synovial chondromatosis (Fig. 4A), but failed to prove that the region of posterior calcification communicated with the joint (Fig. 4B). Surgical excision confirmed the presence of intraarticular synovial osteochondromatosis, and a posterior and anterior synovectomy was performed. The patient recovered without complications.

Two months later radiographs showed recurrent calcifications. The patient was treated with physical therapy and observed. Four years later the recurrent osteochondromatosis had become extensive (Figs. 5 and 6).

Discussion

The first description of synovial chondromatosis appeared in 1813 when Laennac hypothesized that loose joint bodies originated from synovial and subsynovial tissues. Brodie suggested an extrasynovial origin in 1836. Lexer, in 1907, proposed that activated embryonal cell rests in the synovial membrane were the source of intraarticular chondromatosis [22]. Jaffe and others first championed the current view that "concrete histologic evidence of cartilagenous metaplasia of the synovial

Additional imaging studies

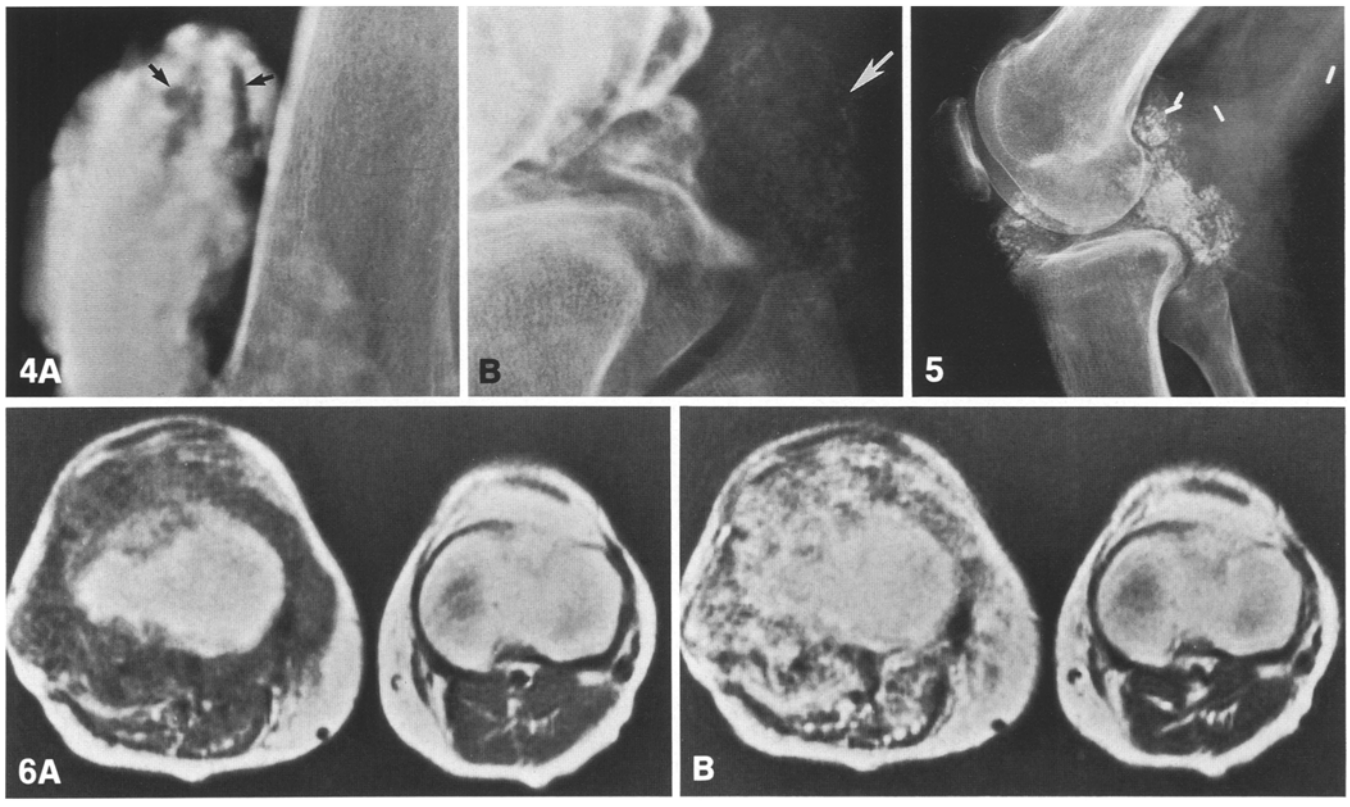

Fig. 4A, B. Single contrast arthrogram. **A** Observe the multiple filling defects (*arrows*) in the suprapatellar bursa. **B** The arthrogram fails to demonstrate a communication from the joint to the area of the calcifications (*arrows*)

Fig. 5. A lateral radiograph 4 years after synovectomy shows recurrent calcified nodules measuring only a few millimeters in diameter present throughout the joint

Fig. 6. MRI. **A** An axial T1-weighted image (Se 343/30) shows a large mass of inhomogeneous signal intensity filling and surrounding the right knee joint. Erosion of the tibial plateau is present. **B** An axial T2-weighted image (Se 2232/85) shows the mass to remain inhomogeneous, with areas of fluid with both very high signal intensity and numerous punctate areas devoid of signal representing the calcifications of synovial osteochondromatosis. The high signal intensity within the muscles is thought to represent marked edema of muscle

connective tissue" is necessary for the diagnosis, but nodules may become either calcified or ossified at which time the lesion is termed synovial osteochondromatosis [9].

Synovial (osteo)chondromatosis (SOC) is known to develop from synovial metaplasia, but the underlying etiology remains unknown [23]. An infectious cause is unlikely since a synovial inflammatory reaction is not observed. Since this disease occurs in adulthood, an embryologic cellular origin is unlikely. Trias et al. [23] reported that a history of trauma was absent or denied in 67% in their Canadian patients. A neoplastic theory would account for the occasional recurrence and exceptionally rare malignant transformation. Most investigators now accept a theory of hyperplastic metaplasia, first developed by Freund in 1937 [7].

Due to the confusion about the etiology of SOC, various terms are used to describe this entity [23]. Synovial chondrosis, occasionally used by the French and Italians, implies degenerative exfoliation. The Canadians prefer synovial chondrometaplasia, descriptive of the current theory of metaplasia of synovial cells into cartilage. Other terms alluding to the entity are articular eccondrosis, synovial chondromatosis, and synovial osteochondromatosis. Some have even referred to it as osteochondritis dissecans or even simply loose bodies. The most popular term in the United States is synovial chondromatosis (or osteochondromatosis).

Primary SOC appears twice as often in men as in women, and occurs most frequently between the ages of 30 and 50 [1, 23]. A familial incidence is extremely rare [23]. One report indicates that the knee is affected in two-thirds of cases followed in order by the hip, elbow, wrist, ankle, shoulder, temporomandibular, and interphalangeal joints [22]. Two reports list the shoulder to be the second most affected joint [1, 23].

Secondary SOC is characteristically associated with trauma, degenerative joint disease, and neurological disease, with the patients ranging in age from 6 to 88 years; the majority are under 20 and over 60 years of age. The most commonly involved joints in secondary SOC in descending order are the knee, hip, elbow, shoulder, and ankle [11]. Secondary SOC can often be polyarticular while primary SOC is virtually always monoarticular [11]. Extraarticular synovial chondromatosis (tenosynovial chondromatosis) is rare and appears most often unifocally in the foot, hand, wrist, and biceps tendon sheath, while the knee is the most commonly involved major location [10, 16]. No age or predilection for a gender has been identified for extraarticular synovial chondromatosis.

Patients with either primary or secondary SOC typically present with a greater than 2-year history of intermittent, progressive joint pain which may become constant [23]. Clicking, grating, and locking of the affected joint are common [1]. Loose bodies may compress a nerve, causing a peripheral neuropathy [23]. Swelling, decreased range of motion, and muscular atrophy may also occur [16, 23]. Patients with extraarticular SOC present with slowly enlarging, painful soft tissue masses with occasional decrease in range of motion [10]. Physical examination of patients with SOC reveals single or multiple palpable loose bodies, tenderness, crepitus, local muscular atrophy, boggy synovitis, and occasionally effusion [22].

The various imaging (plain films, arthrography, CT, and MRI) manifestations of primary SOC can be divided into early and late stages based on the demonstration of calcification. Before calcification is visible, radiographs may show only a vague fullness near the joint or in the bursa [1, 22]. However, SOC without visible calcified or ossified densities is the exception rather than the rule [9]. Radiographs of secondary SOC are less often without calcification [11]. Advanced SOC typically reveals innumerable, stippled calcified nodules which may vary in size from several mm to as much as 1 cm. However, nodules of uniform size are the rule in any individual patient [16]. Calcified (or ossified) nodules of secondary SOC are usually less regular in shape, larger in size, and contain "ring-like" calcifications unlike those of primary SOC [1, 11]. Various investigators have unsuccessfully attempted to determine how many loose bodies are necessary for a radiological (or other imaging) diagnosis of SOC. For example, Trias et al. [23] reported more than five loose bodies seen in 55% of proven cases. Joint effusion and synovial thickening may be observed in the radiograph [16, 22]. Although the integrity of the articular surfaces is usually intact, occasionally nodules will cause pressure erosion of the subchondral or periarticular bone as shown in our case [11]. The defects will have a sclerotic rim, but no periosteal new bone formation. These secondary degenerative changes occur rarely and are due to long-standing mechanical trauma [16, 22, 23]. Extraarticular SOC appears as a soft tissue mass near or removed from an adjacent joint [10]. This mass usually contains small, stippled opaque densities or several large opaque bodies that are closely approximated, creating a lobulated appearance [10]. Bony erosion is unusual in extraarticular SOC [10]. The resulting imaging appearance may be identical to that of a soft tissue chondroma [16].

In our case, the MRI was useful in demonstrating the extent of the disease, but could not assist in establishing the diagnosis. In fact, the diffuse abnormal signal intensity within the soft tissues surrounding the knee and adjacent erosion of the tibial plateau would favor the diagnosis of malignancy rather than an aggressive benign process. The punctate foci of low signal intensity within the tumourous mass imply the presence of either calcium or dense fibrous tissue [15], but determination of their nature was best accomplished radiographically. The areas of high signal intensity within the mass in the T2-weighted image are thought to represent marked edema of muscle tissue.

Computed tomography may aid in the diagnosis and determination of the extent of the lesion [10, 23]. Since the early 1970s, arthrography has been employed to differentiate synovial chondromatosis from other disorders of the knee joint. Crittenden [4] reported multiple, spheroid filling defects in arthrographic studies of SOC, and Prager [18] reported that these defects occasionally coalesce to form larger masses within the joint. Arthrographic evidence of uncalcified, radiolucent intraarticular nodules excludes extraarticular SOC. In

our case, failure to fill the popliteal cyst by intraarticular injection was attributed to obstruction o the neck of the cyst by osteochondromatous bodies.

The differential possibilities for this case were varied. The opacities were numerous, uniform in size, and small [16]. A fewer number of loose bodies would be expected with degenerative joint disease [4, 16]. Synovial hemangioma may show multiple filling defects on arthrography but only a few calcifications on plain films. Synovial sarcoma shows a dystrophic type of calcification in about one-third of cases and a large mass on arthrography and MRI, generally near but not in the joint [16]. Less than 3% of synovial sarcomas are actually intra-synovial [12]. Confluent, frond-like or fungating bodies would be expected in pigmented villonodular synovitis [4] which lacks radiographically evident calcification [2]. Lipoma arborescens, a rare lobulated fatty tumor, produces nodules which are irregular and ill-defined. The latter diagnosis would also be unlikely because of the lack of characteristic palpable soft nodules [16] and findings of fatty deposits on CT and MRI. Para-articular chondroma could simulate the radiographic and CT findings. However, para-articular chondroma is excluded by the numerous intra-articular filling defects in the arthrogram [13]. Post-traumatic osteochondromas within a popliteal cyst might be considered [5]; however, post-traumatic osteochondromas are less numerous and more densely calcified. Synovial chondromatosis limited to a popliteal bursa was a consideration prior to the arthrogram.

The most critical entity to exclude was chondrosarcoma – the initial diagnosis. Primary chondrosarcoma of soft tissues or a joint is an extreme rarity, with the exception of a specialized variant of chondrosarcoma known as the chondroid tumor (extraskeletal myxoid sarcoma). Extraarticular myxoid chondrosarcomas (chondroid sarcomas) are, in fact, usually found in the thigh or popliteal fossa [19]. Within the joints and soft tissues cartilage lesions are benign to about the 98th percentile [12]. Yet it is important to exclude an intramedullary chondrosarcoma with extension into adjacent soft tissues or a joint. CT and MRI scans should clarify the diagnosis in such cases.

Could this have been SOC that had undergone transformation into chondrosarcoma? Perry reported the only conclusive case to date of malignant transformation. In his case, grade II undifferentiated chondrosarcoma was found in SOC of the knee 24 years after the original diagnosis [17]. In our case, malignant chondrocytes were not seen by any of the consulting skeletal pathologists. However, the key to the radiographic diagnosis of intraarticular SOC was the presence of the multiple filling defects detected on arthrography. Histological verification of the diagnosis was established by the identification of chondrometaplasia of the synovium.

In *summary*, a 32-year-old man presented with a mass behind the right knee. The original plain films showed only calcification behind the right knee. Excisional biopsy was incorrectly interpreted as a soft tissue chondrosarcoma. Arthrography and review of the histological sections were important in establishing the correct diagnosis of intraarticular synovial osteochondromatosis extending into a Baker's cyst. This case illustrates the tendency for synovial osteochondromatosis to recur even after synovectomy.

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