Odyssey of an Elbow Synovial Chondromatosis

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abstract

Synovial chondromatosis of the elbow is an uncommon condition. However, a chondrosarcoma arising from the former is remarkably rare. The authors report a case of an elbow chondrosarcoma secondary to synovial chondromatosis in a 38-year-old woman. Before the development of chondrosarcoma, the patient underwent 3 operations and 3 sessions of radiosynovectomy because of continuous recurrence of synovial chondromatosis on the left elbow. After the last radiosynovectomy, magnetic resonance imaging and biopsy showed a grade II chondrosarcoma secondary to synovial chondromatosis. The patient underwent further surgery and custom-made arthroplasty because of aseptic loosening of the prosthesis. Four months after the last intervention, 3 subcutaneous nodes appeared on the patient’s elbow and were histologically found to be a recurrence of chondrosarcoma (grade III). Amputation by disarticulation of the shoulder was performed in addition to biopsy of another subcutaneous node on the abdomen. The biopsy showed metastasis of chondrosarcoma. At final follow-up, the patient had lung metastasis 7 years after the initial diagnosis. A reason for the manifestation of primary synovial chondromatosis and its progression to chondrosarcoma has not been found. Synovial chondromatosis progressing to chondrosarcoma in the elbow was reported in only 1 case, with no clear initial diagnosis. The role of radiosynovectomy in the development of chondrosarcoma is unknown, and no reports have described the treatment of elbow synovial chondromatosis. Although synovial chondromatosis is benign, its metaplastic nature is a marker of possible malignancy, especially with signs of recurrence and aggression. The role of radiosynovectomy in malignant changes should be examined in future studies. [Orthopedics. 2015; 38(1):e62-e67.]

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Synovial chondromatosis of the elbow is an uncommon condition. In this disorder, cartilaginous or osteocartilaginous loose bodies are produced in the joint space without evidence of underlying injury to the cartilage or synovium.\textsuperscript{1} It is considered a benign metaplastic proliferative state of the synovium that can affect fibroblasts in synovial joints, tendons, and bursa.\textsuperscript{2}

Chondrosarcoma arising from benign disease is exceedingly rare. There is no well-established reason for the manifestation of primary synovial chondromatosis and its development in a chondrosarcoma.\textsuperscript{3} The question has been asked whether synovium undergoes a metaplastic change that leads to neoplastic disease or if chondrosarcoma is the primary lesion but is misdiagnosed in its early stages.\textsuperscript{4,5} Only 1 case of elbow chondrosarcoma arising from synovial chondromatosis was published in the English literature by Bertoni et al\textsuperscript{4} in 1991; however, in this case, there was no clear initial diagnosis.

The current report describes a rare case of elbow chondrosarcoma secondary to primary synovial chondromatosis that was confirmed from the early stages with magnetic resonance imaging (MRI) and histologic examination. After several sessions of synovectomy and radiation therapy, the chondromatosis underwent malignant transformation. The patient underwent attempted surgical excision, had disarticulation of the shoulder, and experienced progression with extraskeletal metastatic lesions. At final follow-up, the patient had lung metastasis 7 years after the initial diagnosis.

**Case Report**

A 38-year-old woman presented to the orthopedic upper-limb clinic of the authors’ hospital in June 2006 with worsening symptoms of pain, swelling, and restriction of range of motion of the left elbow (range of motion, 15°–120°; normal pronation-supination), all of which had begun 8 months earlier. The patient had no other symptoms or health issues, had no allergies, and took no medication. Initial radiographs showed signs of synovial chondromatosis, and MRI showed synovial chondromatosis (Figure 1). The patient...
The patient’s symptoms were not alleviated, and the disease persisted. An MRI in March 2007 showed recurrence, and in June 2007 open synovectomy was performed. Because of continuing symptoms, 1 month later, the patient decided to seek another type of treatment and underwent 3 sessions of radiosynovectomy with rhenium-186 sulphur colloid at a dosage starting at 74 MBq and increasing to 111 MBq (74, 90, and 111 MBq). The mean amount of radiation absorbed by the synovium was 130 Gy. Pain and swelling of the elbow were not reduced, and MRI was obtained in January 2008. Increased synovial inflammation, with formation of multiple cysts around the joint, was found. Another open synovectomy was performed in February 2008. For almost 11 months, the patient showed mild improvement but was not symptom-free. The patient was almost pain-free for the first 3 months but then had a gradual increase in pain. By the end of the 11th postoperative month, she had the same level of pain that she had before the last operation. Pain was continuous and moderate to severe. Likewise, range of motion was deteriorating, and all gains in extension and flexion after the last operation (extension-flexion, 5°-125°) were lost (extension-flexion, 20°-100°) 11 months postoperatively.

An MRI obtained in July 2009 (Figure 3) showed synovial chondromatosis, and no destruction of the joint cartilage or bone was reported. Open synovectomy was performed in September 2009 (the third attempt at an open synovectomy), and the resected tissues were sent for histologic evaluation. The results showed a grade II chondrosarcoma secondary to synovial chondromatosis (Figure 4).

The patient was informed about this rare occurrence. The results of tumor staging procedures (bone scan and computed tomography scan of the lungs, abdomen, and pelvis) showed localization of cancer around the elbow joint and no signs of metastasis. The patient elected to undergo resection of the mass. Therefore, total elbow arthroplasty was performed with a custom-made prosthesis in July 2010 (Stanmore, Elstree, United Kingdom) (Figure 5). During surgery, all nerves were identified and protected, but the extensor mech-
anism of the elbow was affected because of resection of the olecranon and part of the triceps tendon. The tumor was located at the distal third of the humerus and the most proximal fourth of the radius and ulna. Therefore, resection of bone and soft tissue was performed carefully to surround the chondrosarcoma within normal bone and soft tissue. In November 2011, the patient had pain during active elbow flexion and was unable to use her hand in daily activities. Physical examination showed pain during active flexion and during resistance against internal and external rotation of the arm (when resistance was placed distal to the elbow, with the elbow at 90°). No soft tissue swelling or erythema was present, white blood cell and C-reactive protein counts were within normal limits, and radiographs showed radiolucent lines around the humeral component. Open biopsy of the humerus was performed in the same month, and tissue samples around the humeral stem were obtained for microbial culture and histologic evaluation. Findings were negative for microbial infection and tumor recurrence around the humeral component; therefore, the treating surgeon made the diagnosis of aseptic loosening.

However, because of financial constraints, the patient was unable to obtain a new prosthesis and health insurance did not cover all of the cost. The patient consented to a customized 2-stage procedure. In December 2011, the humeral component was removed and replaced with a custom-made spacer of bone cement. Afterward, the humeral stem was drilled so that it could act as a locked intramedullary nail, with a screw placed near its end. The process of investigating all options with the health insurance company and seeking alternative approaches took nearly 4 months. In April 2012, the customized cement spacer was removed and the sterilized humeral component was reinserted. Under an image intensifier, 2 screws were placed in the humeral stem, 1 successfully, in the pre-made holes. During both procedures (humeral stem removal and reinsertion), tissue was obtained for microbial culture and histologic evaluation. Findings were negative for infection or tumor recurrence.

After the last operation, the patient was initially pain-free, her arm was stable, and she could perform simple daily activities (eg, picking up food and lifting it to her mouth, holding and moving light objects) and range of motion of the implant was unrestricted. The patient was asymptomatic until the fifth postoperative month (September 2012), when she noticed 3 subcutaneous nodules, 1 in the left distal humerus and 2 in the proximal forearm, and a “hard tissue feeling” at the back of the humerus. Open biopsy showed grade III chondrosarcoma (Figure 6). At that point and after tumor staging, no further metastases were found. The patient underwent chemotherapy, which did not control the tumor. According to the oncology report, the patient received cyclo-
phosphamidem (tablets 1×2 orally on days 1-7 and days 15-21) and sirolimus (also known as rapamycin) (tablets 1 mg orally on days 1-7, 1 mg ×2 on days 8-14, and 1 mg ×3 on days 15-21).

The patient was informed of the need for radical surgical treatment. In February 2013, disarticulation from the shoulder joint was performed. During the same procedure, a subcutaneous nodule in the left middle abdominal side that had been noticed earlier was removed and sent for biopsy. The biopsy findings showed another extraskeletal recurrence. At final follow-up, 7 years after the initial operation, a left lung metastasis was found and an oncologist assumed responsibility for care of the patient.

**Discussion**

Synovial chondromatosis is an idiopathic metaplastic disorder of the synovium. Multiple nodules of metaplastic hyaline cartilage are found within the synovial membrane of a joint or are detached and act as intra-articular loose bodies. Chondrosarcoma is a malignant bone tumor that normally develops from cartilaginous tissue but can also arise de novo in extraskeletal tissue. Histologically, in synovial chondrosarcoma, the stroma shows myxoid modification and the chondrocytes show marked cytologic atypia.

The most important finding that supports the diagnosis of synovial chondrosarcoma is permeation of bone. Synovial chondrosarcoma arising from chondromatosis is 1 of the rarest types of chondrosarcoma.

The current patient represents the only reported case in the literature with histologically confirmed synovial chondrosarcoma of the elbow that demonstrates malignant change. The patient had synovial chondromatosis, but the report does not clearly state whether the finding was histologically confirmed before the first operation, which was total elbow replacement. The article also does not specify why this procedure was chosen as the initial treatment. Bertoni et al. noted the challenge associated with distinguishing synovial chondromatosis from chondrosarcoma and also the difficulty in identifying primary and secondary chondrosarcoma.

It is unknown whether synovial chondrosarcoma can develop de novo or as a metaplastic malignant change. There have been 5 reports in the literature of primary synovial chondrosarcoma without a chondromatosis stage. Muramatsu et al. presented a case of primary elbow chondrosarcoma in a 64-year-old man who underwent elbow arthroplasty after 2 years of uncontrolled right elbow pain. Preoperative MRI showed propagation of synovium inside the joint space and erosive changes on the radial head. Muramatsu et al. also produced a table with all documented cases of chondrosarcoma secondary to synovial chondromatosis. Of the 51 cases presented in the table, 17 involved the hip, 10 involved the knee, 2 involved the shoulder, 1 involved the ankle, and 1 involved the elbow. Bertoni et al. It is impossible to know the pace or pattern of development of primary synovial chondrosarcoma or its de novo existence. Because it is rare, little information is available and there have been no large cohort studies.

The diagnosis of synovial chondrosarcoma should not be based on MRI findings alone but should be confirmed by histologic examination. According to published studies, there are 3 stages of synovial chondrosarcoma, increasing in aggressiveness from grade I to grade III. In the current case, the tumor progressed from stage II to stage III. It is possible that the change in stage indicated an acceleration of growth and metastatic potential. It is impossible to determine whether radiosynovectomy contributed to the malignant change in chondromatosis or the aggressiveness of chondrosarcoma. To the authors’ knowledge, there is no published report of radiosynovectomy in a patient with synovial chondromatosis of the elbow and there are no reports so far that show growth of chondrosarcoma with this technique in patients with chondromatosis or synovitis.

The ability of chondrosarcoma to endure, recur, and produce extraskeletal subcutaneous lesions was shown in the international literature. The avascularity that characterizes this tumor shows that the neoplastic cells are almost independent of the extrinsic blood supply. Subcutaneous involvement has been reported. In 1979, Malek et al. described a solitary subcutaneous metastasis in a patient with chondrosarcoma 7 years after tumor excision. In 1995, Damron et al. reported a subcutaneous lesion on the ipsilateral forearm of a patient with previously resected metacarpal chondrosarcoma. No other metastatic growths were found.

When chondrosarcoma is found, the therapeutic plan should be radical and amputation above the affected joint should be discussed with the patient. Chondrosarcoma of the elbow has a poor prognosis. In the series of Bertoni et al., lung metastases occurred in 5 of 10 patients. In a review of the literature on synovial chondrosarcomas and case report by Muramatsu et al., 11 of 35 (31.4%) patients had metastasis and 8 (22.8%) died of disease. The numbers would probably be higher with longer follow-up because of the apparent slow progress of the tumor. Late diagnosis and inadequate first treatment occurred in many cases. In the current case, the patient’s young
age, high activity level, and unwillingness to undergo radical surgery were the factors that prevented the surgeon from performing an amputation when chondrosarcoma was diagnosed. These were also the reasons for performing custom-made total elbow arthroplasty and for finding a solution when aseptic loosening occurred as a result of using the same components. The reason for early implant failure in this case is unknown. The authors’ hypothesis is that 2 causes could contribute to aseptic loosening of the humeral component. One factor might be the patient’s young age and possible overuse of the arm, and the other could be a low-grade infection, although no infection was detected in any tissue sample obtained during surgery.

Although this condition is rare, the orthopedic surgeon should always be alert when patients have persistent recurrent pain of the elbow, even when MRI shows benign disease. As with all tumors, definitive early diagnosis and radical excision are the keys to survival. The metaplastic nature of synovial chondromatosis, even though it is benign, is a marker of possible malignancy, especially if findings indicate recurrence and aggression. Further studies are needed to examine the role of radio-synovectomy in patients with these malignant changes.

REFERENCES