Synovial Chondromatosis of Spine: Case Report and Review of the Literature

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Abstract. Primary synovial chondromatosis is an uncommon, benign entity involving the synovial lining of larger joints. In this metaplastic process, islands of chondrocytes are clustered throughout the synovium, forming nodules that jut into the joint cavity. Clinical manifestations include pain, swelling, and decreased range of motion of associated extremities. Synovial chondromatosis may be confused with other degenerative joint diseases as well as chondrosarcoma. The occurrence of synovial chondromatosis in the spine is rare. Ten cases of this spinal variant have previously been published and this report represents the eleventh case. Here we briefly review the literature and discuss the histopathologic pitfalls and differential diagnosis.

Keywords: synovial chondromatosis, chondrosarcoma, tumoral calcinosis, hamartoma

Introduction

Primary synovial chondromatosis is an uncommon, benign process in which the synovial lining of diarthrodial joints undergoes metaplasia, producing multiple discrete islands composed of chondrocytes. Osteocartilagenous nodules develop and may protrude into the joint cavity. The nodules may also be seen in tendon sheaths and extraarticular bursae [1,2]. Pieces of ectopic tissue are often dislodged and accumulate in the joint space. These loose bodies are responsible for many of the clinical symptoms. Although similar debris is observed in arthritic processes, synovial chondromatosis differs since it lacks an underlying degenerative etiology. Synovial chondromatosis typically involves one large joint, common sites being the knee, shoulder, elbow, and hip [1,4]. Microscopically, clustered chondrocytes show varying nuclear polymorphism, binucleate cells, increased mitotic activity, and prominent cellularity [1,2].

Relatively few cases of primary synovial chondromatosis in the spine have been reported. Search of the English language literature revealed ten published cases [5-13], of which five arose in cervical facet joints [9-12]. One case showed progression into underlying bone, signifying the importance of this entity in differential diagnosis [9]. Although synovial chondromatosis is generally considered to be benign, cases of chondrosarcoma arising from synovial chondromatosis have been reported [11,14]. Approximately 6% of cases may undergo malignant transformation [15]. In one instance, foci of sarcoma were found in an affected hip only after metastasis to the spine had occurred [3]. Chondrosarcoma rarely may arise in synovial chondromatosis of the spine [11,16]. Removal of loose bodies from the joint space is highly effective in alleviating symptoms, but local recurrence of synovial chondromatosis is not uncommon, occurring in approximately 17% of cases. Recurrent synovial chondromatosis is usually secondary to incomplete removal of the synovial lining [1,2,15].
Case Report

Clinical history and imaging studies. A 44-yr-old man presented with complaints of a painful neck mass that he had first noticed 6 months previously. He reported that the mass was tender and that the pain had progressively worsened to involve the right shoulder and biceps. He denied numbness or tingling but he noted weakening of his right grip. The patient’s medical history was significant for chronic renal failure following a kidney transplant 14 years previously.

Physical examination revealed a tender, fixed mass approximately the size of a grape in the cervical region on the right side of the neck. Cranial nerves two through twelve were intact, as was sensation to light and sharp touch. The finger extensors and flexors of the right hand exhibited full strength and reflexes were normal bilaterally.

Computed tomography (CT) and magnetic resonance imaging (MRI) showed a bony tumor with a large exophytic component on the right lateral aspect of the C1 and C2 vertebrae, with compression of the right vertebral artery (Fig. 1, panels A and B). At surgery, the patient underwent a transcondylar far lateral approach to the posterior cranial fossa with C2 laminectomy, right facet-ectomy, decompression and mobilization of the right vertebral artery, en bloc resection of the tumor, C1 to C4 posterior instrumentation, and fusion with an allograft (Fig. 1, panel C).

Intraoperatively, following the initial exposure and before the laminectomy was performed, an avascular, yellow, nodular mass was identified that had fleshy and granular areas. The tumor was carefully dissected and >90% was resected. The vertebral artery had been skeletonized at that level and was decompressed by removal of the tumor. This was followed by appropriate instrumentation to stabilize the spine.

Histopathologic findings. Grossly, the pathology specimen consisted of fragments of grey-white soft tissue with multiple gray-white irregular nodules that ranged in size from approximately 1 to 20 mm in largest dimension. The tissue was fleshy with areas of granularity. Routinely processed, paraffin-embedded, H&E-stained microscopic sections revealed discrete clusters of hyaline cartilage and calcification within areas of proliferative synovium and fibrous tissue (Fig. 2, top panel). There were multiple isolated foci of dystrophic calcification. The surrounding synovium was reactive (Fig. 2, middle panel) with a rim of multinucleated giant cells encasing the islands. At high magnification, chondrocytes within the myxoid cartilaginous matrix were occasionally plump and pleomorphic (Fig. 2, bottom panel). Although some cells in the cartilaginous areas were histopathologically worrisome, the tumor was considered benign.

Discussion

Diagnosing synovial chondromatosis on frozen sections can be extremely difficult given the non-diagnostic findings of fibrous and synovial tissue, foci of dystrophic calcification, and reactive histiocytes, particularly when examined in isolation. When viewed in aggregate with the clinical and radiographic information, synovial chondromatosis is likely to be diagnosed correctly. However, it may be easily mistaken for various benign and malignant entities such as tumoral calcinosis, hamartoma, degenerative joint disease, extraskeletal chondroma, and chondrosarcoma.
Given our patient’s history of renal transplant, tumoral calcinosis was high on the list of differential diagnoses, especially since tumoral calcinosis closely resembles synovial chondromatosis histologically. Tumoral calcinosis is not an intra- or peri-articular process and it is manifest in the soft tissue [15]. Grossly, tumoral calcinosis is typically a single, multilocular cystic mass. Microscopically, histiocytes surrounding calcified areas along with fibrosis and granulation tissue are seen. This reactive process is also observed in synovial chondromatosis, but in addition there is extensive synovial proliferation. Tumoral calcinosis lacks this finding as well as the presence of cartilage. Synovial chondromatosis also tends to be nodular and not cystic.

In degenerative joint disease, osteocartilaginous loose bodies that are produced grossly resemble those in synovial chondromatosis [4]. The former, however, lack chondrocyte clustering and nuclear polymorphism. The prominent nuclear polymorphism, along with nesting and hypercellularity, that is observed in synovial chondromatosis warrants the inclusion of chondrosarcoma in the differential diagnosis. To distinguish between the two, the joint must be examined. If the clusters of chondrocytes (otherwise known as an “island pattern”) are present within the confines of the joint, a diagnosis of chondrosarcoma may be excluded.

Another entity that should be included in the differential diagnosis is extraskeletal chondroma. Although it may be observed on tendons or tendon sheaths, this benign cartilaginous mass arises primarily in the soft tissue of the hands and feet, without involvement of bone [1]. Soft-part chondromas are not seen in the synovium of larger joints. Like synovial chondromatosis, hamartomas are characterized by islets of cartilage [4]. However, multiple cysts and giant cell proliferation are also present. Maturation of cartilage into bone is also a common feature of hamartomas.

A search of the English language literature identified ten previous reports of synovial chondromatosis of the spine [5-13]. A review of the topic was published by Fechner and Mills [17]. The present case is similar in most respects to the prior descriptions (Table 1). The mean age of patients at presentation is 41 yr (range 22-68 yr) and there is

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**Fig. 2.** H&E-stained sections of the cervical tumor. Top panel: At low magnification (40x), the nodular arrangement is evident, containing islands of hyaline cartilage and calcification with surrounding synovium. Middle panel: The synovium and fibrous tissue are proliferative with scattered multinucleated giant cells that rim the cartilage clusters. Focal dystrophic calcification is also evident (100x). Bottom panel: At high magnification (200x), the cartilage is seen to contain chondrocytes with pleomorphic nuclei.
no sex predilection. Most cases involve the upper spine with the cervical region being the most frequent site. The lesions tend to develop laterally and often involve the facet joints.

Synovial chondromatosis is an uncommon condition, especially when it involves the vertebral column. The present case report highlights the important features of this entity and the key differential diagnoses to be considered when examining such a patient. Correlation of the clinical, radiologic and histopathologic findings greatly facilitates arrival at the correct diagnosis.

References