

Benign extraosseous cartilage tumours of the hand and wrist

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Abstract

Benign extraosseous cartilage tumours of the hand and wrist comprise soft tissue chondromas, synovial chondromatosis and tenosynovial chondromatosis. These tumours can significantly affect patients as they are often painful, functionally limiting and cosmetically displeasing. Although each tumour is generally considered to be a distinct entity, they share radiological and histopathological similarities. Occasionally, all three tumours may be seen in the same patient. This is an important consideration because of the risk of recurrence that may not necessarily occur at the same anatomical site but instead extend to different sites, such as a tendon sheath and/or joint.

Keywords

Benign cartilage tumours, chondroma, synovial chondromatosis, tenosynovial chondromatosis

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Introduction

Extraosseous cartilage tumours comprise soft tissue chondromas, synovial chondromatosis and tenosynovial chondromatosis. They are three benign cartilage tumours that generally affect patients in mid-adult life. When they occur in the wrist and hand, they usually present as slowly growing masses that are sometimes painful and can result in functional impairments. Patients are often concerned about the appearance of the hand or wrist, especially when the tumour is large and located on the dorsal surface where it is likely to be obvious to others. Although each tumour is considered to be a separate entity, they share clinical, radiological and histopathological features, and occasionally, all three tumours are seen in the same patient. The characteristics and treatment of these extraosseous benign cartilage tumours will be discussed.

Soft tissue chondromas

Soft tissue chondromas, sometimes referred to as 'chondromas of soft parts', generally occur between the ages of 30 and 60 years, with both sexes equally affected (Dellon et al., 1978; Zlatkin et al., 1985). They usually involve the hands and feet, with up to 80% occurring in fingers, particularly on the flexor aspects (Dellon et al., 1978; DelSignore et al., 1990; Floyd and Troum, 1995; Lichtenstein and Goldman, 1964). They are generally painless and slow growing

masses, with most being less than 3 cm in size when the patient is first examined (Plate et al., 2003; Zlatkin et al., 1985). Soft tissue chondromas tend to be mobile, well demarcated and firm to palpation (Plate et al., 2003; Zlatkin et al., 1985). They are believed to originate from synovial tissue in tendon sheaths or joints, although tissue metaplasia has also been suggested as the aetiology (Bansal et al., 1993; Floyd and Troum, 1995; Steiner et al., 1994; Zlatkin et al., 1985). The radiographic appearance is usually an extraosseous soft tissue mass with clearly defined borders in close proximity to a joint (Zlatkin et al., 1985) (Figure 1). Calcifications within the tumour occur in 33–70% of cases, and in chronic cases the tumour can cause pressure erosion on an adjacent bone (Chung and Enzinger, 1978; Floyd and Troum, 1995; Plate et al., 2003; Zlatkin et al., 1985).

Histologically there is well differentiated, mature, hyaline cartilage arranged in lobules (Floyd and Troum, 1995; Plate et al. 2003). At the periphery of these lobules, epithelioid and multinucleated cells can be seen. Chondrocytic cells are identified in the lacunae. If numerous, this variant is called

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Figure 1. Radiographs of a patient with a soft tissue chondroma demonstrating an extraosseous mass with well demarcated boundaries near the MCP joint.

chondroblastic chondroma. Myxoid, osseous or fibrous change can also be seen in soft tissue chondromas (Plate et al., 2003; Zlatkin et al., 1985) (Figure 2). Since soft tissue chondromas essentially contain mature chondrocytes, they stain positive for S-100 protein and vimentin.

Treatment is by surgical excision, which is usually uncomplicated since the tumours are encapsulated (DelSignore et al., 1990; Floyd and Troum, 1995). Malignant transformation has not been reported, although recurrences occur in approximately 20% of cases (Chung and Enzinger, 1978; DelSignore et al., 1990; Zlatkin et al., 1985). When there is a recurrence, repeat excision is usually indicated.

Synovial chondromatosis

Synovial chondromatosis is characterized by focal metaplastic cartilage formation within the synovial membrane of a joint, bursa or tendon sheath (Campeau and Lewis, 1998; Hermann et al., 1995; Kenan et al., 1993; Lewis et al., 1974; Rompen et al., 1999). Although usually affecting large joints such as knees, hips and shoulders, synovial

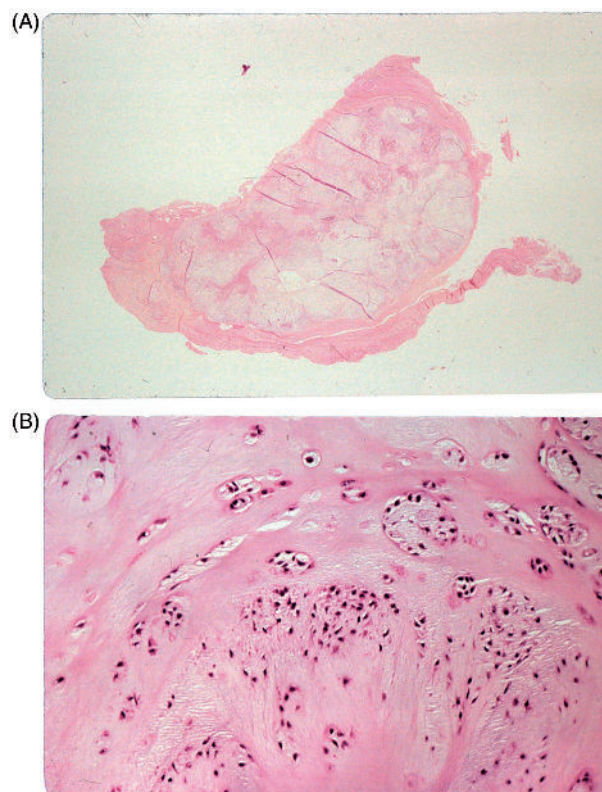


Figure 2. (a) Low and (b) high power histological sections of soft tissue chondroma (H&E stain).

chondromatosis can also develop in wrists and the small joints in hands. Patients are usually male, whereas soft tissue chondromas occur with equal frequency in both sexes. However, the age distribution of synovial chondromatosis is similar to that of soft tissue chondromas in that both conditions tend to occur in the third to fifth decades. Synovial chondromatosis is characterized by pain and joint swelling that results in a functional impairment when severe. Although the involved joint is enlarged, the chondroid nodules are usually too small to be palpated.

Radiographs often fail to demonstrate any pathology in the early stages of the disease but later will usually show radiopaque bodies of varying degrees, depending on the extent of calcification, and sometimes ossification of the cartilaginous nodules (Rogachefsky et al., 1997; Rompen et al., 1999). Although conventional radiographs provide little if any useful information in the early stages of the condition, magnetic resonance imaging (MRI) will usually show an increase in joint fluid (Rogachefsky et al., 1997; Rompen et al., 1999). CT arthrography can also be used to make the diagnosis. Later, MRI can delineate the number and size of the intra-articular bodies. Joint destruction and erosion of the underlying bone is common in chronic cases (Ono et al., 1994; von Schroeder and Axelrod, 1996).

Based on the clinical and radiographic appearance of the joint, the differential diagnosis includes pigmented villonodular synovitis, crystal deposition disease, inflammatory arthropathy and soft tissue chondrosarcoma (Rompen et al., 1999; von Schroeder and Axelrod, 1996). The definitive diagnosis is made on microscopic examination of the synovial specimen. Grossly the nodules within the synovium vary from 1 mm to several centimetres. Microscopically, the nodules display variable degrees of cellularity (Polster et al., 2005; Villacin et al., 1979) (Figure 3). Chondrocytes are plump and bi-nucleated, clustered, with rare but typical mitotic figures.

Although spontaneous regression of synovial chondromatosis has been reported, treatment is usually by surgery. Milgram (1977) classified synovial chondromatosis into three phases of development – early, transitional and late – that determined the type of surgery that was necessary. For the early phase in which there is active synovial disease but no loose joint bodies, a surgical synovectomy is recommended. For the transitional phase when there is both synovial disease and loose bodies, a synovectomy and excision of the loose bodies should be carried out. In the late phase that is 'burned out' with loose bodies but no synovial disease, removal of the loose bodies without a synovectomy is indicated. Most cases of synovial chondromatosis present in the transitional phase and the treatment is by excision of loose bodies and synovectomy. The objectives of surgery are to make a definitive histopathological diagnosis and by excision of the tumour, reduce the incidence of joint destruction. Surgery is also recommended to prevent malignant sarcomatous transformation and metastases that have been reported in

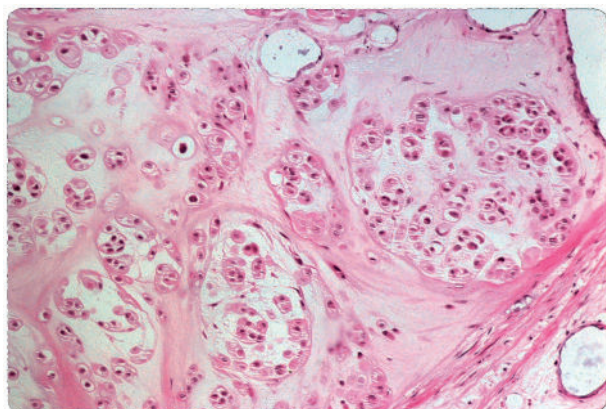


Figure 3. Specimen from a patient with synovial chondromatosis demonstrating nests of cartilaginous tissue with cytological atypia (H&E stain).

rare, aggressive forms of the tumour (Perry et al., 1988; von Schroeder and Axelrod, 1996).

In most cases of synovial chondromatosis surgery is effective, although some patients do not recover complete joint motion and some have residual pain due to damage to the articular surfaces. Recurrences have been reported in up to 60% of patients and in almost all such cases a repeat synovectomy is indicated (Ueo et al., 2004; Villacin et al., 1979).

Tenosynovial chondromatosis

Tenosynovial chondromatosis is closely related to synovial chondromatosis, with the tumour arising in the synovial lining of a tendon sheath rather than the synovial lining of a joint (Fetsch et al., 2003). Tenosynovial chondromatosis occurs almost exclusively in hands and feet (Fetsch et al., 2003; Minsinger et al., 1985). Although it can occur at any age, the condition most commonly develops in mid-adult life, similar to soft tissue chondromas and synovial chondromatosis. However, unlike patients with synovial chondromatosis, patients with tenosynovial chondromatosis have fewer symptoms. Typically, there is a soft tissue mass that develops slowly on the flexor aspect of a hand or digit which is usually non-tender and rarely causes functional impairment. Consequently, the diagnosis and treatment of tenosynovial chondromatosis is often delayed (Fetsch et al., 2003; Minsinger et al., 1985). Radiographically, the lesion may be identified by the presence of calcifications or ossifications in the extra-articular soft tissues (Figure 4).

There is a higher rate of recurrence after surgery for tenosynovial chondromatosis than synovial chondromatosis, especially for those tendon sheath tumours that have 'skip' lesions elsewhere in the sheath that are not visualized at surgery. The histopathology of the tumour is similar to synovial chondromatosis: there are multiple lobular cartilaginous nodules containing nests of chondral cells within the tenosynovium and the chondrocytes often show atypia (Fetsch et al., 2003) (Figure 5). Since flexor tendon sheaths are in intimate contact with the phalanges, any firm mass in the sheath is adjacent to a phalanx and therefore can frequently cause pressure erosion of the bone. The high rate of recurrence indicates the importance of complete excision, including any visible satellite lesions (Fetsch et al., 2003; Minsinger et al., 1985).

Case report

Similarities among the tumours were noted in 17 tumours reviewed in the Department of Pathology



Figure 4. Radiographs from a patient with tenosynovial chondromatosis showing extra-articular lesions in the soft tissues.

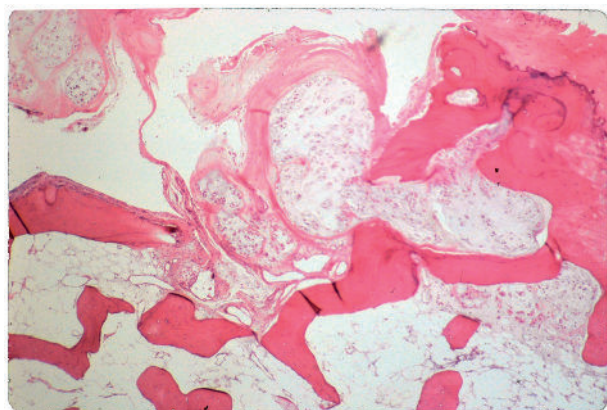


Figure 5. Histological specimen from a patient with tenosynovial chondromatosis, demonstrating bone erosion in the area of the articular end of the bone (H&E stain).

at our hospital and suggest a common pathological origin. One such example was a 28-year-old man who presented with a slightly painful, enlarging soft tissue mass of 3 months duration on the palmar-ulnar aspect of the left hand at the metacarpophalangeal (MP) joint of the little finger. The mass was considered to be a recurrence of a tumour that had been excised from that site more than 3 years earlier at another hospital. Radiographs at that first operation were reported to be normal, and hospital records

documented that a 'soft tissue chondroma' was excised from the soft tissues over the dorsum of the MP joint.

The recurrent mass was firm, multi-nodular and slightly tender on examination. Radiographs of the hand showed a radio-dense circumscribed mass between the little and ring metacarpals, and scalloping of the head/neck junction and the mid-portion of the little metacarpal (Figure 6a). The scalloping at the latter site was probably from the soft tissue mass, which was not attached to the metacarpal but was in close proximity to it and produced pressure erosion of the cortex. There was also bone formation at the base of the proximal phalanx of the little finger. At surgery, in order to obtain complete visualization of the tumour that appeared to have several components, a mid-axial longitudinal incision was made along the ulnar side of the proximal phalanx and MP joint of the little finger and which was then continued proximally along the entire length of the little finger metacarpal. Cartilaginous tumour tissue was first excised from the soft tissues adjacent to the ulnar side of the metacarpal. There was a separate circumscribed tumour mass in the soft tissues between the little and ring metacarpals that measured 5.0×3.5 cm, containing similar cartilaginous tissue that also had numerous calcifications. Figure 6b illustrates the gross specimen.



Figure 6. Case report. (a) Radiographs of the hand showed a radio-dense circumscribed mass between the little and ring finger metacarpals, and scalloping of the head/neck junction and the mid-portion of the little finger metacarpal. (b) The gross specimen demonstrating cartilaginous tissue with numerous calcifications.

Further distally, there was an additional tumour adjacent to the MP joint and proximal phalanx of the little finger. The tumour at these sites arose from the MP joint, consistent with synovial chondromatosis, and from the flexor tendon sheath of the little finger, consistent with tenosynovial chondromatosis. Microscopic sections of the tumour from the joint and tendon sheath as well as from the separate

mass between the metacarpals all showed similar findings: lobulated cartilaginous tissue with clusters of chondrocytes, and varying amounts of calcification. A thorough excision of all tumour tissue was performed and postoperatively the patient regained complete digital mobility except at the MP joint of the little finger, where flexion was limited to 50°. At a follow-up examination more than 2 years later

there was no evidence of recurrence. He was contacted by telephone more than 5 years later and he reported that he had no further problems with his left hand.

Discussion

Soft tissue chondromas, synovial chondromatosis and tenosynovial chondromatosis are benign extraosseous cartilage tumours. They are uncommon lesions but they frequently involve the hand and wrist when they do occur. They have overlapping clinical presentations and similarities in radiological appearance and histopathology. They often limit function, and may be painful and aesthetically displeasing.

The case described illustrates the common origin of benign extraosseous cartilage tumours, in that all three tumours reviewed were present. It also shows that recurrence after surgery is a significant risk. A soft tissue chondroma can recur as a distinct entity or it can recur and involve other anatomical sites such as a flexor tendon sheath and joint, as in the patient described. It is therefore important that at the initial operation a careful and thorough excision of all tumour tissue is done. Recurrences can occur even if this is done and follow-up examinations should continue for several years at least.

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Conflict of interests

None declared.

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