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CASE REPORT

Chondrosarcoma arising within synovial chondromatosis of the lumbar spine

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Abstract

Synovial chondromatosis is an uncommon benign neoplasm that usually affects large appendicular joints and only rarely the spine. There are only a few small series and case reports documenting malignant transformation of synovial chondromatosis into secondary chondrosarcoma, typically within the hip in the setting of recalcitrant disease and multiple recurrences. Chondrosarcoma arising in synovial chondromatosis of the spine is exceedingly rare, with only one previously published case report involving the craniocervical junction. We present a case of chondrosarcoma arising within synovial chondromatosis of the lumbosacral spine, with the diagnosis made at the time of initial presentation. We describe the clinical, imaging, and histopathological findings and review diagnostic criteria for this difficult diagnosis.

Keywords Synovial chondromatosis · Chondrosarcoma · Spine · Paraspinal mass · Facet joint

Introduction

Synovial chondromatosis is an uncommon benign disease that usually affects large appendicular joints, most commonly the hip and knee. The disease is characterized by exuberant subsynovial hyaline cartilage nodule formation, in the absence of a significant underlying pathological condition of the joint. The nodules eventually extrude through the synovium, often detaching and calcifying. In the large appendicular joints, the radiographic findings typically consist of a joint effusion with synovitis and innumerable discrete round calcified/cartilaginous loose bodies of fairly uniform size scattered throughout the joint. They may be associated with intra-articular and juxta-articular erosions [1]. Synovial chondromatosis occurring in the spine is rare. The largest series of spinal synovial chondromatosis was recently published, describing the imaging findings in 28 cases [2]. Until that time, only 11 cases were reported in the literature [3–12]. In the combined 38 cases, just over half involved the cervical spine. The remaining cases were evenly split in incidence between the thoracic and lumbosacral spine.

Chondrosarcoma arising within synovial chondromatosis is also rare, occurring in 2.6–6.4% of cases of synovial chondromatosis [13–16]. Almost exclusively it involves the hip and knee in the setting of recalcitrant disease, with multiple episodes of recurrence and a history of multiple surgical excisions [15, 16]. To our knowledge, there has been only one other reported case of chondrosarcoma arising within synovial chondromatosis of the spine, occurring at the craniocervical junction and described as "foci of low-grade chondrosarcoma" [8], which were present at the time of initial diagnosis. However, little description was provided of the pathological findings or the criteria used to make the diagnosis. The diagnosis of chondrosarcoma arising in synovial chondromatosis is challenging, both for the radiologist and the pathologist, owing to the significant overlap in the appearance of benign synovial chondromatosis and low-grade chondrosarcoma.

We report a de novo case of chondrosarcoma arising within synovial chondromatosis of the lumbar spine at the time of initial presentation, which is extremely rare. Previous studies have shown most cases to be diagnosed in the setting of

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recurrent recalcitrant disease, with gradual histological progression from a benign to a malignant state.

Case report

A 38-year-old woman presented with a 10-year history of right-sided lower back pain, which was initially attributed to a motor vehicle accident that had occurred 14 years earlier. She had no history of malignancy. Physical examination revealed a deep-seated tender mass in the right lower lumbar paraspinal region. She had no objective neurovascular deficit in her lower extremities. Presenting lumbar (L) spine radiographs demonstrated a large, mineralized soft-tissue mass along the right posterior lower lumbar spine and upper sacrum (S) with punctate and confluent calcifications suggestive of chondroid matrix (Fig. 1). Computed tomography (CT) of the lumbar spine confirmed the right posterior paraspinal mass and demonstrated to better advantage the character of the mineralized matrix, with multiple discrete punctate calcifications compatible with chondroid matrix (Fig. 2). Most of these calcifications were small, round, and uniform in size; however, there were also scattered areas of more confluent, mass-like mineralization. Inferiorly, there was an unmineralized component of the mass located along the sacrum and adjacent ilium. The mass was in close proximity to and abutted the right L4–5 facet joint with chronic-appearing erosions along the outer cortical surfaces of the articular processes. The articular surfaces of the facet joint were preserved. Subsequent magnetic resonance imaging (MRI) showed a large, heterogeneous soft-tissue mass in the right lower lumbar paraspinal region measuring up to 9 cm in the craniocaudal dimension, abutting and closely associated with the right L4–5 facet joint (Fig. 3). The margins were irregular with the mass insinuating into neural foramina at multiple levels. The mass was predominantly intermediate in signal intensity on T1-weighted images, but contained scattered foci of low signal intensity. On T2-weighted images, the mass was lobulated and demonstrated predominantly heterogeneous hyperintense T2 signal, also with scattered punctate low-signal foci correlating with the calcifications on CT and radiographs. Gadolinium-enhanced images demonstrated areas of solid heterogeneous enhancement and scattered areas of non-enhancing myxoid change with punctate nodular peripheral enhancement, which was more marked inferiorly. The constellation of imaging findings was compatible with a cartilage tumor.

Given the imaging features of a cartilage-containing soft-tissue mass in this location, the differential diagnosis would include synovial chondromatosis arising from a facet joint. Given the size, heterogeneity, and infiltrative margins with mass-like regions of enhancement, chondrosarcoma arising in synovial chondromatosis should also be considered. Additional differential considerations would include chondrosarcoma arising in an osteochondroma where the stalk has been obliterated by the sarcoma, juxtacortical chondrosarcoma, or extraskeletal mesenchymal chondrosarcoma.

Tissue obtained from a CT-guided needle biopsy histologically showed a hyaline cartilage tumor, but the sample was insufficient to determine if it was benign or malignant. A subsequent open incisional biopsy was diagnostic of chondrosarcoma arising within synovial chondromatosis (Fig. 4). Metastatic workup, consisting of a chest CT and

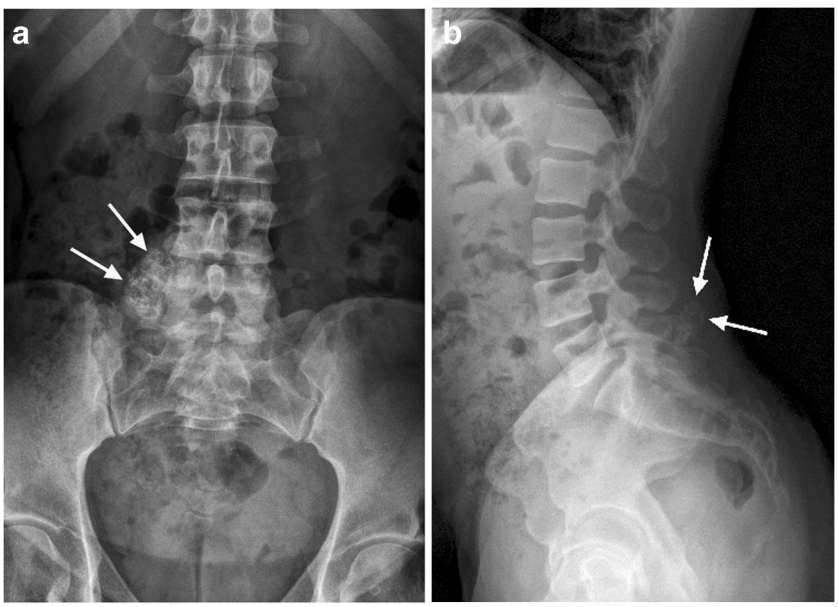

Fig. 1 A 30-year-old woman with chondrosarcoma of the lumbar spine. **a** Anterior–posterior and **b** lateral radiographs of the lumbar spine demonstrate a large, mineralized soft-tissue mass (*arrows*) along the right posterior lower lumbar spine and upper sacrum

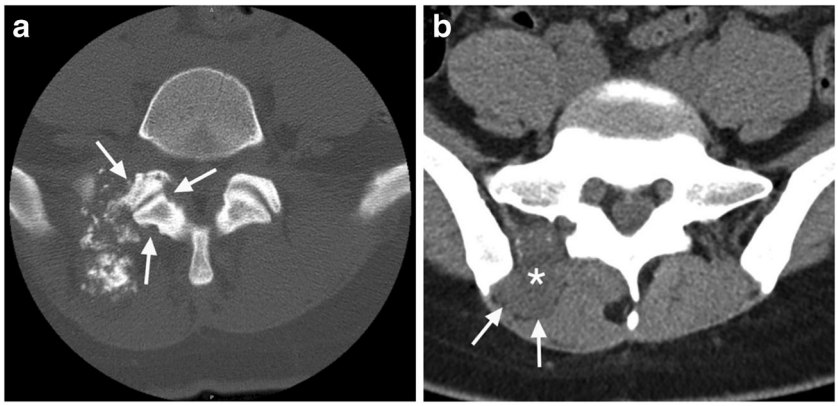

Fig. 2 **a**, **b** Axial unenhanced CT images demonstrate a right posterior paraspinal soft-tissue mass with extensive mineralized matrix and multiple discrete punctate calcifications compatible with chondroid matrix (**a**). The mass is in close proximity to and abuts the right L4–5 facet joint with chronic-appearing erosions along the outer cortical surfaces of the articular processes (*arrows* in **a**). However, the facet joint line is preserved. Inferiorly, there is an unmineralized component along the sacrum and adjacent ilium (*arrows* and *asterisk* in **a**)

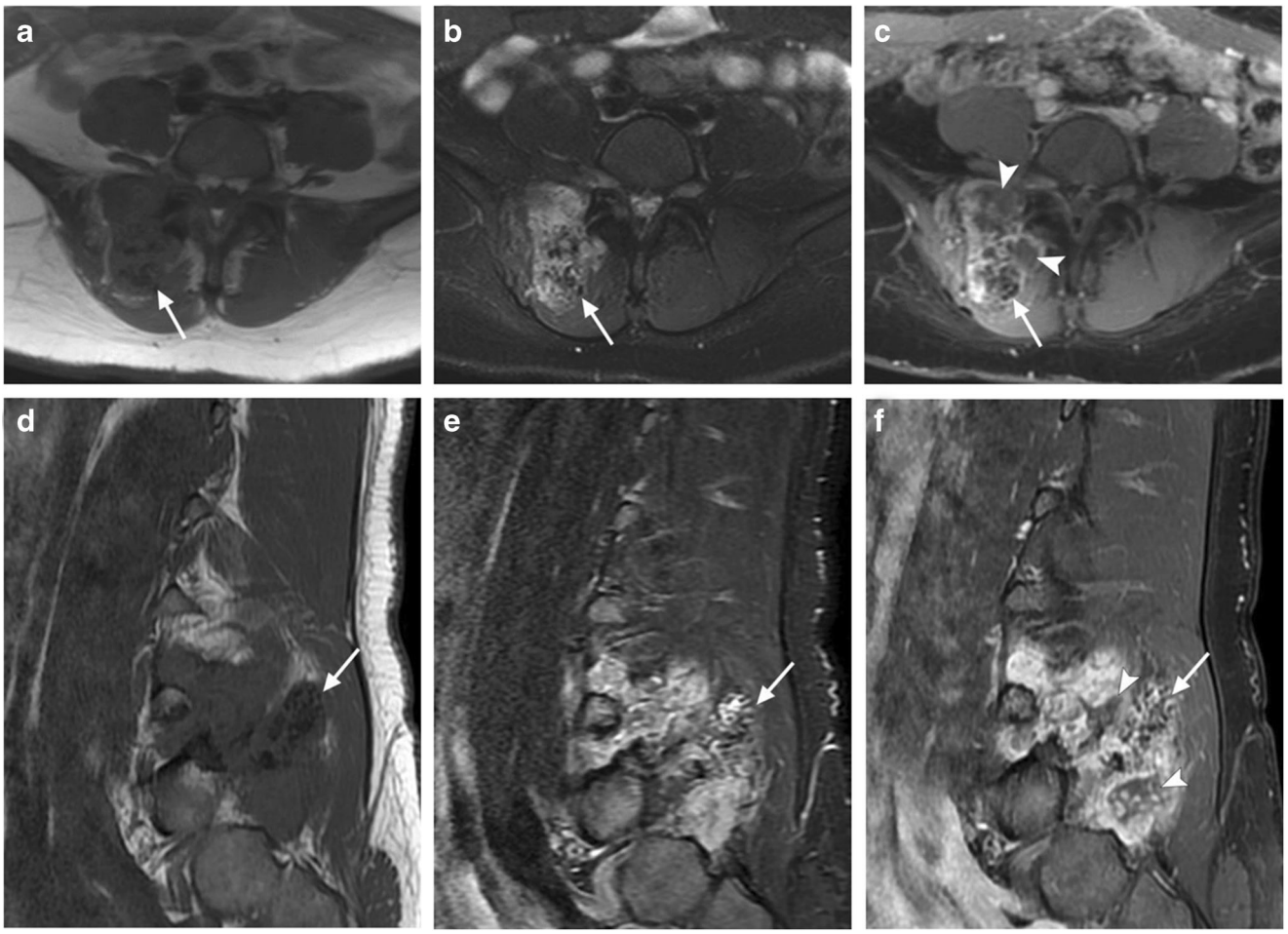

Fig. 3 **a** Axial and **d** sagittal T1-weighted, **b**, **e** short tau inversion recovery (STIR), and **c**, **f** post-contrast spoiled gradient MR images demonstrate the large size and irregular and infiltrative margins of the mass, which extends from L4 to S2 and measures nearly 9 cm in the cephalocaudal direction. The mass is heterogeneous on all sequences, but predominantly isointense to skeletal muscle on T1 and hyperintense on STIR with low signal intensity punctate foci on T1, STIR, and post-contrast images correlating with the areas of mineralized chondroid matrix noted on CT and radiographs (*arrows* in **a**–**f**). Post-contrast images demonstrate areas of solid heterogeneous enhancement and scattered areas of non-enhancing myxoid change with punctate nodular enhancement, more marked inferiorly (*arrowheads* in **c** and **f**). The constellation of findings is compatible with a cartilage tumor

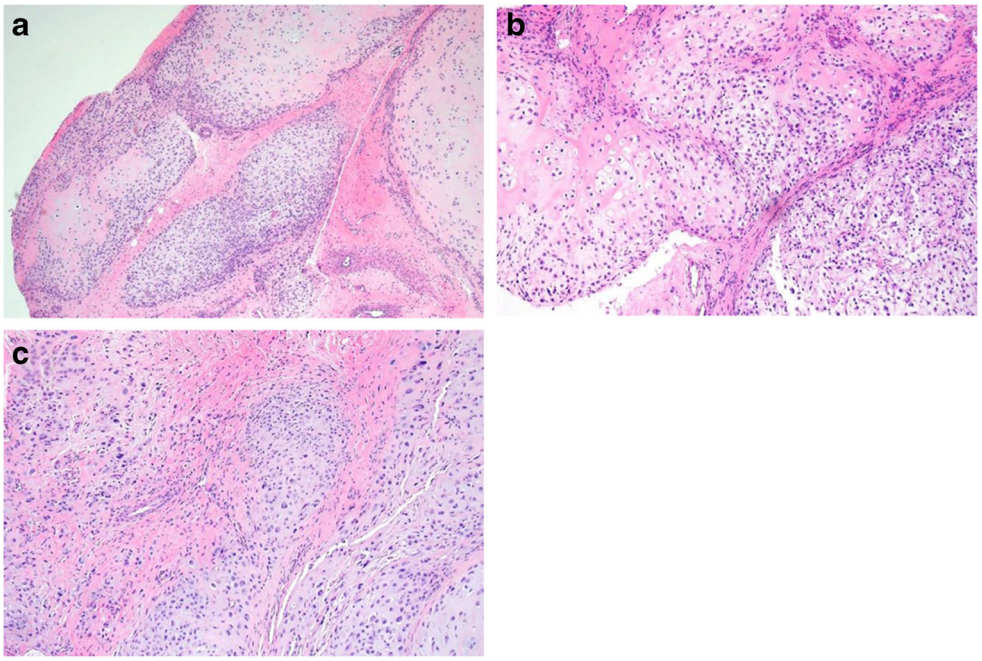

Fig. 4 **a**, **b** Photomicrographs of open incisional biopsy tissue. Some areas, particularly centrally, within the nodular masses of hyaline cartilage, resemble synovial chondromatosis, but most demonstrate diffuse sheets of cells with loss of a clustering growth pattern. **c** Malignant chondrocytes with prominent nuclear enlargement and pleomorphism permeate into surrounding soft tissue

radionuclide whole-body bone scan, was negative for evidence of metastatic disease. A staged wide resection of the tumor was performed with partial corpectomies at L4 and L5, right hemilaminectomies from L3 through S1, and posterior instrumented fusion from L2 through the sacrum and ilium (Fig. 5). Final pathology demonstrated chondrosarcoma, grade 2 (with areas of grade 3), arising in synovial chondromatosis (Fig. 6). Surgical margins were negative for tumor. At 5 years' followup, there was no evidence for residual, recurrent, or metastatic disease.

Fig. 5 Postoperative anterior–posterior lumbar spine radiograph. The patient underwent wide resection of the tumor with resection of portions of L3–S1 and posterior instrumented fusion

Discussion

Synovial chondromatosis in the spine is rare and has unique imaging features, quite distinct from the classic findings of synovial chondromatosis occurring in appendicular joints [2]. In the spine, it typically presents as an epidural soft-tissue mass with neural foraminal extension, although posterior paraspinal involvement, as in this case, is not uncommon. The classic, multiple uniform, round calcifications typically seen in synovial chondromatosis of large appendicular joints and also present in the subject of our study was noted to occur in a few patients (28%) with spinal synovial chondromatosis in a previous investigation [2]. In fact, calcifications were not a dominant finding, with nearly half showing no detectable calcifications at all. Typically, masses associated with synovial chondromatosis in the spine were reported to be close in proximity to a facet joint, but not usually centered upon the joint as was demonstrated in our case. Although chronic erosions were commonly seen on adjacent and nearby cortical surfaces, the facet joint was usually preserved, and this was true for our subject as well. In general, MRI findings were typical of a cartilage lesion. Most cases in the literature demonstrated intermediate, or intermediate and dark, T1 signal and heterogeneous T2 signal, often with discrete foci of dark T2 signal. Additionally, most cases demonstrated thin peripheral and septal enhancement with scattered nodular enhancement. Although the MRI features in this case were similar in many ways to reported findings in the series of synovial chondromatosis of the spine, the mass in our case was much more heterogeneous, irregular, and larger in size, measuring up to 9 cm, compared with the reported maximum dimension

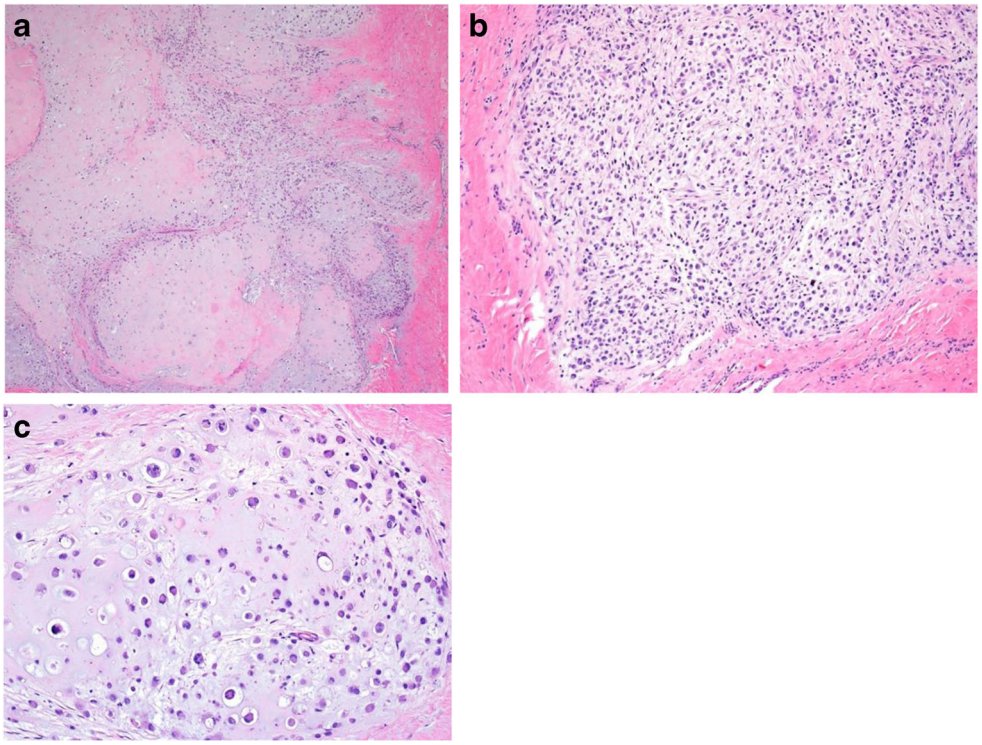

Fig. 6 Photomicrographs of the resected specimen. **a**, **b** Hyperchromatic chondrocytes coalesce into sheets at the periphery of the nodules where they extend into soft tissue. **c** Chondrocytes with marked cytological pleomorphism beyond what is seen in synovial chondromatosis

of 3.4 cm for the previously reported cases of benign synovial chondromatosis in the spine. In addition, our case of malignant transformation had areas of distinct solid and heterogeneous internal enhancement in contrast to the thin peripheral pattern of enhancement described previously [2].

In addition to synovial chondromatosis and chondrosarcomatous degeneration of synovial chondromatosis, the differential diagnosis based on imaging in this case would include primary chondrosarcoma and secondary chondrosarcoma, such as chondrosarcoma arising within an osteochondroma. However, chondrosarcomas of the spine are most commonly located in the thoracic spine and typically present as a large destructive mass involving bone and/or soft tissue, with or without characteristic cartilage matrix, and often show evidence of a predominant myxoid component. In contrast to this case, chondrosarcomas of bone, either primary or secondary, tend to show evidence of more aggressive bone destruction, usually appearing to arise from the bone, either centrally or from a surface lesion, such as a pre-existing osteochondroma [17, 18]. Although it would be difficult to exclude a juxtacortical chondrosarcoma arising from the surface of the posterior elements of the spine, or an extraskeletal mesenchymal chondrosarcoma, the close association with the facet joint, the characteristic chronic extrinsic bony erosions, and the numerous round uniform calcifications typical of synovial chondromatosis should suggest the presence of a component of synovial chondromatosis of the spine and aid in narrowing the differential diagnosis. In addition, the size, infiltrative margins within the soft tissues, and the heterogeneous enhancement pattern with areas of both solid enhancement and peripheral nodular enhancement surrounding areas with myxoid features should alert to the possibility of chondrosarcoma arising within synovial chondromatosis.

Two recent studies examined the incidence of chondrosarcoma arising in synovial chondromatosis. In a retrospective study of 78 such patients, Evans et al. [15] found malignant degeneration to chondrosarcoma in 5 cases of synovial chondromatosis (6.4%). McCarthy et al. [16] retrospectively examined 155 cases of synovial chondromatosis and found chondrosarcoma in 4 cases (2.6%). Of the 9 total cases for both series, 7 occurred in the hip, 1 in the knee, and 1 within the elbow. Finally, 7 of the 9 cases of chondrosarcoma occurred in the setting of recalcitrant disease, with a history of multiple recurrences of synovial chondromatosis.

The diagnosis of chondrosarcoma arising in synovial chondromatosis is clinically, radiographically, and histologically challenging. There are no reported reliable distinguishing imaging findings that differentiate benign synovial chondromatosis from chondrosarcoma arising in synovial chondromatosis. Furthermore, distinction between these entities is challenging, with both image-guided needle and open biopsy. According to McCarthy et al. [16] and two recent literature reviews [19, 20], clinical and imaging features suggestive of chondrosarcoma arising within synovial chondromatosis include multiple and rapid recurrences, increases in volume and extent of disease in recurrent cases, muscle infiltration, and satellite lesions remote from the surgical scar. However, in our experience, recurrences are not

uncommon in benign disease. In addition, after previous resections where tissue planes have been disrupted, it is difficult to differentiate between multifocal recurrence of benign implants due to tissue contamination, versus infiltration related to malignant transformation. An important imaging feature suggesting malignant transformation is frank cortical destruction with marrow invasion, as opposed to chronic extrinsic pressure erosions typically seen in benign synovial chondromatosis [16, 19, 20]. In our experience, loss of the uniform round calcifications and development of more bizarre mineral patterns can be predictive of malignancy as well. In the case presented here, there was an unmineralized area within the large paraspinal soft-tissue mass that lacked the uniform round calcifications present throughout much of the tumor. As previously noted, the typical uniform round calcifications typical of synovial chondromatosis in large appendicular joints, although present in this case, are often absent in synovial chondromatosis in the spine.

Histologically, synovial chondromatosis demonstrates nodules of hyaline cartilage containing clusters of chondrocytes with nuclear enlargement and hyperchromasia, nuclear crowding, binucleation, and focal minimal myxoid change. These features resemble those of chondrosarcoma, thus creating a diagnostic dilemma when trying to determine unequivocal transformation to chondrosarcoma, particularly with a limited amount of biopsy tissue. Evans et al. [15] looked at 5 cases of chondrosarcomatous degeneration among 78 cases of synovial chondromatosis. In 2 of the 5 cases, repeat biopsies suggested benign disease; however, malignancy was confirmed in these cases, but only after hindquarter amputations were performed owing to severe refractory pain in the setting of recalcitrant disease. Another pitfall of inadequate biopsy tissue occurs when foci of malignant transformation are missed and the sampled tissue represents only areas of synovial chondromatosis.

The literature lacks consistent histological criteria for the diagnosis of chondrosarcoma arising within synovial chondromatosis. In 1991, Bertoni et al. [13] published the first series of chondrosarcoma of the synovium designed with the intent of defining criteria for separating synovial chondrosarcoma from synovial chondromatosis. In their series of 12 cases, the most helpful histological features of malignancy were loss of a clustering growth pattern, myxoid change in the matrix, areas of necrosis, and spindling at the periphery of the chondroid nodules. Synovial chondromatosis may push into surrounding tissue and erode bone; however, diffuse infiltration of soft tissue and a destructive permeative growth pattern with encasement of bone trabeculae provide strong support for malignant transformation to chondrosarcoma. McCarthy and colleagues [16] required histological evidence for periarticular soft-tissue and/or an infiltrative growth pattern within bone for inclusion in their series of 4 synovial chondrosarcomas. Histological grading of cartilaginous tumor is associated with considerable interobserver variability among pathologists [21]. The challenge is even greater in the setting of synovial chondrosarcoma due to histological overlap between synovial chondromatosis and low-grade chondrosarcoma. Therefore, the diagnosis is more reliably made when the histological features fall into the category of a high-grade (2 or 3) chondrosarcoma. Most cases in the literature are reported to be grade 2 chondrosarcomas [19].

In conclusion, we present a case of high-grade chondrosarcoma arising in synovial chondromatosis of the lumbar spine. Synovial chondromatosis of the spine is unusual, and the diagnosis of malignant transformation of synovial chondromatosis is exceedingly rare. This case is unique in that malignancy was diagnosed at the time of the initial presentation compared with earlier larger studies that showed most cases of malignant transformation of synovial chondromatosis occurring in the setting of recurrent disease, often with a history of multiple recurrences, and most often involving the hip. The optimal treatment plan requires a multidisciplinary approach, with careful consideration of clinical, radiographic, and histopathological features.

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Compliance with ethical standards

Conflicts of interest The authors have no conflicts of interest to disclose.

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