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Cartilage on the Move: Extra-Articular Synovial Chondromatosis (Reichel's Disease) of the Wrist - A Rare Case Report with Review of Literature

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ABSTRACT

Background: Extra-articular synovial chondromatosis (Reichel's disease) is a rare, benign metaplastic disorder of synovial tissue that produces cartilaginous, often calcified, nodules within tendon sheaths and bursae outside a joint capsule. It overwhelmingly favours the knee; involvement of the wrist is exceptional, with only a handful of cases described worldwide.
Case Presentation: A 58-year-old homemaker presented with a slow-growing swelling over the medial aspect of the left wrist of three years' duration, with pain on movement for the preceding eight months and no history of trauma. Examination revealed a firm, oval, 3.5 × 2.5 cm swelling fixed in the vertical plane and mobile transversely, with restricted wrist flexion. Radiographs showed a calcified soft-tissue swelling medial to the distal ulna without bony erosion. The lesion was excised under wrist block and was found adherent to the flexor carpi ulnaris tendon, extra-articular and extra-synovial in location. Histopathology confirmed hyaline cartilage tissue with chondrocytes and foci of bony trabeculae, consistent with extra-articular chondromatosis.
Conclusion: Extra-articular chondromatosis of the wrist is a diagnostic rarity that should be included in the differential of any peri-articular calcified soft-tissue swelling. A high index of suspicion, appropriate imaging, and complete surgical excision achieve cure and allow histopathology to exclude the principal - and rare - differential of synovial chondrosarcoma.

Keywords: *Synovial chondromatosis; Reichel's disease; extra-articular; wrist swelling; tenosynovial chondromatosis; flexor carpi ulnaris; case report*

INTRODUCTION

Synovial chondromatosis is a benign, metaplastic disorder in which nests of cartilage-forming cells arise within the synovial lining of joints, bursae, and tendon sheaths. When this metaplastic process occurs entirely outside a joint capsule -

within a tendon sheath or bursa rather than the synovium of the joint itself - the condition is termed extra-articular (or tenosynovial) synovial chondromatosis, also eponymously known as Reichel's disease, after the description of peri-articular chondroma-like nodules distinct from the more familiar intra-articular form.

The disorder is uncommon, with an estimated prevalence in the order of 1 per 100,000 population, and shows a male predominance of roughly 2:1, typically manifesting in the fourth to sixth decades of life. The knee overwhelmingly predominates as the site of involvement, accounting for the majority of reported cases, while small joints such as the wrist contribute only a small minority. Fewer than thirty cases of the extra-articular variant have been documented in the world literature to date, underscoring the rarity of this presentation. We report a case of extra-articular chondromatosis arising adjacent to the flexor carpi ulnaris tendon at the wrist in a 58-year-old woman, a location and demographic seldom described, and review the salient literature on its pathogenesis, imaging, and management.

CASE REPORT

History

A 58-year-old female homemaker presented to the surgery outpatient department with a slow-growing swelling over the medial aspect of the left wrist joint of three years' duration, associated with pain over the swelling for the preceding eight months. The swelling was insidious in onset, initially measuring approximately 0.5 × 0.5 cm, and had gradually progressed to its current size of 3.5 × 2.5 cm. The pain was aggravated by movement of the left wrist joint and relieved by rest. There was no history of preceding trauma, fever, or constitutional symptoms.

Examination

Local examination revealed a well-defined, oval swelling over the medial aspect of the left wrist joint measuring 3.5 × 2.5 cm, with an uneven surface and normal overlying skin, which was pinchable. There was no local rise in temperature, but mild tenderness was elicited over the swelling. The swelling was firm in consistency, fixed in the vertical plane, and mobile in the transverse plane, with restriction of movement noted on wrist flexion.

INVESTIGATIONS

Plain radiography of the left wrist joint demonstrated a calcified soft-tissue swelling medial to the distal end of the ulna, without evidence of underlying bony erosion or intra-articular extension, favouring an extra-articular calcified lesion over a primary intra-articular process.

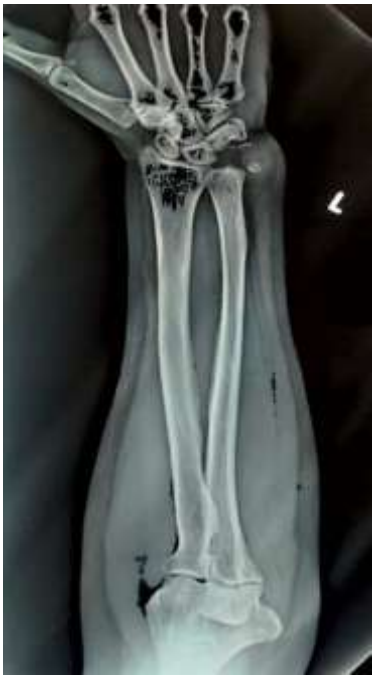

Figure 1. Plain radiograph of the left wrist showing a calcified soft-tissue swelling medial to the distal ulna, with no bony erosion.

OPERATIVE FINDINGS

After appropriate pre-operative marking (Figure 2), excision of the swelling was performed under wrist block anaesthesia. Intra-operatively, a calcified, irregularly shaped swelling measuring 4 × 3.5 cm was found densely adherent to the tendon of the flexor carpi ulnaris, confirming its extra-articular, tenosynovial location (Figure 3). The lesion was excised in toto with preservation of the underlying tendon.

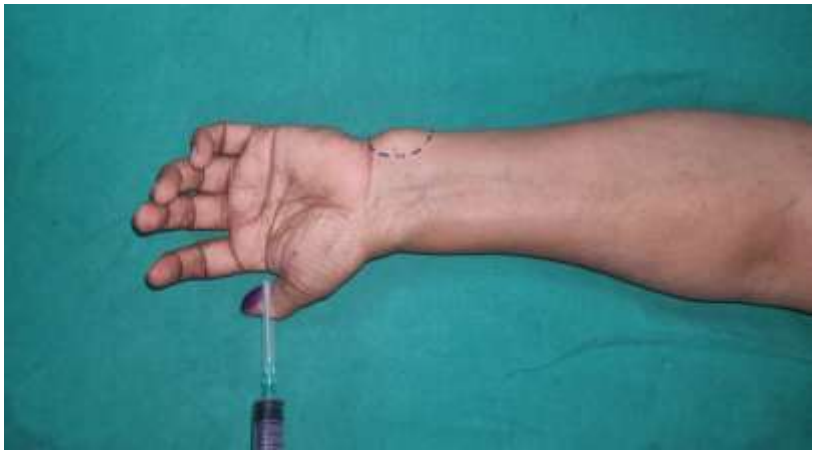

Figure 2. Pre-operative marking of the swelling over the medial aspect of the left wrist.

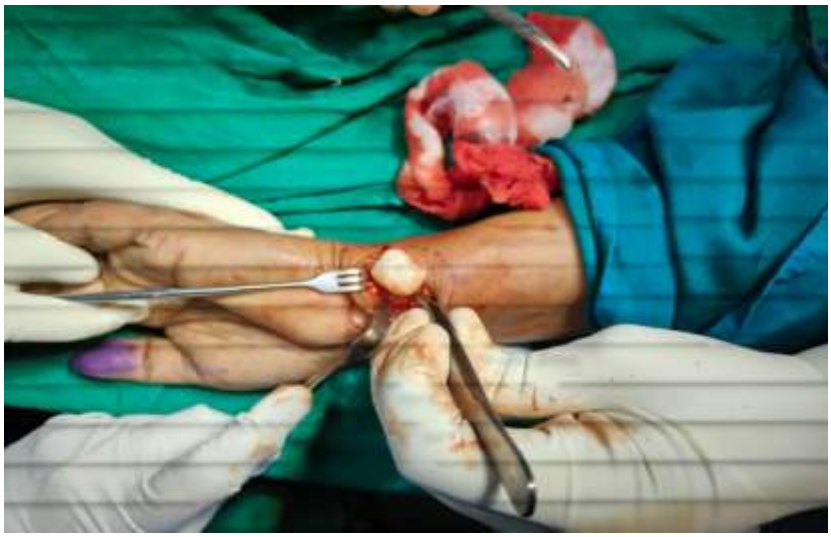

Figure 3. Intra-operative photograph showing the calcified swelling being dissected free from its adherence to the flexor carpi ulnaris tendon.

Figure 4. Gross excised specimen — a firm, irregular, calcified nodule.

HISTOPATHOLOGY

Microscopic examination of the excised specimen revealed tissue lined by a fibrous capsule, with the subepithelium showing hyaline cartilage tissue laid down with chondrocytes arranged in clusters (chondrosites), along with foci of bony trabeculae, confirming the diagnosis of extra-articular chondromatosis. No cytological atypia, increased mitotic activity, or features suggestive of malignant transformation were identified.

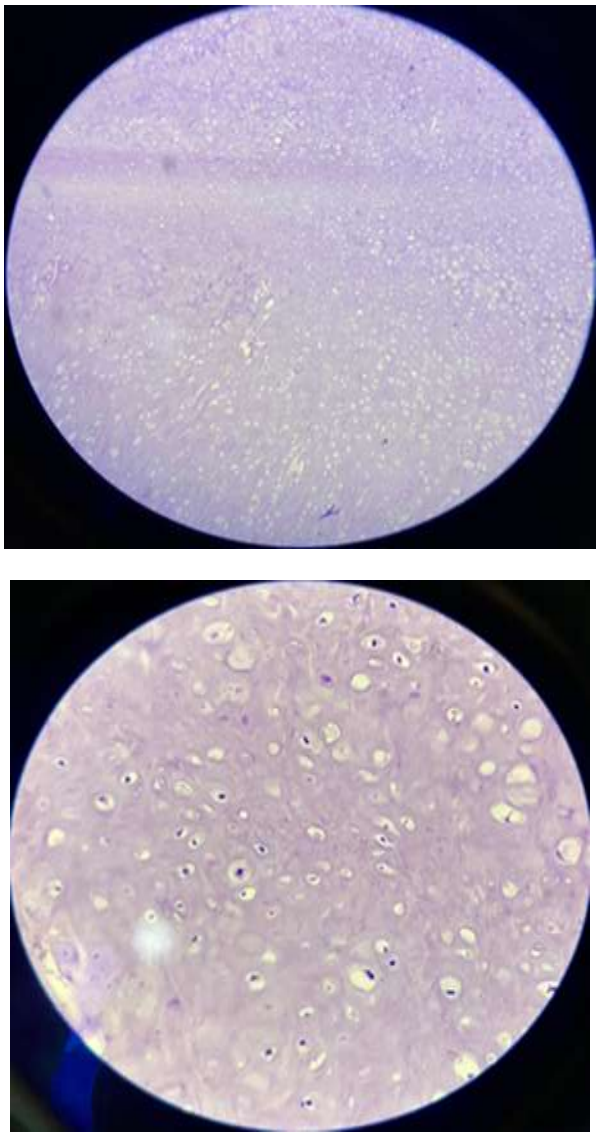

Figure 5. Photomicrograph showing hyaline cartilage nodules with chondrocytes embedded in lacunae, without atypia (H&E).

The patient had an uneventful post-operative recovery with complete resolution of pain and swelling and no early recurrence at follow-up.

DISCUSSION

Definition and nomenclature

Synovial chondromatosis is a benign proliferative disorder characterised by metaplastic transformation of subsynovial connective tissue into cartilage-forming nodules, which may calcify or ossify and can eventually detach as loose bodies. The condition is classically divided into a primary form - an idiopathic, monoarticular metaplastic process of the synovium - and a secondary form, which develops in a joint already damaged by degenerative arthritis, trauma, neuropathic arthropathy, or inflammatory arthritis, where cartilage or

bone fragments are shed into the synovium rather than arising de novo. When this metaplastic process is confined to synovial tissue lying outside the joint capsule - the lining of a tendon sheath or a bursa - the term extra-articular (or tenosynovial) synovial chondromatosis is applied; this is the entity historically eponymised as Reichel's disease, in distinction to the far more common intra-articular form.

Epidemiology

Primary synovial chondromatosis is uncommon, with a reported prevalence of roughly 1 in 100,000, a male-to-female predilection of approximately 2–4:1, and peak incidence in the third to fifth decades of life - the present patient, at 58 years, therefore lies at the older end of the usual range. The knee is by far the most frequently affected site, accounting for approximately 40–50% of reported cases in large series, followed by the hip, shoulder, elbow, and ankle; the wrist, metacarpophalangeal, and interphalangeal joints are affected only rarely. The extra-articular form is rarer still than its intra-articular counterpart, and involvement of the wrist and hand tendon sheaths - as in the present case, arising in relation to flexor carpi ulnaris - represents one of only a small number of such cases described in the literature to date.

Aetiopathogenesis

The precise aetiology of synovial chondromatosis remains uncertain. The leading hypothesis proposes metaplasia of pluripotent mesenchymal cells within the sub-synovial connective tissue into chondroblasts, which form cartilaginous nodules that may subsequently calcify or ossify and separate from the synovial surface as loose bodies capable of continued nutrition from synovial fluid. Cytogenetic studies have identified clonal chromosomal rearrangements - most consistently involving chromosome 6 and, in some series, alterations of fibroblast growth factor receptor-3 signalling - supporting a true neoplastic or metaplastic clonal process rather than a purely reactive one in at least a subset of cases. In the extra-articular form, an identical metaplastic process arises within the synovial-like lining of a tendon sheath or bursa, unrelated to any joint pathology, and - as illustrated by the present case - may become densely adherent to the adjacent tendon, here the flexor carpi ulnaris.

Milgram's classification

Milgram described three progressive histological and radiological phases of primary synovial chondromatosis. Phase I is an early, active intrasynovial disease without free loose bodies. Phase II is a transitional phase in which active intrasynovial disease coexists with loose bodies. Phase III represents multiple free osteochondral loose bodies with quiescent synovial disease. This staging correlates with imaging appearances: radiographs are most useful in Phase III, once calcification of the nodules renders them radio-opaque, whereas earlier, non-calcified phases may be radiographically occult and require ultrasonography or MRI for detection. The densely calcified lesion seen radiographically and confirmed on histology in the present case corresponds to a later, calcified phase of disease.

Clinical features

Patients typically present with an insidiously progressive, painless or mildly painful peri-articular swelling, sometimes with restriction of joint movement, stiffness, or a palpable mass, as in the present case where pain developed only after several years of painless swelling. Symptoms are often present for a protracted period - commonly several years - before diagnosis, reflecting the slow growth rate and non-specific nature of early symptoms. On examination, extra-articular lesions tend to be firm, well-circumscribed, and - as in this case - fixed along the long axis of the underlying tendon while remaining mobile transversely, a finding that can be a useful clinical clue to their tenosynovial rather than purely subcutaneous or intra-articular origin.

Imaging

Plain radiographs may show characteristic multiple ring-and-arc or popcorn-like calcifications when the nodules are calcified, as seen medial to the distal ulna in this patient, but can appear entirely normal in early, non-calcified (Phase I) disease. Ultrasonography can demonstrate a lobulated, hypoechoic soft-tissue mass with echogenic foci corresponding to calcification, and can assess its relationship to adjacent tendons. MRI is the most sensitive modality, typically showing a lobulated mass of intermediate-to-low T1 and high T2 signal, with signal voids corresponding to calcified or ossified foci, and is particularly valuable for delineating the extent of disease, its relationship to neurovascular structures, and for excluding features suspicious for malignant transformation, such as marrow invasion or aggressive bone erosion, prior to surgery.

Histopathology

The diagnostic histological hallmark is the presence of well-circumscribed hyaline cartilage nodules containing chondrocytes, which may be arranged in clusters within lacunae, embedded in a fibrous or synovial-lined stroma, with variable calcification and, as demonstrated in the present specimen, foci of bony trabeculae reflecting endochondral ossification within the nodules. Benign nodules are characterised by a relatively uniform cellularity, clustering of chondrocytes typical of normal cartilage growth, and an absence of significant cytological atypia, necrosis, or infiltrative permeative growth - features that were reassuringly absent in this case and which are essential to exclude before a benign diagnosis can be confidently rendered.

Differential diagnosis

- **Synovial chondrosarcoma:** A juxta-articular or intraosseous mass with malignant potential; distinguished from benign chondromatosis by cytological atypia, marrow or cortical invasion, permeative growth, and a more aggressive clinical course.
- **Ganglion cyst:** A common, benign, mucin-filled swelling near a joint or tendon sheath; typically, cystic and transilluminant on ultrasound, and lacks calcification on radiographs.
- **Giant cell tumour of tendon sheath:** A soft-tissue tumour of the tendon sheath that is usually solid and non-calcified, without the characteristic ring-and-arc calcification seen in chondromatosis.
- **Tumoral calcinosis / calcific tendinitis:** Periarticular calcification most often affecting the shoulder, distinguished by its typical location and lack of a discrete cartilaginous nodule on histology.
- **Osteochondroma:** A benign cartilage-capped bony outgrowth continuous with the underlying bone cortex and medulla, distinguishable radiologically by this continuity, unlike the purely soft-tissue origin of extra-articular chondromatosis.

In the present case, the absence of continuity with the underlying ulna on radiographs, the tenosynovial adherence noted intra-operatively, and the bland histopathology together excluded these mimics and supported the final diagnosis.

Management

Complete surgical excision of the lesion, with removal of all loose bodies and the affected portion of synovium or tendon sheath, is the treatment of choice and is generally curative, as was achieved in this patient by excision under regional (wrist block) anaesthesia. Where the intra-articular form is treated arthroscopically, extra-articular and tenosynovial disease - being outside the joint capsule - is more often approached through open excision, as performed here, to allow safe dissection from the adherent tendon and complete clearance of the lesion. Reported recurrence rates after synovectomy vary widely in the literature, being generally higher for tenosynovial than for purely intra-articular disease, which underscores the importance of complete

excision and continued clinical follow-up even after apparently successful surgery.

Prognosis and malignant potential

Synovial chondromatosis, whether intra- or extra-articular, follows a benign course in the great majority of patients. However, malignant transformation to synovial chondrosarcoma, though rare, is a recognised long-term complication of the primary intra-articular form, reported in the region of 1–6% of cases in large series, typically after a prolonged interval - often two decades or more - from initial diagnosis, and is heralded by multiple recurrences, worsening pain, rapid growth, or aggressive imaging features such as bone marrow invasion. Malignant transformation of the purely extra-articular, tenosynovial form is exceptionally rare and has been described chiefly in case reports rather than large series; nonetheless, meticulous histopathological assessment for atypia - as was undertaken in this case - and clinical follow-up remain essential in every patient.

Why this case merits report

- Wrist involvement in synovial chondromatosis is itself uncommon, reported in only a small fraction of published series, most of which describe the intra-articular form.
- Purely extra-articular (tenosynovial) disease adherent to the flexor carpi ulnaris, as demonstrated here, is rarer still, with fewer than thirty cases of extra-articular chondromatosis reported worldwide across all joints.
- The combination of an atypical site, an older-than-typical patient age, and radiologic-pathologic correlation adds a useful data point to a very sparse existing literature and reinforces the need to consider this diagnosis in any calcified peri-articular wrist swelling.

CONCLUSION

Extra-articular synovial chondromatosis (Reichel's disease) of the wrist is an exceedingly rare entity that should nevertheless be considered in the differential diagnosis of any slow-growing, calcified, peri-articular soft-tissue swelling, particularly one showing fixity along the axis of an adjacent tendon. Plain radiography can be diagnostic once the lesion calcifies, but cross-sectional imaging and histopathological confirmation remain essential both to secure the diagnosis and to exclude the rare but important differential of synovial chondrosarcoma. Complete surgical excision, as performed in this patient, is curative and carries an excellent prognosis, though long-term follow-up is prudent given the recognised, if rare, potential for recurrence and malignant transformation in this family of disorders.

Declaration by Authors

Ethical Approval: Not applicable (case report); written informed consent for publication was obtained from the patient.

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Conflict of Interest: No conflicts of interest declared

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