Synovial chondromatosis of the hand: Three case reports and literature review

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Synovial chondromatosis is a rare condition that is probably caused by synovial connective tissue metaplasia. It is very rare in the hand and wrist and because of its low prevalence and nonspecific symptoms, synovial chondromatosis can present diagnostic difficulties for the hand surgeon and may lead to a delay in treatment. We review the literature and report three additional cases of synovial chondromatosis in the hand.

Key Words: *Hand; Osteochondromatosis; Synovial chondromatosis*

Synovial chondromatosis is a rare condition in which multiple cartilaginous nodules develop in the synovial membrane of joints, bursae or tendon sheaths as a result of subsynovial connective tissue metaplasia (1). Although this condition usually involves large joints such as the knee, it has in very rare instances been reported in the hand and wrist. Because of its low prevalence and nonspecific symptoms, synovial chondromatosis can present diagnostic difficulties for the hand surgeon and may lead to a delay in treatment. We review the literature and report three additional cases of synovial chondromatosis in the hand encountered in the practice of the senior author (AT).

CASE PRESENTATIONS

Case 1

A 66-year-old woman who was seen in 1997 complained of increasing swelling on the dorsum of the right index finger of two years' evolution. The swelling had become increasingly painful over the previous two months, restricting the range of motion and daily activities. There was no history of trauma or overuse. Examination revealed a soft mass over the ulnar aspect of the proximal interphalangeal (PIP) joint of the right index finger, with no involvement of the skin. Radiographs of the index finger showed soft tissue swelling with some indentation on the dorsum of the proximal phalanx. Surgical excision revealed a 1.2×0.4 cm area of firm, whitish, cartilaginous tissue. Histological examination demonstrated mature hyaline cartilage surrounded by fibrous tissue and partly lined by synovium, which was consistent with synovial chondromatosis (Figure 1).

La chondromatose synoviale de la main : Trois rapports de cas et une analyse bibliographique

La chondromatose synoviale est une maladie rare probablement causée par une métaplasie des tissus conjonctifs synoviaux. Elle est très rare dans la main et sur le poignet et, en raison de sa faible prévalence et de ses symptômes non spécifiques, elle peut présenter des problèmes diagnostiques pour le chirurgien de la main et entraîner un report du traitement. Nous avons analysé la documentation scientifique et présentons trois autres cas de chondromatose synoviale de la main.

Two years later, the lesion recurred in the same location, with dull aching pain. Examination revealed a small, firm, mobile cyst on the radial aspect of the PIP joint extending along the radial, dorsal and volar sides of the joint. Radiographs showed two bony defects on the proximal aspect of the middle phalanx and the distal aspect of the proximal phalanx. Surgical re-excision revealed white mucoid and nodular soft tissue. Pathological examination once again demonstrated nodular cartilaginous tissue consistent with synovial chondromatosis.

Three years later, the nodules recurred a second time on the right index finger (Figure 2), with scalloping of the neck of the proximal phalanx (Figure 3). Multiple round lesions were removed from the joint space (Figure 4). No recurrence was observed after one year of follow-up.

Case 2

A 44-year-old man presented in 1986 with a swelling over the right third metacarpal bone near the metacarpophalangeal (MCP) joint. There was no history of pain or trauma. The mass had gradually increased in size without affecting range of motion. A bone scan showed increased activity over the joint.

A dorsal incision was made, revealing a large encapsulated soft tissue tumour under the extensor tendon measuring 2 cm in diameter. This extended across the joint into the proximal phalanx. The capsule contained multiple fragments of cartilaginous material. Pathology revealed hyaline cartilaginous tissue lined by synovium, which was consistent with synovial

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Figure 1) Case 1. Histopathology of the mass on the dorsum of the proximal phalanx. Notice the mature hyaline cartilage surrounded by fibrous tissue and partly lined by synovium (hematoxylin and eosin stain, original magnification $\times 50$)

Figure 2) Recurrent synovial chondromatosis on the dorsum of the proximal interphalangeal joint of the right index finger

Figure 3) Scalloping of the volar cortex of the neck of the proximal phalanx due to synovial chondromatosis

chondromatosis. Surgical excision of the remaining synovium through a volar approach was carried out.

Six years later, the tumour recurred over the right third MCP joint. Radiographs demonstrated extraosseous calcifications (Figure 5) and the bone scan revealed increased uptake over the same area. Surgical excision yielded hard pearly nodules (Figure 6). Histology demonstrated mature hyaline cartilage with varying degrees of ossification, which was consistent with synovial chondromatosis (Figure 7).

Two years later, the lesion recurred and was surgically resected. All fragments of extraosseous and synovial enchon-

Figure 4) Excised fragments of synovial chondromatosis

Figure 5) Extraosseous ossifications seen at the neck of the third metacarpal (synovial chondromatosis)

dromatosis tissue were removed in addition to a capsule of synovial tissue. There was no recurrence for the subsequent two years of follow-up.

Case 3

A 34-year-old woman presented with complaints of pain and swelling over her right index finger. There was no history of injury and the swelling had been gradually increasing in size over the previous eight years. The swelling was located primarily over her PIP joint, with some limitation in range of motion. Radiographs demonstrated a large soft tissue mass with

Figure 6) *Excised synovial chondromatosis*

Figure 7) *Case 2. Histopathology of the mass over the right third metacarpal bone near the metacarpophalangeal joint. Notice the nodules of hyaline cartilage showing focal ossification with calcification (hematoxylin and eosin stain, original magnification $\times 50$)*

small calcifications over the ulnar aspect of a normal PIP joint (Figure 8).

A mid-lateral incision was made. The tumour was excised down to the periosteum (Figure 9). Parts of the collateral ligament were also involved and were removed. Pathology revealed fibrous tissue containing cartilaginous areas with foci of bone with calcification (Figure 10).

Ten years after the excision there was no recurrence.

LITERATURE SEARCH

A literature search for publications discussing synovial chondromatosis of the hand or wrist was conducted through PubMed using the following search strategy: (synovial chondromatosis OR synovial osteochondromatosis OR chondromatosis OR osteochondromatosis) AND (hand OR wrist OR finger OR metacarpophalangeal OR interphalangeal OR phalan* OR carpophalangeal OR carpal OR radiocarpal OR radioulnar). Abstracts and articles in English were included. References of relevant papers were reviewed. A separate search was also conducted for general articles on the disease.

The literature search yielded 167 publications, seven of which did not have an English abstract. Fifty-seven relevant articles were identified from the remaining 160 abstracts. After reviewing their reference lists, a total of 96 papers discussing synovial chondromatosis in the hand from the years 1902 to 2000 were identified. All were case reports, which limited the

Figure 8) *Extraosseous calcification on the ulnar side of the proximal interphalangeal joint of the index finger*

Figure 9) *Intraoperative view of synovial chondromatosis*

quality of evidence available from which to draw definite conclusions.

DISCUSSION

Synovial chondromatosis refers to the development of cartilage from synovial connective tissue in which secondary calcification and ossification commonly occur. Multiple loose bodies are seen when cartilaginous foci become pedunculated and detached. Cases affecting the hand and wrist are rare, with one retrospective case series reporting a prevalence of 7.5% among 53 synovial chondromatosis cases (2) compared with 74% in the knee. One hundred two cases of synovial chondromatosis of the hand were found in the literature (Table 1).

Clinical presentation

The clinical diagnosis of synovial chondromatosis can be difficult because of its low incidence and nonspecific symptoms. The greatest number of hand lesions appear to be located in the finger and more frequently involve the flexor surfaces (3). Primary synovial chondromatosis in the hand and wrist occurs most commonly from the ages of 30 to 60 years, with both sexes being affected equally. The dominant hand seems to be

Figure 10) Case 3. Histopathology of the mass overlying the right index finger. Notice the fibrous tissue containing cartilage showing foci of bone with calcification (hematoxylin and eosin stain, original magnification $\times 5$)

involved preferentially (3). The majority of patients report no significant trauma to the affected area, and clinical evidence of infection or other inflammatory reactions of the synovium is usually lacking. Patients may present with pain, swelling, stiffness and/or limitation of motion in the affected joint due to the presence of loose bodies and an active synovium. These symptoms were demonstrated in our three cases. Examination of the joint may reveal tenderness, crepitus, effusion and palpable nodules, as well as a limited range of motion or locking. Other associated symptoms that have been reported include stenosing tenosynovitis from an osteochondroma of the tendon (4,5) and extreme pain due to a loose body over an extensor tendon (6).

The differential diagnosis includes rheumatoid arthritis, chronic infection (including tuberculosis), trauma (osteochondral fracture), osteochondritis dissecans, osteoarthritis, neuropathic arthritis and gout (7).

It is also important to differentiate the features of synovial chondromatosis from a chondrosarcoma. Clinically, the malignant lesion has a faster growth rate and is more invasive, causing more pain and swelling as it enlarges. Histologically, the malignant tumour can be differentiated by the loss of a solid matrix. Other features consistent with chondrosarcoma include bone destruction, diffuse hypercellularity with more significant nuclear hyperchromasia, and spindling of tumour cells at the periphery of the lobules (8). These were absent in all three cases presented.

Investigations

Roentgenographic changes are usually nonspecific, including the presence of a soft tissue mass, scattered calcifications or a radiopaque tumour, depending on calcification or ossification of cartilaginous nodules in the lesion (9). There have also been reports of new bone formation. This feature was demonstrated in two of our cases. All three had osteophyte formation, which may have been related to the tumour. Although the metaplastic changes of the synovium are self-limited, damage to the joint from multiple loose bodies may lead to early degenerative

TABLE 1
Published cases of synovial chondromatosis in the hand

Site	Reported cases (n)	Reference
Finger		
Interphalangeal joint	12	3,6,7,26-32
Flexor tendon	27	3-5,18,33-43
Extensor tendon	13	3,12,35,36,43-48
Other or unspecified	3	10,27,49
Metacarpophalangeal joint	14	3,5,10,43,49-55
Palm	5	1,35,49,56,57
Wrist	28	3,8,13,16,27,28,33,35,45,46,48,57-68

arthritis. There have also been reports of diagnostic imaging studies using ultrasound, computed tomography and magnetic resonance imaging (9).

Pathology

The histopathological analyses of the three cases reported in the present article demonstrate the common features of synovial chondromatosis. On gross pathological examination, synovium can be affected focally or diffusely, with multiple nodules of varying sizes concentrated near the synovial-cartilaginous junction. Occasionally, a giant solitary cartilaginous nodule is seen (10). In all of our three cases, numerous cartilaginous nodules were present intraoperatively with small foci of calcification. Microscopic examination demonstrates focal, circumscribed areas of hyaline cartilage embedded within synovial connective tissue. Certain authors require intrasynovial involvement to be present before making a diagnosis of synovial chondromatosis (6,11). This feature was present in two of our three cases. Some nodules also contain bony trabeculae and mature osteoid with bone marrow elements (termed osteochondromatosis). Cytological features include enlarged chondrocytes, hypercellularity and pleomorphic nuclei. Despite these features of aggressive growth, the natural history of the disease is one of slow and usually self-limiting progression. Milgram (8) classified the disease process into three phases: early, with active intrasynovial disease but no loose bodies; transitional, with active disease and loose bodies; and late, with multiple loose bodies but no intrasynovial disease.

Although the exact pathogenesis of the condition is unknown, it has been postulated that multipotential synovioblasts undergo multifocal metaplasia to form chondroblasts, leading to the formation of cartilaginous nodules. Cells resembling fibroblasts are arranged to form a primitive chondroid matrix, which supports the growth of nodular cartilage. This is either by proliferation of interstitial cells or by surface growth of new cells differentiating near the surface to form a matrix.

Management

It has been repeatedly observed that pain and other symptoms are relieved by the evacuation of the loose bodies, and complete excision of the lesion is commonly accepted. However,

further surgical treatment in terms of synovectomy to prevent recurrence has been controversial. Some authors (12,13) advocate the removal of loose bodies with synovectomy, while others recommend the removal of loose bodies alone (14). Two of our cases demonstrated recurrences twice each, with or without synovectomy. The lone published case series specifically involving the hand demonstrated a recurrence rate of 24% (five of 21 cases), even with synovectomy (3). A retrospective study of 13 patients with synovial chondromatosis of the knee observed a lower rate of recurrence after complete synovectomy (zero of eight with synovectomy versus three of five without) (15). This has not been confirmed in other retrospective studies (2). Because of equivocal recurrence with or without synovectomy, Maurice et al (2) recommended synovectomy only for symptomatic relief from pain and swelling, and the removal of loose bodies for mechanical symptoms. Larger controlled studies are needed to determine the most appropriate treatment, but, unfortunately, the low incidence of disease precludes randomized trials.

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Complications

Complications of synovial chondromatosis include secondary degenerative osteoarthritis and mechanical locking, with limitation of function due to loose bodies in the joint space. Nerve compression by the mass has also been reported (16). Controversy remains as to whether synovial chondromatosis undergoes malignant change to become a chondrosarcoma. While no such cases have been reported involving the hand or wrist, there have been several cases of chondrosarcomatous transformation in the knee (17-25).

CONCLUSIONS

Synovial chondromatosis of the hand is rare, usually presenting as a lesion with nonspecific clinical features. Surgical excision and histopathological examination are, thus, required for diagnosis. Management consists of complete excision and removal of loose bodies. Complete synovectomy is advocated; however, recurrence is still common. Further evidence is required to improve diagnosis and management.

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