Abstract. A 38-year-old woman presented with a very large synovial chondromatosis arising from the interphalangeal joint of the thumb. The tumor surrounded the interphalangeal joint and its size was approximately 3 cm. The range of motion was almost fused. Plain radiographs showed an extra-osseous tumor shadow with calcifications. The patient had no local recurrence at one year of follow-up after marginal resection of the tumor and synovectomy. Because this case showed no osteoarthritic changes, it can be classified as primary synovial chondromatosis with a tumor-like nature. Based on the clinical course and pathological report, our synovial chondromatosis case can be classified as Milgram stage 3. Our case report is unique in three respects: i) origin from the interphalangeal joint of the thumb, ii) a single lump of mass and iii) extremely large size. To our knowledge, a similar type of synovial chondromatosis has not been previously reported in the English literature.

Synovial chondromatosis (SC) is a rare disorder characterized by synovial metaplasia. SC can lead to the development of numerous osteocartilaginous bodies in any joint, tendon sheath or bursa. The lesion is commonly monoarticular and tends to involve major joints such as the knee and hip joints (1). In the upper extremity, SC has been described in the shoulder and elbow (2), however it rarely occurs in the digits. To our knowledge, 19 cases of osteochondromatosis have been reported to date, with 8 arising from the interphalangeal joint (3-10).

We report on a unique case of a very large SC arising from the inter-phalangeal (IP) joint of the thumb. This tumor was larger than all previously reported cases. We discuss the differential diagnosis and appropriate treatment for this very rare lesion.

Case Report

A 38-year-old woman noticed a hard tumor in the dominant right thumb. The lack of any pain or impairment of daily activities meant that she did not attend a clinic until three years later. The range of motion (ROM) of the IP joint became progressively worse and the tumor gradually became larger until she finally consulted an orthopedic clinic and was introduced to the Yamaguchi University Hospital.

Upon physical examination, the thumb showed major swelling due to a bony, hard tumor. The tumor surrounded the IP joint and the proximal phalanx and its size was approximately 3 cm. The ROM of the IP joint was almost fused at a position of 10 degrees flexion.

Plain radiographs showed an extra-osseous tumor shadow with calcifications around the IP joint and proximal phalanx (Figure 1). However, the IP joint itself appeared normal. The proximal phalanges showed scalloping signs due to compression by the hard tumor. The tumor showed a homogenous low signal in T1-weighted magnetic resonance imaging (MRI) which was enhanced heterogeneously by gadolinium. It also showed a connection to the IP joint. On T2-weighted MRI, the tumor mass showed a high signal in most areas, but with some areas showing a low signal. Pre-operative open-biopsy was performed with a strong suspicion of chondrosarcoma. Histologically, the tumor cells showed mild pleomorphism of the nuclei and mitosis. The pathological diagnosis was synovial osteochondromatosis.

Marginal tumor resection was performed through a bilateral, longitudinal incision. The whitish tumor with cartilaginous hard tissue arose from the IP joint and expanded between the flexor and extensor tendon and the proximal phalanges (Figure 2). The IP joint was opened and total synovectomy was performed. The articular cartilage did not show any signs of severe invasion. The final histopathology reported articular osteochondromatosis rather than chondrosarcoma (Figure 3).
Figure 1. Pre-operative plain radiographs showed an extra-osseous tumor shadow with calcifications around the IP joint and proximal phalanx.

Figure 2. The whitish tumor with cartilagenous hard tissue arose from the IP joint and expanded between the flexor and extensor tendon and the proximal phalanges.
The patient had no local recurrence and the ROM improved to 50 degrees in flexion and 10 degrees in extension at one year of follow-up after surgery. She showed a high degree of satisfaction and suffered no disability.

**Discussion**

SC of the hand occurs very rarely. To our knowledge, only 8 cases with SC in the IP joint of the hand have so far been documented (3-10) (Table I). All cases were females and their age ranged from 13 to 67 years. Three out of these 8 cases involved the thumb. The first report of SC arising in the thumb was by Lewis et al. in 1974 and in a 28-year-old woman (3). The development of SC may be separated into primary and secondary types (2). Primary SC has a tumor-like character and chondroid metaplasia with cytological atypia can be seen within the synovium. This type of SC has the potential to transform into malignant chondrosarcoma. Secondary SC may be associated with repetitive trauma and degenerative changes to the joint. This may result from osteoarthritis,
osteoochondral fracture or osteochondritis. Because our case showed no osteoarthritic changes of the IP joint with small fragments, it can be classified as primary SC with a tumor-like nature. The histological appearance included mild atypia.

Milgram described three stages of progression for SC (1). Stage 1 consists of active synovial disease with osteochondral bodies. Stage 2 consists of a transitional stage where there is active synovial disease with osteochondral bodies in the synovial tissue. Stage 3 consists of a dormant stage in which there are osteochondral loose bodies but no active synovial disease. Based on the clinical course and pathological report, our SC case can be classified as Milgram stage 3.

The extent of surgical treatment for primary SC remains controversial. Out of the 8 reported cases of SC arising in IP joints (3-10), 2 cases of recurrence have been documented (5, 8). Although recurrences have been noted even after synovectomy, the removal of loose bodies alone may be insufficient and hence extensive synovectomy should be performed as much as possible. Arthrodesis may be indicated only when the affected joint shows severe osteoarthritis.

Malignant transformation of SC into synovial chondrosarcoma is extremely rare, with a risk of malignant change of 5% (2). We have previously reported on a very rare malignant change of SC to chondrosarcoma in the elbow joint (2). To our knowledge, however, no cases of malignant change have been reported in the hand region.

Our case is unique in three respects, i) origin from the IP joint of the thumb, ii) a single lump of mass, rather than small bodies, and iii) extremely large size expanding into both flexor and extensor sides. To our knowledge, a similar type of SC has not been previously reported in the English literature. Although our SC case is extremely rare, clinicians should be aware of its existence so that an accurate diagnosis can be performed together with early intervention.

**References**


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