Extensive tenosynovial chondromatosis of the wrist: a case report

Mikiko Shimomura, Mitsuru Nemoto, Kazuno Moriyama, Natsuko Kounoike, Akira Takeda

Department of Plastic and Aesthetic Surgery, Kitasato University School of Medicine

We report a case of a 60-year-old male patient with extensive tenosynovial chondromatosis of the wrist arising from the extra-articular tissues. Tenosynovial chondromatosis is known to recur postoperatively with a high frequency because complete synovectomy is difficult. We estimated the area of extensive tenosynovial chondromatosis lesions using 3D computed tomography (CT), which enabled clear visualization of the calcified nodules forming the tenosynovial chondromatosis. Extensive synovectomy was performed to excise the lesions on the entire circumference of the wrist removing the nodules on the basis of the 3D-CT findings. Nevertheless, the lesions recurred postoperatively after 5 years. Complete synovectomy for extensive tenosynovial chondromatosis arising from extra-articular tissues is difficult because of the likelihood of postoperative recurrence years after the surgery, making long-term follow-up a must.

Key words: tenosynovial chondromatosis, synovectomy, wrist

Introduction

Tenosynovial chondromatosis is a proliferative cartilaginous or osteocartilaginous lesion that arises from metaplastic synovial tissue, tenosynovium, or bursal lining.1,2 Tenosynovial chondromatosis has been previously reported in the literature by various names, such as synovial chondromatosis,3 synovial chondrometaplasia,4 tenosynovial osteochondroma,5 and soft tissue chondroma,6 for which accurate pathological and clinical features have been difficult to distinguish. The rate of recurrence is largely different among proliferative cartilaginous lesions that occur in the hands, especially between tenosynovial chondromatosis, synovial chondromatosis, and soft-tissue chondroma. However, these lesions are likely to be confused. Patients should receive proper treatment based on a correct diagnosis arrived at from the information on the individual characteristics of the lesions. In the present study, we report clinical features of a patient with a rare, long-term, tenosynovial chondromatosis spreading over the entire circumference of the wrist and a 5-year postoperative recurrence.

Case report

A 60-year-old male patient, an office worker, noticed masses in his right wrist gradually growing larger over a period of 5 years and finally visited our hospital. Elastic hard subcutaneous masses protruding on both ulnar and radial sides of the right wrist were recognized. Palpable small pebble-like indurations were present on the entire circumference of the wrist (Figure 1). The patient had no pain or tenderness in the wrist, however, there was a slight limitation in range of motion. A plain radiograph revealed images of multiple small calcifications accumulated predominantly on the ulnar side of the right wrist (Figure 2). Magnetic resonance imaging (MRI) showed a mass at a specific location from the subcutaneous area appearing to infiltrate between the flexor tendons. A mass showing a high signal in T2-weighted images internally contained nodules with low signals in both T1- and T2-weighted images. There were cortical erosions of the carpal bones such as the triquetrum and hamate, which were located adjacent to the mass. These images were suggestive of tenosynovial chondromatosis. Although the patient did not want surgery at first, he later agreed to undergo surgery because the limitation in the range of motion of the right wrist had gradually advanced. The surgery was decided to be
performed after confirming the location and size of the lesion with 3D computed tomography (CT). CT images revealed subcutaneous proliferating soft tissues that accompanied small calcifications on the ulnar side of the wrist and cortical erosions of the carpal bones, such as the trapezium, trapezoid, capitiate, pisiform, and triquetrum. The 3D-CT images revealed that not only had calcified nodules amassed on both the ulnar and radial sides of the wrist, but also small calcified nodules had spread to both the volar and dorsal sides of the carpal bones (Figure 3A,B).

Surgery was performed under general anesthesia. The carpal tunnel was released through an approach from the volar side of the wrist, being careful not to damage the median nerve, and the nodules infiltrating into the carpal tunnel were excised along with the synovium-like tissues. Because a mass on the ulnar side infiltrated all the way into the inside of the hypothenar muscle layer and also infiltrated into the intercarpal spaces among the carpal bones as well, the mass was excised along with a part of the hypothenar muscles and some carpal bones (Figure 4A,B). Through an approach from the dorsal side of the

Figure 1. Macroscopic appearance of the right hand and wrist at the initial hospital visit. Elastic hard subcutaneous masses protruding on both ulnar and radial sides of the wrist were recognized. Small pebble-like indurations were palpable on the entire circumference of the wrist.

Figure 2. Plain radiograph. Multiple small calcifications were observed predominantly on the ulnar side of the wrist. Cortical erosions of the carpal bones on the ulnar side were recognized.

Figure 3. 3D-CT images. These findings were useful to confirm the localization of tenosynovial chondromatosis.

A. On the ulnar side of the wrist, small, calcified nodules with versatile shapes were recognized.

B. Small, calcified nodules spread on both the volar and dorsal sides of the carpal bones.
wrist, a mass containing nodules that were located among the extensor tendons attached to the carpal bones was excised. A mass on the radial side that infiltrated between the flexor carpi radialis tendon and the flexor digitorum superficialis tendons were also excised with complete removal of all the nodules. Histopathological findings demonstrated that synovium-like tissues covered the hyaline cartilage that proliferated in a lobular pattern in the region from immediately below the dermis to the subcutaneous tissues. Because bones with calcification and fatty marrow were observed within the proliferated hyaline cartilage, the lesion was histopathologically diagnosed as tenosynovial chondromatosis (Figure 5A,B).

Extensive synovectomy for tenosynovial chondromatosis on the entire circumference of the wrist was performed with the removal of all the nodules that were detected in the plain radiograph, MRI, and 3D-CT scans. Nevertheless, 64 months later, new, small calcifications were observed in a plain radiograph (Figure 6). Although the likelihood of recurrence was strongly suspected, the patient did not want to undergo a second surgery because he had no pain or limitations in range of motion of the wrist joint. He continues to work and there are no inhibitions with his activities of daily life.

A. There were multiple nodules spreading in a lobular pattern in the region from immediately below the dermis to the subcutaneous tissue and continued to spread from deep inside the carpal tunnel to the ulnar side of the carpal bones.

B. Macroscopic appearance of the excised mass from the ulnar side of the wrist on the right and the excised calcified nodules on the left.

A. Nodules composed of hyaline cartilage were covered by synovium-like tissues.

B. Proliferated hyaline cartilage accompanied calcification and included bone components, leading to the histopathological diagnosis of tenosynovial chondromatosis.

Figure 5. Histopathological findings (H&E stain, ×100).
Synovial chondromatosis is a proliferative cartilaginous and/or osteocartilaginous lesion, which arises from metaplastic synovial tissue, tenosynovium, or bursal lining in between and around the joints, with a predilection for the knee, hip, and elbow, and rare in the hand, and wrist. The lesions are classified by their origin into intra-articular synovial chondromatosis and extra-articular tenosynovial chondromatosis. Intra-articular synovial chondromatosis of the hand and wrist can occur in a monoarticular joint or multiple joints concurrently, and its occurrence in the wrist is more frequent in the order of the distal radioulnar joint, the metacarpophalangeal joints, and the interphalangeal joints. Extra-articular tenosynovial chondromatosis occurs frequently in the dominant hand of patients in their 40s and 50s with a near equal frequency in the male-female ratio. The predilection sites are more frequent in the order of the fingers, wrist, and palm. It is more frequent in flexor tendons than in extensor tendons.

The main symptoms of tenosynovial chondromatosis are pain, swelling, and limitation in range of motion. Some patients present with accompanying trigger finger or carpal tunnel syndrome. Because there are many asymptomatic patients, sometimes it takes time before reaching a diagnosis. In the present case, the patient had no symptoms other than swelling, and it was longer than 5 years from the onset until the surgery was performed because of the gradual advancement of limitation in range of motion of the joints in the wrist.

For the diagnostic imaging of tenosynovial chondromatosis, plain radiograph, MRI, and CT are performed. Plain radiograph reveals many small multiple calcifications. It is sometimes difficult to recognize lesions without ossification by plain radiograph, whereas MRI is useful for the detection of nodules. T1-weighted MRI images revealed a mass of tenosynovial chondromatosis as a region with intermediate signal intensity, within which nodules were visualized with low intensity. T2-weighted images showed the mass as a region with high intensity, within which nodules were visualized with low intensity. CT provided accurate visualization of calcified nodules and revealed the presence or absence of cortical erosions. In the present case, cortical erosions of the carpal bones including the trapezium, trapezoid, capitate, pisiform, and the triquetrum were recognized. 3D-CT images revealed the accumulation of calcified nodules on the ulnar and radial sides of the wrist, and the presence of small, calcified nodules on both the volar and dorsal sides of the carpal bones, as well. These findings provided significant information to determine the area and likely extent of the surgery.

In these types of cases, histopathological examination is required for a definitive diagnosis. Histopathologically the proliferation of hyaline cartilage in a lobular pattern was detected in the region from immediately below the dermis to deep within the subcutaneous tissues. The hyaline cartilage included calcifications, and bone components with fatty marrow were also seen. Although imaging findings revealed cortical erosions of the carpal bones, histopathologically the nucleoli of chondrocytes were indistinctive, and mitosis was not observed, indicating no findings suggesting malignancy. It is difficult to distinguish tenosynovial chondromatosis from synovial chondromatosis and soft-tissue chondroma only by histopathological findings. Because synovial chondromatosis shows intra-articular growth, it is possible to distinguish it from tenosynovial chondromatosis, showing extra-articular growth, by their pathogenesis. Soft-tissue chondroma is a solitary and well-circumscribed lesion that occurs in relatively young individuals from 10 – 39 years of age. Other diagnoses making up the differential should include periosteal chondroma, tumoral calcinosis, hydroxyapatite deposition disease, and hemangiomatosis. These diseases can be discriminated by their clinical courses, imaging, and histopathological findings.

There is a report that synovial chondromatosis undergoes malignant transformation to chondrosarcoma, although that finding is less common. The reported cases that underwent malignant transformation were intra-articular synovial chondromatosis occurring in the knees or elbows. To our knowledge, from experience and
extensive research in the literature, there have been no incidences or reports that extra-articular tenosynovial chondromatosis of the hands or wrists undergo malignant transformation, although the rate of postoperative recurrence is high.

The current gold-standard therapeutic procedure for tenosynovial chondromatosis is synovectomy with the excision of, if at all possible, all the multiple nodules. However, complete synovectomy of the hands and wrists is especially difficult. And unless it is complete, there is the risk of a high rate of recurrence. In the literature, the recurrence rate of proliferative cartilaginous lesions of the hands was 88% in tenosynovial chondromatosis, 36% in synovial chondromatosis, and 18% in soft-tissue chondroma. In cases of tenosynovial chondromatosis, complete synovectomy is possible when the nodules are localized in a relatively small area. However, complete synovectomy in this present case of tenosynovial chondromatosis, with the removal of all the nodules located on the entire circumference of the wrist, was exceedingly difficult.

To prevent a recurrence, it is important to obtain accurate information on the location of all the lesions prior to the surgery. CT and MRI are useful to confirm the location of the lesions. Particularly, in this case, 3D-CT was outstanding for the preoperative imaging, making it possible to clearly visualize the calcified nodules on the entire circumference of the wrist. We performed extensive synovectomy with the removal of all the nodules circumferentially on the basis of those 3D-CT images. Unfortunately, as we had suspected, images of new small calcifications were detected in the plain radiograph at the 64-month follow-up. However, the patient did not elect to undergo a second surgery because he had no pain or limitations in the range of motion in his wrist. We are carefully watching his condition with regular follow-up examinations because we still suspect there may be a recurrence of the tenosynovial chondromatosis.

There have been no reports that tenosynovial chondromatosis of the hand has ever undergone a malignant transformation. There were no histopathological malignant findings in this case, and currently the patient remains asymptomatic with no complaints of inhibitions with his work or in his activities of daily life.

**Conflicts of Interest:** None

**References**