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CASE REPORT

Primary synovial chondrosarcoma of the hip joint in a 45-year-old male: case report and literature review

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Abstract Synovial chondrosarcoma is a rare tumor, seen most commonly arising from antecedent synovial chondromatosis, the more common benign entity. The distinction between the two can be difficult on the basis of clinical, imaging, and histologic criteria. The authors report a case of pathologically proven synovial chondrosarcoma of the hip in a 45-year-old male initially treated for presumed synovial chondromatosis. The case is made more unusual by the fact that no evidence of co-existent synovial chondromatosis was noted at histology. The literature as regards synovial chondrosarcoma, both de novo and secondary cases, is reviewed.

Keywords Synovial chondrosarcoma · Synovial chondromatosis · Intraarticular soft tissue mass · MRI · Histology

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Introduction

Synovial chondrosarcoma (CHS) is an unusual tumor that arises from the synovium of the large joints, particularly the knee and hip. Most reported cases of synovial chondrosarcoma are categorized as secondary originating from pre-existing synovial chondromatosis [1]. Rarely, synovial chondrosarcoma is felt to be primary in origin with no antecedent history of or histological evidence of coexistent synovial chondromatosis [1, 2]. The distinction between synovial chondromatosis and synovial chondrosarcoma can be difficult with significant overlap in both the radiologic and histologic features. The treatment, however, is significantly different with synovial chondromatosis treated with resection of the cartilaginous nodules with or without synovectomy and synovial chondrosarcoma often requiring wide resection, and, in some cases, amputation.

The purpose of this paper is to present an unusual case of primary synovial chondrosarcoma of the hip and to review the clinical, radiologic, and histologic features that can help distinguish this unusual malignancy from the benign entity of synovial chondromatosis.

Case report

The patient was a 45-year-old male who presented with a history of vague pain in his left hip region for 5 years. Prior to being seen at the authors' institution, he had not sought medical advice or been imaged. On clinical examination, he was noted to have slight limitation in range of motion of the left hip and mild discomfort with rotation.

Radiographs and a magnetic resonance imaging examination (MRI) of the pelvis and left hip were performed. The radiographs were remarkable for a large number of

irregularly shaped ossific densities in a periarticular distribution in the left hip (Fig. 1a, b). There was relative preservation of joint space. An area of erosion was noted along the anterolateral margin of the femoral neck on the frog leg lateral view (Fig. 1b) with underlying lytic and sclerotic change. The cortex over the foveal margin of the femoral head was indistinct with suggestion of underlying lucency.

On MRI, there was a large amount of lobular soft tissue centered within the joint space, which was mildly heterogeneous, but predominantly isointense to muscle on T1 (Fig. 2a) and hyperintense on T2-weighted images (Fig. 2b). There were deep areas of excavation along the foveal margin of the femoral head, the medial acetabular wall and in numerous locations along the femoral neck, most prominent anteriorly where the soft tissue material extended into the medullary cavity of the femur.

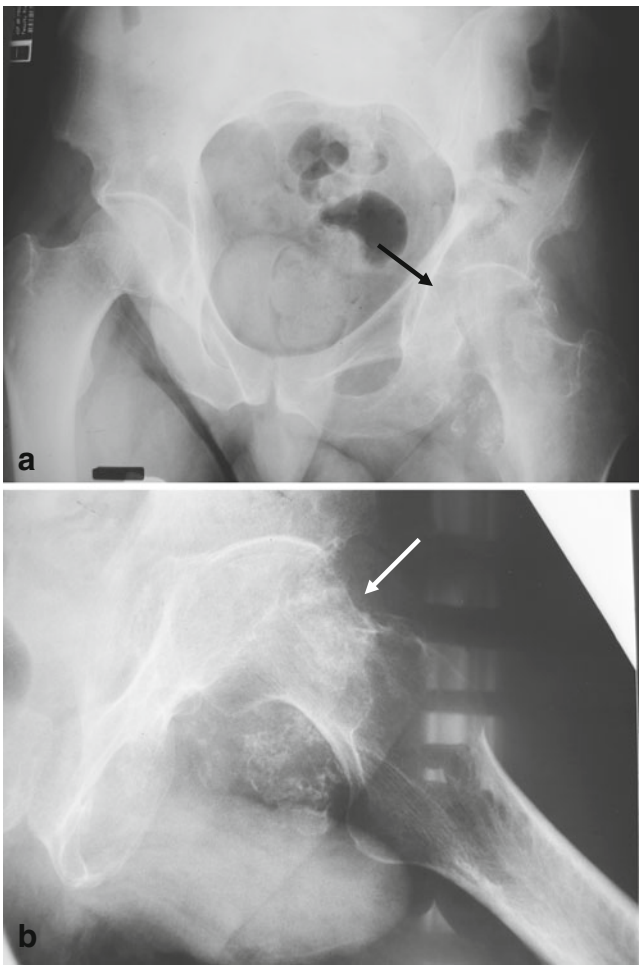

Fig. 1 An AP radiograph of the pelvis (**a**) and frog leg lateral view of the left hip (**b**) demonstrating irregularly shaped ossific densities in a periarticular distribution in the left hip. Note the loss of definition of the foveal margin of the left femoral head on the AP film (*black arrow*) and area of erosion along the anterolateral margin of the femoral neck on the frog leg lateral (*white arrow*)

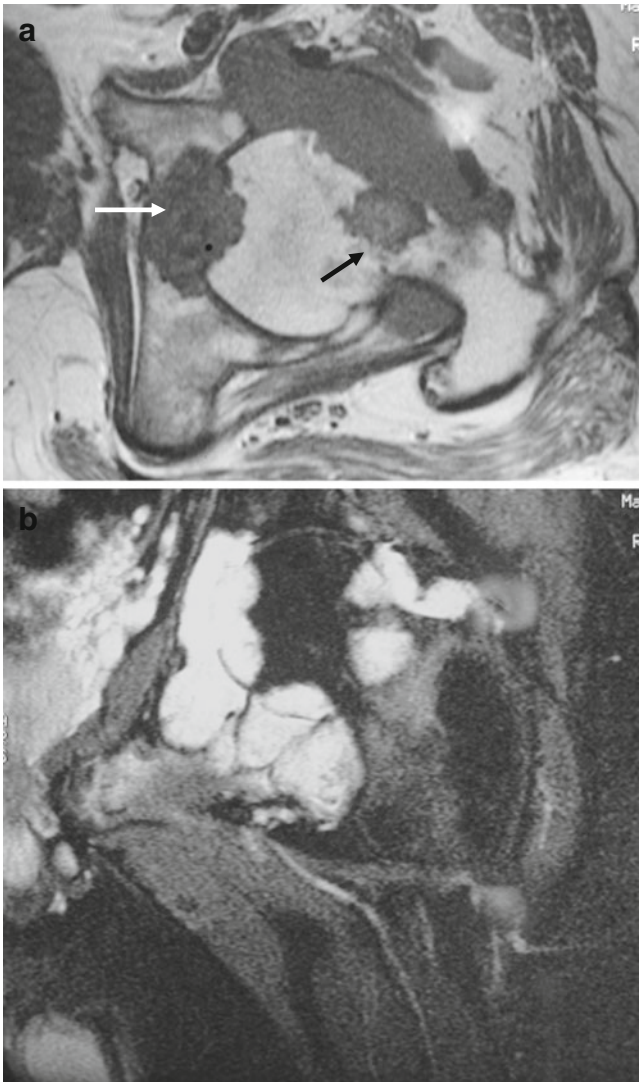

Fig. 2 Axial T1-weighted (**a**) and coronal T2 fat-saturated (**b**) images of the left hip from an MRI demonstrating extensive lobular intra-articular soft tissue predominantly isointense to muscle on T1 and hyperintense on T2. There are multiple areas of erosion along both the femoral and acetabular joint margins. Note the area of central low signal within the central joint space on the axial image (*white arrow*) felt to reflect calcification and the more irregular and "permeative" border of the tumor with the bone along the anterolateral margin of the femoral neck (*black arrow*)

Based on the imaging findings, the patient was felt most likely to have synovial chondromatosis. Due to the extent of disease and erosion of the articular margins, a decision was made by a general orthopedic surgeon to partially excise the lesion and perform a total hip arthroplasty.

Gross examination of the surgical specimen revealed numerous irregular fragments of grayish-white, firm cartilaginous tissue with areas of calcification measuring in aggregate 6×5×4.5 cm (Fig. 3). No discrete nodules, free or attached to synovium, were identified in the tissue. The resected femoral head and neck were also received and

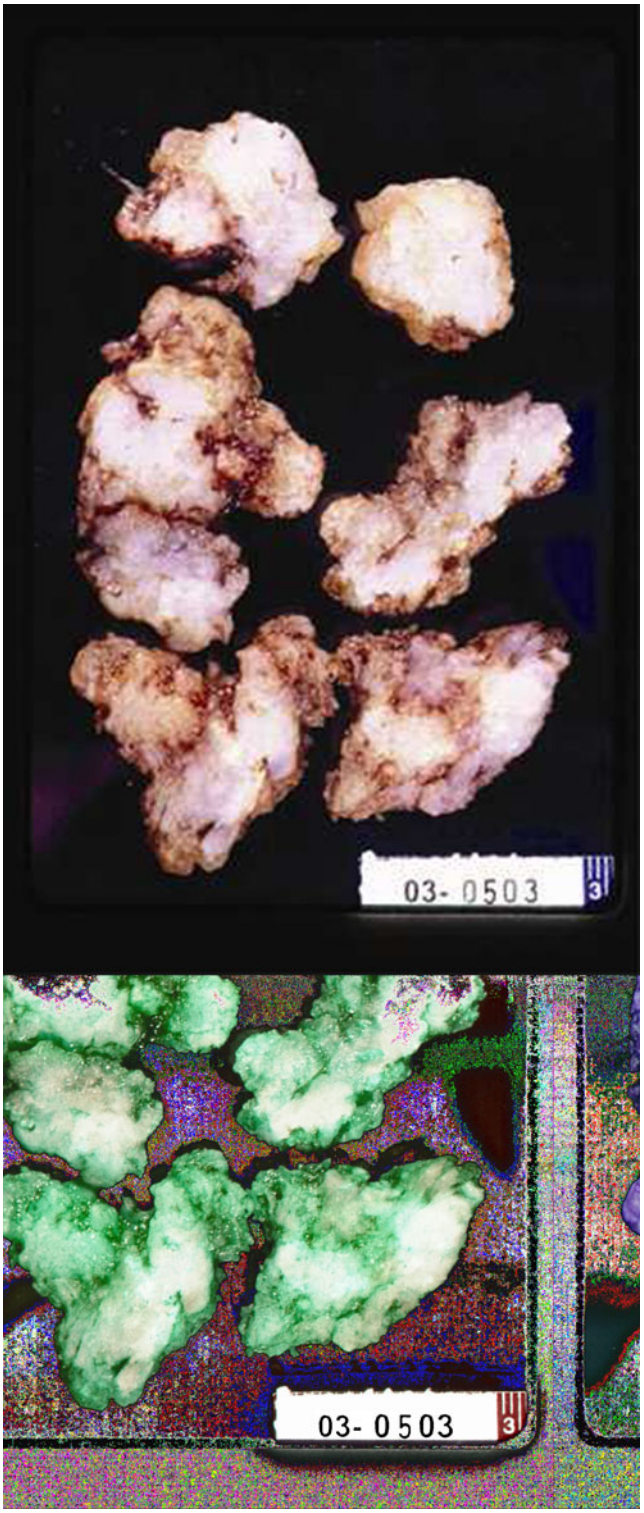

Fig. 3 Tumor from the hip joint showing fragments of grayish-white, firm tissue with a cartilaginous appearance and a gritty cut surface. No nodular structures typical of synovial chondromatosis are seen

demonstrated two separate areas of cartilaginous tumor at the articular surface of the femoral head in the region of the fovea and along the posterior aspect of the neck. On coronal sections, the tumor in the fovea measured 3×3×1.5 cm and demonstrated irregular borders with invasion into the subchondral bone. The tumor along the posterior aspect of the femoral neck measured 1.5×1.5×1 cm and had similar features with irregular margins, cortical destruction, and invasion of the underlying medullary cavity (Fig. 4a). The specimen x-ray confirmed the ill-defined borders of the tumor in the femoral head (Fig. 4b).

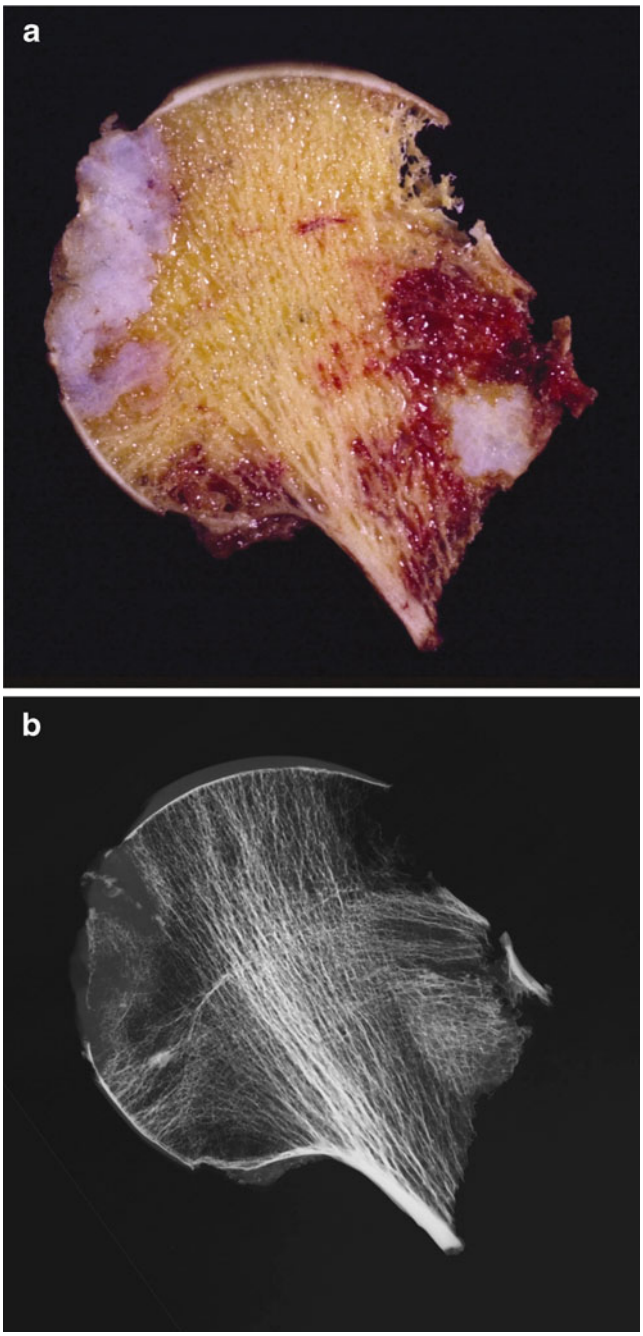

Fig. 4 **a** Coronal section and **b** corresponding radiograph of the femoral head and neck demonstrating foci of cartilaginous tumor in the fovea and the posterior neck, which are infiltrating and destroying the bone. Note the irregular invasive borders of the tumor

Microscopic sections revealed a low grade CHS exhibiting a moderate degree of cellularity and poorly defined nodular pattern of growth. The majority of the lesion consisted of cartilage cells within lacunae with uniform distribution in the tissue and occasional cluster formation. There was mild cellular atypia with mildly pleomorphic nuclei, focal areas of open chromatin and small nucleoli, and occasional binucleated cell. Focal calcification of the matrix was seen. No mitoses were identified (Fig. 5a). The majority of the tumor fragments, however, demonstrated peripheral areas of intermediate grade 2 CHS with prominent atypical nuclei ranging in size from round to

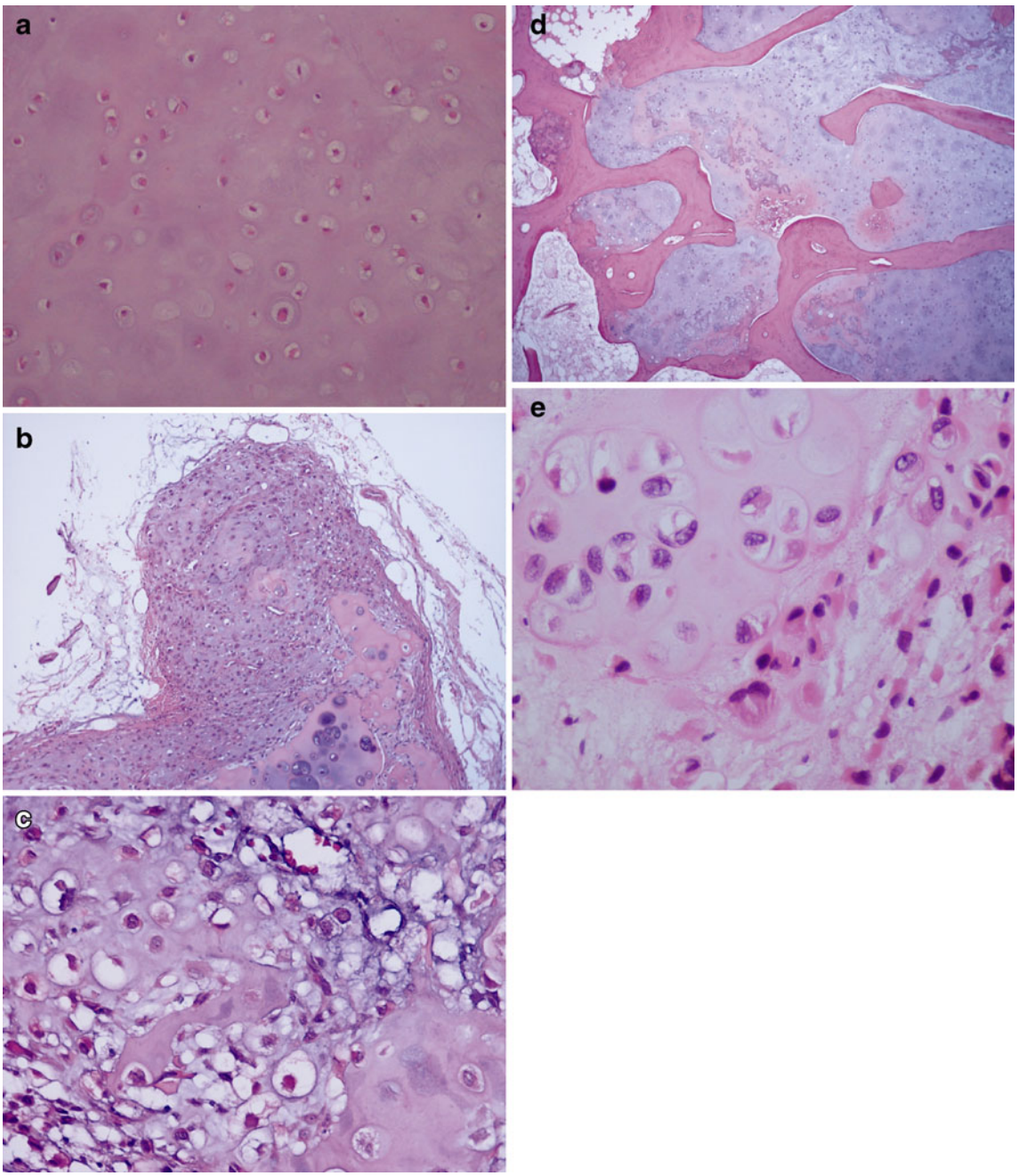

Fig. 5 **a** Representative micrograph of the tumor showing a low grade CHS with a benign histologic appearance. **b** Peripheral portion of the tumor in the hip joint showing increased cellularity, cellular atypia, and decreased matrix. Compare this with the well differentiated CHS at the bottom. **c** High-power view of the cellular area at the periphery with atypical cells having high nuclear-to-cytoplasmic ratio, open chromatin, and conspicuous nucleoli, consistent with CHS grade 2. **d** Low-power view of the femoral bone showing extensive infiltration of the marrow spaces by tumor, consistent with a low grade CHS. **e** High-power view of chondrosarcoma from the acetabular region demonstrating malignant chondrocytes with large atypical nuclei

ovoid or elongated with conspicuous nucleoli and high nuclear-to-cytoplasmic ratio. In some of these areas, the tumor cells appeared to be infiltrating freely into the surrounding soft tissue. The pericellular lacunae were sparse, and there was a decreased amount of pale blue matrix, which showed focal necrosis and liquefactive changes. No mitoses were seen (Fig. 5b, c).

The intraosseous components of the tumor within both the foveal and neck regions demonstrated histologic changes similar to those in the intra-articular portions. The majority consisted of well differentiated CHS with extensive marrow permeation, entrapment of pre-existing bone by tumor and focal areas of trabecular destruction (Fig. 5d). As with the intra-articular portion, there were peripheral elements that demonstrated an intermediate degree of malignancy (grade 2).

Normal synovial tissue was rarely seen and small fragments of membranous tissue resembling synovial membrane were present in the joint. There was no histological evidence of synovial chondromatosis.

A few months later, the patient was referred to an orthopedic tumor surgeon who removed the total hip replacement and performed a wide resection of the acetabulum and soft tissues around the hip joint. In addition, an extensive curettage resection and cryosurgery were performed of the femoral canal. The acetabulum was reconstructed with an acetabular cage and the femur with a long stem cemented femoral component and femoral strut allograft.

All tissue fragments removed during the second surgical procedure had similar features to the original tumor including permeation of the acetabular bone and invasion into the soft tissue. Small nodules of highly cellular grade 2 CHS were also present. No mitoses were seen (Fig. 5e). The degree of nuclear proliferative activity of the CHS was studied by immunohistochemistry using the Ki67 mononuclear antibody. The percent proportion of Ki67-positive nuclei was determined by counting 1,000 cells from representative areas. The proliferative index in the well differentiated areas was 0.3% and in the grade 2 areas was 4.8%.

Because of the lack of previous history of synovial chondromatosis and the absence of gross and histological evidence of synovial chondromatosis in the two surgical procedures, we concluded that this was a case of primary synovial chondrosarcoma grade II, with invasion of the femoral head, neck, and acetabulum. The bone involvement was felt to be secondary as the lesion was centered within the joint with superficial involvement of the osseous margins, as is noted in other joint-centric processes. A primary CHS of the bone with secondary extension into the joint would be expected to be centered within the bone of origin with involvement of both articular margins unlikely.

Discussion

Synovial CHS is a rare tumor reported most frequently in the knee followed by the hip, ankle, and elbow [1, 3]. Involvement of the small joints of the hands is exceedingly rare, and there have been reports of involvement of the temporomandibular joint [2, 4].

The majority of synovial CHS are secondary occurring in areas of pre-existing synovial chondromatosis. Reports of the estimated incidence of malignant transformation to chondrosarcoma in synovial chondromatosis range from 1 to 5% [5–7]. In 1996, Anract et al. suggested that the criteria for establishing synovial chondrosarcoma arising from synovial chondromatosis should consist of histologic proof of pre-exisiting synovial chondromatosis, histologic confirmation of the diagnosis of synovial chondrosarcoma, and areas of both entities discovered side by side in the same resected specimen [8]. Campanaci et al. conducted a review of the literature in 2008 and found only 39 cases of synovial CHS of which 35 were confirmed to arise histologically from synovial chondromatosis [9]. They found another four cases that were felt to be of primary origin with no histologic evidence of co-existent synovial chondromatosis [1, 2, 10]. These primary tumors involved the knee, hip, ankle, and metacarpophalangeal joint of the thumb. A fifth possible case was reported as extraskeletal myxoid chondrosarcoma arising from the synovium of the knee, although it is not clear whether a thorough search for any concomitant areas of synovial chondromatosis was performed [11]. Of those cases where the histology was reported, 95% were of low to intermediate grade (21% grade 1 and 74% grade 2) and only 5% were considered high grade [9].

The largest series of synovial CHS was published in 1991 by Bertoni et al. who collected 10 cases from the files of the Mayo Clinic and reviewed 12 cases previously reported in the literature [1]. Similar to the other reports in the literature, 7 of the 10 cases were of intermediate grade 2. Bertoni et al. suggested that certain histological features appeared to be unique to synovial CHS and could be used to help distinguish this entity from synovial chondromatosis. These included chondrocytes arranged in sheets without cell clustering, cellular atypia, crowding of cells with round and spindle cell appearance, myxoid changes, necrosis, arrangement of cells in unicellular files or ribbons and bone marrow permeation. They made the distinction between the “pushing margins” seen in the bone erosions of synovial chondromatosis and true permeation of the trabecular bone with filling up of the marrow spaces, a feature of synovial chondrosarcoma.

Synovial chondromatosis and synovial chondrosarcoma have radically different behaviors and prognosis. Synovial chondromatosis, though it can be locally aggressive and has

a tendency to recurrence, has no metastatic potential. The usual treatment consists of removal of the cartilaginous nodules with or without synovectomy. If the extent of erosive disease is such that the integrity and function of the joint have been compromised as was originally believed to be the case with our patient, a decision might be made to perform an arthroplasty. Synovial chondrosarcoma, on the other hand, is a malignant lesion with a similar prognosis to its skeletal counterpart with a reported incidence of metastases of up to 29% [9, 12]. The treatment for this tumor in most cases involves wide or radical resection or amputation.

Because of the differences in prognosis and treatment for these two entities, attempts have been made to identify features that might aid in differentiating between them preoperatively. Many authors, however, have found significant overlap with no definitive clinical or radiologic criteria of malignancy that can be used to distinguish the two [1, 7, 12–14].

Clinically, both may present with pain, swelling, and restricted range of motion. There has been some speculation that rapid recurrence of synovial chondromatosis following surgical treatment may be a harbinger of malignant degeneration [7, 12, 15]. Though it is true that there are reports of synovial chondrosarcoma confirmed histologically only after several recurrences of synovial chondromatosis, the overall recurrence rate for synovial chondromatosis is reported to be as high as 23%, the vast majority of which are benign [7]. Some authors have suggested that rapid deterioration of the clinical course in a patient with suspected synovial chondromatosis should be regarded with suspicion [8].

The distinction between these two entities on imaging can be similarly difficult. Both can present with nodular or conglomerate intraarticular soft tissue masses with or without mineralization, which can cause osseous erosion and extend into the extraarticular soft tissues. With respect to synovial chondromatosis specifically, erosions have been noted in up to 30% of cases on radiographs and 80% of MRIs [16, 17]. The erosions with synovial chondromatosis can be quite extensive, particularly in low capacity joints such as the hip, ranging in depth from a few to 24 mm and resulting in a tapered, “apple core” appearance to the femoral neck [6, 7]. These are felt to be pressure erosions due to the pushing margins of the lesion at the bone interface [16]. Some authors have made the distinction between these pressure erosions and areas of true cortical destruction and marrow permeation, considered to be an imaging feature suggestive of malignancy [1, 7]. This finding, though helpful when seen on MRI and present in our case, does not seem to be a common feature in the cases reported to date.

Retrospectively, although there was nothing specific about the clinical presentation of our patient, the appearance on MRI should have created suspicion as to the possibility of malignancy given the finding of aggressive cortical destruction and permeation of the marrow space. The diagnosis of primary synovial chondrosarcoma, however, was made on histology following the original arthroplasty and the patient was taken back for a more extensive resection. At follow up 7 years after surgery, there has been no evidence of local or systemic recurrence. The patient is pain free and has full motion in his hip. He ambulates without a cane or other assist device. There is no evidence of loosening. The femoral strut allograft has healed well. He continues to be monitored yearly with plain radiographs of the hip and a CT scan of the chest.

In summary, we present an unusual case of primary synovial chondrosarcoma in a 45-year-old male with a 5-year history of vague hip pain. This rare malignant tumor in most cases arises from pre-existent synovial chondromatosis, and differentiating between these two entities on clinical and imaging grounds can be very difficult. Rapid deterioration of clinical course or multiple recurrences in presumed synovial chondromatosis may be clinical features that can herald malignant transformation. With respect to the imaging findings, frank cortical destruction and marrow permeation may suggest synovial chondrosarcoma. Ultimately, this difficult distinction may rest with the pathologist who can use the criteria first described by Bertoni to confirm the diagnosis and guide the treatment. Open biopsy is recommended prior to definitive surgical resection in cases with any features considered suggestive of malignancy.

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