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SYNOVIAL CHONDROSARCOMA ARISING FROM SYNOVIAL CHONDROMATOSIS OF THE KNEE

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We report the case of a 51-year-old woman who had suffered from right knee pain and stiffness for 40 years. Her symptoms had gradually worsened over the past 5 years. One year previously, when she first visited our clinic, plain radiographs and CT scan images had revealed synovial chondromatosis over the right knee. At the current admission, follow-up MRI showed synovial masses around the knee and worsening of endosteal cortical scalloping in the patella, femoral condyle, and tibial plateau. After diagnosis on the basis of frozen sections, the patient had total excision of the lesions and total knee athroplasty. Histological examination revealed synovial chondromatosis in the joint cavity and grade 1 chondrosarcoma invasion into the adjacent bone. In long-standing synovial chondromatosis, presentation with aggravated symptoms and deterioration on imaging findings should alert clinicians to the potential for malignant change.

Key-words: Knee, neoplasms – Sarcoma.

Malignant transformation of synovial chondromatosis to chondrosarcoma is documented but rare (1, 2). The clinical features alone are not helpful for distinguishing them, and no definite imaging criteria can differentiate synovial chondromatosis from low-grade synovial chondrosarcoma (2-5). Synovial chondromatosis is very difficult to distinguish from secondary synovial chondrosarcoma solely on clinical, radiological or histological criteria. The diagnosis is usually made on the basis of a combination of clinical, imaging and histological criteria, as we did in our present case.

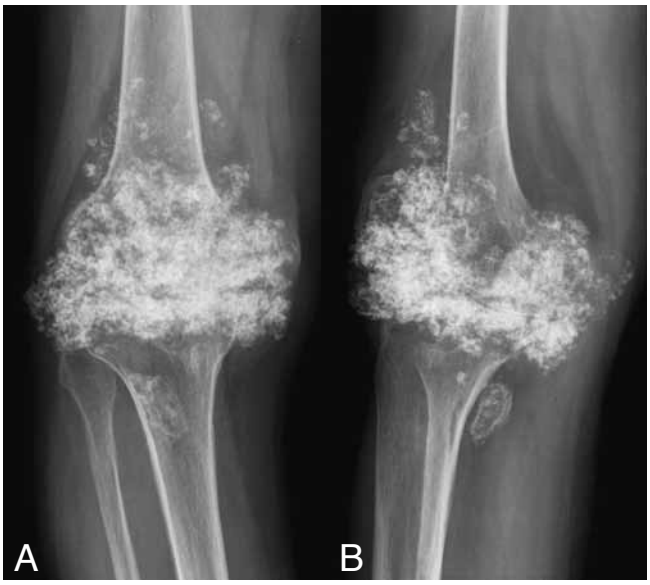

Fig. 1 — A. Anteroposterior and B. lateral projection of knee radiographs show extensive intra-articular synovial chondromatosis.

Case report

A 51-year-old woman had a 40-year history of right knee pain and stiffness. She had injured her right knee and had been treated with surgery 27 years earlier. Her symptoms had gradually worsened over the past 5 years until she had become unable to walk. One year earlier, she had visited our outpatient clinic, when plain radiographs of the knee showed numerous intra-articular mineralized bodies of similar sizes over the right knee (Fig. 1). Computed tomography (CT) scan images revealed both calcified and ossified nodules (Figs. 2A, 2B). No evidence of degenerative osteoarthritis was noted.

At this admission, physical examination revealed a large, hard, fixed mass over the right knee, with a surgical scar. Her right knee was swollen, tender, and completely stiff at full extension. The knee had a zero-degree range of motion. Follow-up X-rays of the knee revealed no remarkable interval change of the synovial chondromatosis. Magnetic resonance imaging (MRI) showed deterioration in the distribution of intra-articular synovial masses (Figs. 2C, 2D and 3). The synovial mass was lobulated, with fibrotic septa delineated by a small joint effusion, and largely involved the cruciate ligaments and the posterior capsule, with endosteal cortical scalloping in the patella, femoral condyle and tibial plateau. There were also lobular nonenhancing structures, with hyperintense signal on T2-weighted images, consistent with intra-articular cartilage. The calcified bodies were depicted as signal voids whereas ossified nodules were shown as high-signal-intensity marrow surrounded by low-signal-intensity cortical bone. The chest X-ray was negative.

A frozen biopsy section was examined three days later and

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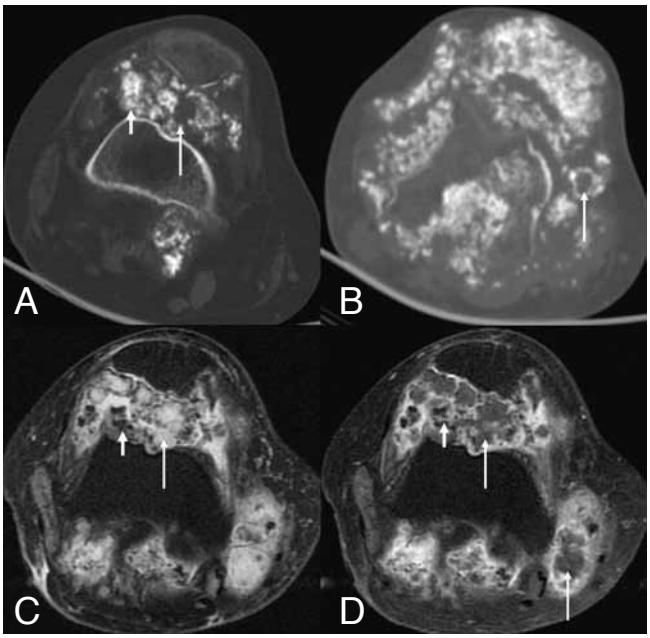

Fig. 2 — A, B. Axial CT images confirm many calcified bodies (small arrow) and central lucent nodules (long arrow) in the suprapatellar bursa and some in the posterior capsule. C. Axial proton-density and D. postgadolinium MRI images 14 months later show multiple lobular cartilaginous masses (long arrow) (corresponding to central lucent nodules on CT images), with patellar erosions, and anterior and posterior extension signal-void calcifications (small arrow).

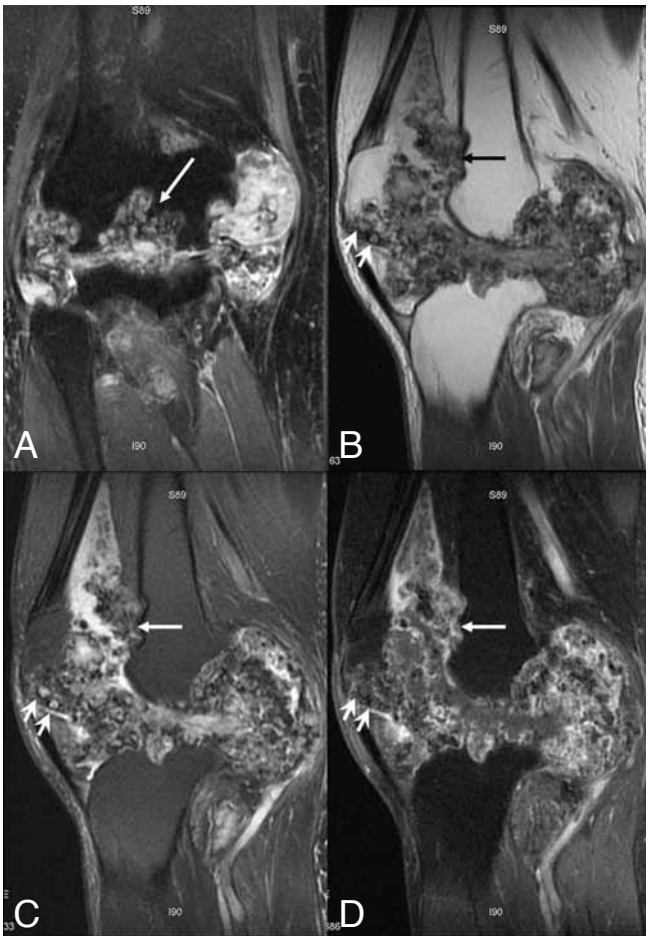

Fig. 3 — A. Sagittal proton-density MRI image shows extensive synovial mass with multiple bone erosion (arrow). B. T1-weighted MRI image, C. T2-weighted image, and D. postgadolinium image show numerous signal-void calcifications, both attached to and free from the synovium, with erosion about the adjacent bone (long arrow), and joint effusion (hyperintense signal on T2-weighted image but nonenhancing on postgadolinium image) and synovial thickening with obvious enhancement on postgadolinium image. There are some ossified bodies with MRI signal similar to that of bone marrow (short arrows). Note that the histologically proven intra-articular area of malignant transformation adjacent to the patellar erosion shows nonspecific minimal enhancement.

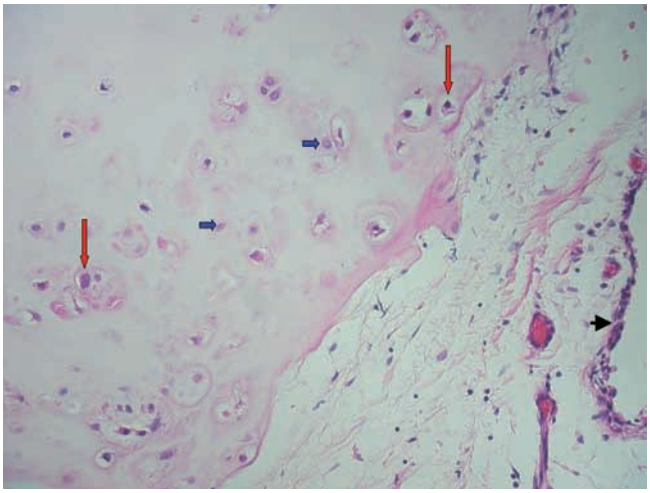

Fig. 4 — Photomicrograph shows nodular proliferation of chondroid tissues containing benign (short arrows) and malignant (long arrows) chondroid cells in the synovial tissue lined with synovial cells (arrowhead).

showed low-grade chondrosarcoma. Total excision of the lesion and total knee arthroplasty were then performed. The histological diagnosis was grade 1 chondrosarcoma arising from synovial chondromatosis (Fig. 4). There were multinodular cartilaginous islands and loose bodies in the joint cavity. Stromal myxoid changes and atypical chondrocytes with loss of clustering and spindling were noted. The chondrosarcoma had low cellularity, mild nuclear atypia, chromatin clumping, little mitotic activity, and invasion into the patellar, femoral and tibial bone marrow.

Discussion

Primary synovial chondromatosis is a benign, proliferative and metaplastic disease arising from the synovial membrane (6). It is characterised by the formation of multiple cartilaginous bodies of equal size, sometimes with calcification or ossification, in the synovium of joints, tendons, and bursae; similar findings were noted in our case. A useful differentiating feature is that the primary form is relatively uncommon and synovial masses are the predominant feature, whereas secondary synovial chondromatosis is rela-

tively common and is typified by the presence of cartilaginous bodies against a background of degenerative joint disease (1, 2, 6).

Primary synovial chondromatosis is preponderant in middle-aged and elderly male patients, with a mean age of 41 years (range, 17-76 years) and a male-to-female ratio of 1.5 to 1.0 (1, 2). The knee is the most common site of involvement (1). Clinically, symptoms include pain, limitation of the range of joint movement because of detached proliferating nodules, swelling due to synovial thickening or effusion, and palpable masses. All of the symptoms are nonspecific.

One study reported that 50% of patients with synovial chondromatosis had radiographic evidence of marginal bone erosion (2). CT may show noncalcified bodies, and long-standing osteochondromatosis presents as an aggressive lesion attached to the adjacent bone, but plain X-ray cannot distinguish bone from overlapping ossified or calcified mass lesions. On MRI, the typical T1-weighted findings of synovial chondromatosis are multiple signal-void lesions in the joint caused by the calcification of the loose bodies. MRI reveals more erosion than radiography (80% compared with 50%) (2), and 45% of patients have joint effusion (1). MRI typically demonstrates lobulated lesions and is the best imaging tool to assess the intramedullary extent of the tumour.

Kramer et al. reported three distinct MRI patterns of synovial osteochondromatosis (7). Pattern A was characterized by lobulated masses with multiple areas of hypointense septa due to fibrosis. Both plain X-ray and CT scan showed no calcification, making diagnosis difficult. Pattern B, the most common pattern, was characterized by pattern A plus foci of signal void, corresponding to calcifications on plain film and CT scan. Pattern C had features of patterns A and B plus foci of peripheral low signal surrounding a central fat-like signal, corresponding to areas of ossification; a similar feature was noted in our case. Milgram classified loose bodies secondary to synovial osteochondromatosis on histology into three subgroups (8): (1) active intrasynovial disease with no loose bodies, similar to Kramer's MRI pattern A; (2) intra-articular cartilaginous nodules with free loose bodies with nidi, consisting of the lobular type of cartilage; (3) multiple loose bodies with nidi, with no active intrasynovial disease, similar to Kramer's pattern C, although this example may be controversial.

Malignant transformation to chondrosarcoma arising from primary chondromatosis accounts for 5-10% of cases (1, 2). Malignant change should be suspected if the patient has a long clinical history or a rapidly enlarging mass or a recurring mass following excision, with evidence of deterioration on imaging (2, 9), as did our patient. A progressive or newly developed periosteal reaction, cortical destruction or erosion, or marrow invasion can indicate potential malignant transformation to chondrosarcoma (2, 9). In retrospect, MRI images showed minimal enhancement of the histologically proven area of malignant transformation adjacent to patellar erosion; this finding is nonspecific. Detection of lung metastases also is a clear imaging sign of malignancy (9).

No definite histological criteria differentiate synovial chondromatosis from a low-grade synovial chondrosarcoma (1, 3, 4). Murphy et al. (6) reported that 28 patients with chondromatosis had histological evidence of atypical cells, and none subsequently developed malignancy. Both had similar features of hypercellular and atypical hyaline cartilage. Signs of malignancy were seen in necrosis within the tumor and permeation of trabecular bone. Anract et al. (10) proposed the following criteria to diagnose malignant transformation: 1) histological diagnosis of synovial chondromatosis established before diagnosis of chondrosarcoma; 2) histologic diagnosis of chondrosarcoma at the same anatomical site as the synovial chondromatosis; 3) diagnosis of chondrosarcoma and synovial chondromatosis in the same resection specimen. Our case was in accordance with the last two criteria.

The treatment of suspected malignant transformation to chondrosarcoma must be a wide resection or an amputation (10). However, the preoperative risk still lies in the misinterpretation of the synovial chondromatosis as chondrosarcoma (10). Long-standing synovial chondromatosis presenting with aggravated symptoms and deterioration on imaging findings should alert clinicians to the potential for malignant change. The diagnosis is usually made on a combination of clinical, imaging and histological criteria.

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