Extra-articular tenosynovial chondromatosis of the right fifth digit in a 59-year-old man:
A case report and literature review.

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ABSTRACT
Tenosynovial chondromatosis is a rare benign disorder characterized by formation of cartilaginous bodies within the synovia of the tendon sheaths. Most commonly present in the hands and feet. Clinical presentation and plain radiography can be inconclusive, which can lead to misclassification, most often confused as a chondroma of soft parts. In this case, we report the clinical, radiologic, and histology of a 59-year-old man who presented with a 1-year history of mass on the right fifth digit with limitation of motion secondary to this condition. Surgical excision revealed multiple cartilaginous nodules of varying size arising from the flexor tendon sheath. The diagnosis was confirmed postoperatively by surgical histopathology. The postoperative course of the patient was uncomplicated and has achieved an excellent functional recovery.

CASE REPORT

A 59-year-old male presented to the Hand Surgery Clinic with a complaint of a painless but irregular mass at the distal interphalangeal joint (DIP) of the right fifth finger that he had noticed since a year ago. He noted that the mass had slowly grown in the last year. The patient reported a slightly reduced range of motion in the DIP joint, however no restrictions in his daily activities. The patient recalled no history of trauma, infection, or overuse of the right hand.

On physical examination, a ~ 2 cm multinodular hard mass was palpable over the dorsum and palmar aspect of the DIP joint of the fifth finger (Fig. 1a). The range of motion of this finger was slightly reduced in flexion due to mass effect. The patient reported no tenderness or lack of sensation over the area of the lesion. The plain radiographs of the fifth finger showed multiple-calcified bodies of varying size at the entire length of the middle phalanx was encompassed by the lesion. (Fig. 1b and c). In addition, volar and dorsal osseous scalloping were noted of the subluxated middle phalanx (Fig. 2).

Routine hematological and biochemical tests were unremarkable. Since chondromatosis rarely occurs extra-articularly, the nodules were preliminarily diagnosed on imaging as a periosteal chondroma.

Surgical excisional biopsy of the nodules was performed under general anesthesia, with a tourniquet applied at the forearm. A longitudinal incision was made on the dorsal aspect of the distal phalanx after a digital block was given.
The dorsal mass was dissected on the ulnar and radial aspect and sent for surgical pathology. A small amount of bone destruction was noted on the radial aspect of the middle phalanx; however, no evidence of invasion identified.

Histology revealed osteocartilaginous nodules attached to soft tissue and benign-appearing cartilaginous cells in lacunae, (Fig. 3), consistent with a diagnosis of tenosynovial chondromatosis (TC). No signs of malignancy or infection were observed.

Complete excisional biopsy was performed one month later. An incision was performed on the palmar aspect of the right fifth digit under the digital nerve block. There were several white and brittle calcified cartilaginous soft tissue masses along the medial and lateral palmar aspect of the right small finger. These masses were densely adherent to the volar plate and flexor digitorum profundus tendon sheath and were safely resected (Fig. 4a and b). Histopathology once again confirmed the diagnosis of TC. The patient’s postoperative recovery was uneventful. No recurrence of the lesion was observed at the time of follow-up 7 months after the operation. Full range of motion was maintained without any complication on activities of daily living.

**DISCUSSION**

**Etiology & Demographics:**

Primary synovial chondromatosis is characterized histologically by formation of hyaline cartilage nodules in the subsynovial lining. Synovial chondromatosis may be classified based on the involvement of joint, tendon sheath, or bursa. Tenosynovial chondromatosis (TC) is the extra-articular form of synovial chondromatosis. Often erroneously classified as a cartilage or soft tissue tumor [1]. TC involves the tendon sheath, especially in the hands or feet [1, 2]. TC is histologically similar to synovial chondromatosis, but TC is less common and has a higher recurrence rate, reported up to 88% in contrast to 60% of the intra-articular form of this disease [1, 2, 3, 4].

TC affects the flexor tendons sheaths more frequently than the extensor [3]. To the best of our knowledge, no significant determinant factors such as trauma, infection, repetitive stress or over usage have been reported to be associated with this entity [2, 4]. According to the data in the literature, only 5 cases of TC involving the fifth digit had been reported (as seen in our patient) [4].

The demographics of TC is not well understood due to the rarity of the condition; however, several published studies provide some useful information. A review of 21 patients by Walker found a male predominance before the fifth decade, and a review of 37 patients over 20-years by Fetsch found after the fifth decade with female: male ratio of >2:1 [2, 4]. Even though most literature agrees that TC predominates around the fifth decade of life, there have been cases of TC reported in patients as young as 7 years of age [2].

**Clinical & Imaging Findings:**

The clinical diagnosis of TC is difficult due to its intermittent nature, slow disease progression and atypical clinical manifestations. Many patients are asymptomatic. However, if symptoms are present, the two most common symptoms are painless swelling over the span of several months to years and mild point tenderness over the lesion. Other symptoms could include decreased range of motion and trigger finger deformity [5].

The plain radiographs of extra-articular TC are usually nonspecific, including the existence of calcifications, soft tissue mass, or ossification in the cartilaginous nodules. Additional findings include scalloping of the underlying osseous structures and new bone formation [2, 4, 6, 7].

Our case presents a patient with nodular mass in the distal fifth finger. Plain film radiographs showed multiple-calcified bodies enclosing a subluxated middle phalanx from its base to the DIP along with volar and dorsal osseous scalloping of the middle phalanx. Other findings, such as erosion of bone surface were also reported [6]. Accurate diagnosis of TC is challenging on plain film radiography alone. There is a tendency to misclassify TC with more common look alike entities such as chondromas of soft parts or periosteal chondromas [4]. Radiographic studies for a diagnosis of periosteal chondroma, often shows a single periosteal mass, frequently with internal calcifications, that sharply scallops the outer cortex of the underlying bone. Soft tissue chondroma is an extra-articular entity with a similar anatomic distribution in the hands and feet. Radiologic appearances often show a mass within the soft tissues and closely adjacent to the underlying bone [8]. However due to limited descriptions and no definite radiologic appearances, a diagnosis of periosteal chondroma was made.

Although not performed in this case, other authors have reported diagnostic imaging studies using ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI) to further evaluate tenosynovial chondromatosis. CT-scan of TC can clearly detect non-calcified loose bodies that are not visible on plain radiographs as well as cortical erosions. MRI is useful to reveal the exact locations of the nodules, and signal intensities differ according to the degree to which the nodules have mineralized. In the reported cases, the nodules show a low to intermediate signal intensity as compared to muscle on T1-weighted images (T1W), with high signal intensity on T2-weighted images (T2W). The contrast enhancement pattern of TC is characteristically of hyaline cartilage lesions of peripheral and septal pattern [2, 4]. Ultrasound is also a modality that is particularly sensitive to identify nodules affecting the tendon sheath [2, 5]. However, if there is suspicion for the presence of soft tissue or skeletal masses, plain film radiography should remain the first study of choice.

According to the review of literature, histopathology frequently demonstrates circumscribed regions of hyaline cartilage nodules adjoining or implanted within subsynovial connective tissue or tenosynovium [4, 5, 6]. Certain authors have reported that TC cartilaginous matrix is more intensely basophilic than in other entities such as chondroma of soft parts, but this is not an explicit defining characteristic [4]. In
our case, gross examination of the excised tissue demonstrated osteocartilaginous nodules attached to connective soft tissue. Histopathology revealed benign appearing cartilaginous cells in lacunae and diagnosis of TC was made.

TC is a rare condition with a differential that includes more common conditions such as chondromas, which makes the diagnosis of this entity more challenging. TC is a rare entity, making it important to present cases such as this one to contribute to the knowledge base. This study highlights the challenges for recognizing TC at a clinical level. Furthermore, this report also explores current literature and points to ways in which these diagnostic challenges can be approached through imaging, as well as histological analysis.

Treatment & Prognosis:
The treatment of choice for tenosynovial chondromatosis is surgical resection. Some authors have highlighted a nonsurgical approach with surveillance of the progression depending mainly on the patient’s preference when the signs and symptoms of swelling, reduced range of motion, and pain are tolerable. The final aim of surgery is complete excision of the loose bodies together with curettage of the bony excavation [9]. TC more frequently involves the flexor surfaces of fingers as seen in the case presented in this article [3, 5, 10]. It has been reported that predominant symptoms such as pain and mechanical deficiencies are relieved by the evacuation of the lesion [9]. Meanwhile, further surgical treatment with dissection of the surrounding synovial membrane has remained controversial. Some authors support the resection of loose bodies with synovectomy, while others recommend the resection of loose bodies alone [11].

The reported local recurrence of TC after resection is high and ranges from 24% and as high as 88% according to a cohort study of 37 cases [2, 4]. The recurrence of TC can take several months to several years. In contrast, chondroma of soft parts and periosteal chondromas have been reported to have marginally lower recurrence rates [4, 12]. To the best of our knowledge, no case reports of patients with malignant cases of TC have been reported. This difference in recurrence rates after surgery is another reason why differentiation of TC from chondroma and malignancies is clinically relevant.

Differential Diagnosis:
Tenosynovial chondromatosis presents a clinical picture of nodular lesions in hands and feet in patients with no history of trauma, repetitive mechanical stress, or over usage. The differential list is broad and listed below are a few conditions that have a similar presentation.

1. Synovial Chondromatosis
Synovial chondromatosis is the intra-articular form of TC [3]. Synovial chondromatosis is a benign neoplastic process characterized by synovial metaplasia and proliferation with resultant formation of multiple intra-articular chordal loose bodies. Plain film radiographs reveal multiple round bodies of similar size and degree of calcification are highly variable. Synovial chondromatosis predominantly appears at the knee, hip, shoulder and elbow [8, 13]. Furthermore, CT imaging of synovial chondromatosis reveals low attenuation of non-mineralized areas of synovial thickening, along with associated joint fluid and the high-water content of the hyaline cartilage neoplastic process. MRI appearances vary, the most frequent pattern is characterized as T1-weighted images intermediate to low, along with high signal intensity on T2-weighted images. On contrast administration, synovial chondromatosis demonstrate peripheral and septal enhancement, in keeping with characteristic of hyaline cartilage lesions [13]. Ultrasonography can present as a heterogeneous mass containing foci of hyperechogenicity. Histology analysis reveals lobular hyaline cartilaginous nodules surrounded within synovial lining. The nodules are frequently hypercellular, nuclear hyperchromasias and enlarged chondrocytes [8, 13].

2. Chondroma of Soft Tissue
Chondroma of soft tissue are benign tumors of cartilaginous tissue that present in a similar anatomic distribution as TC. They also have a propensity to present as a mass lesion adjacent to normal bony or cartilaginous structures. These features complicate differentiation from TC. However, the distinguishing feature of TC is that it is located in the tendon sheath, instead of adjacent to the tendon sheath (which could be the case in a soft tissue/periosteal chondroma) [2]. This level of detail can be seen through cross sectional imaging [2]. In terms of specific imaging findings, plain film radiographs and CT imaging present a well-circumscribed extraskeletal soft tissue mass, often with rings and arcs calcifications and can present with remodeling to the adjacent bone structure [14]. MR imaging demonstrates high signal intensity on T2-weighted images and is variable according to the degree of water content and significant calcification is seen [7, 14]. Chondroma of soft tissue demonstrates variable patterns of enhancement after IV administration of contrast medium [15]. Histology will demonstrate chondrocytes with mild or moderate atypia in a cartilaginous matrix [7].

3. Periosteal Chondroma
Periosteal chondroma is a benign chondroid lesion arising from the periosteum of tubular bones. Demographics and location preference could help clinical differentiation of periosteal chondroma from TC. Periosteal chondroma has higher incidence in childhood and young adulthood as supposed to people in their fifth decade of life, as it is the case for TC. In addition, periosteal chondromas most frequently involve the proximal femur, although hands and feet have also been reported [16]. Once again, TC can be differentiated from periosteal chondroma because TC is located in the tendon sheath rather than adjacent to it [2]. This can be seen through cross-sectional imaging. Radiographic studies also identify a solitary periosteal mass with matrix calcifications, cortical erosion and remodeling of the underlying bony cortex. On MRI imaging there are isointense or low signal intensity on T1-weighted images, and high signal intensity on T2-weighted images. Contrast-enhanced images show predominantly peripheral or ring-and-arc enhancement pattern, which is characteristic of chondroid tissue [17, 18]. Histopathology is characterized by small chondrocytes in cartilaginous matrices arising from the periosteum with insignificant mild focal cellular atypia [12, 19].
4. Giant cell tumor of the tendon sheath (localized tenosynovial giant cell tumor)

The localized tenosynovial giant cell tumor or Giant cell tumor of the tendon sheath is a slow growing benign lesion that arises from the synovial lined-tendon sheath. This lesion is generally present in the extremities and specially in the hands [20, 21, 22]. Radiographs may show multinodular masses that may cause pressure erosions on the underlying bone. MR images will demonstrate signal intensity similar to muscle on T1-weighted images and low signal intensity on T2-weighted images, primarily due to internal collagen bundles, and diffuse patterns of enhancement after intravenous injection of contrast material [20, 21, 23]. Histology shows a polymorphic population of cells with osteoclast-like giant cells and smaller mononuclear stromal cells composed of eccentric nuclei, and eosinophilic cytoplasm [21, 22]. It is also important to highlight there are rarely dystrophic calcifications of this condition [23].

5. Synovial sarcoma

Malignancies could share certain features with TC. Synovial sarcoma can present calcifications, but the mineralization is not chondroid like in TC [2]. In general, plain radiographs show synovial sarcomas as round to oval soft tissue lesions adjacent to the joint and calcification (usually peripheral) possible in up to 30% of cases. Bone erosion can occur, but it is usually indolent, which could lead to confusion with a benign lesion (e.g. TC) [24]. CT scan shows a heterogenous deep mass with an attenuation that is slightly lower or similar to muscle [24]. Differentiating between malignancy and TC can be facilitated by looking for non-specific features present in malignant lesions such as heterogeneity and hemorrhage. MR imaging can reveal these non-specific malignant features. Furthermore, malignancies can invade a tendon sheath, they are not located within the tendon sheath as in the case for TC. MR can provide information about this anatomical distinction between malignancies and TC [2]. More specifically, T1-weighted MR imaging of synovial sarcoma presents multilobulated and heterogeneous lesions with intensity similar or slightly higher to the intensity of muscle [24]. T2-weighted imaging shows high heterogeneity with predominant high intensity [24]. On intravenous administration of contrast medium, synovial sarcoma demonstrates a variable and often heterogeneous than homogenous enhancement pattern [24]. Histologically, synovial sarcoma can be of 3 subtypes: biphasic, monophasic and poorly differentiated. The most common type being monophasic where spindle cells with ovoid/pale nuclei predominate with mild-moderate mitotic activity. Biphasic type contains both epithelial cells forming glands and spindle cells, and poorly differentiated are generally epithelioid in morphology, but with high mitotic activity [24].

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FIGURES

**Figure 1:** A 59-year-old male with tenosynovial chondromatosis of the right fifth finger.

Findings: Preoperative clinical photograph of the palmar aspect (A) of the right-hand demonstrating nodular deformity in the middle phalanx of the fifth finger involving the DIP joint (purple arrow). Note there is sparring the PIP joint (pink arrow) of the fifth finger. A posteroanterior (B) and oblique (C) radiographs showed a multinodular calcified mass surrounding the middle phalanx and distal interphalangeal joint. DIP= Distal Interphalangeal Joint. PIP= Proximal Interphalangeal Joint.

Technique: Posteroanterior and oblique radiographs (Carestream Health DRX-1 System) at kVp 55 and 1.6 mAs.

**Figure 2:** A 59-year-old male with tenosynovial chondromatosis of the right fifth finger.

Findings: Lateral radiograph (A) and magnified lateral images (B, C and D) showed a multinodular calcified mass scalloping the palmar (red) and dorsal (yellow) aspect of the right fifth digit middle phalanx. Note there is subluxation of the middle phalanx, and juxta-marginal erosion of the distal phalanx base (arrow).

Technique: Lateral radiograph (Carestream Health DRX-1 System) at kVp 55 and 8 mAs.

*Radiology Case.* 2021 Aug; 15(8):8-17
Figure 3: A 59-year-old male with tenosynovial chondromatosis of the right fifth finger.

Findings: Photograph (A) of the resected specimens. Hematoxylin and eosin stained histology sections of osteocartilaginous nodules attached to connective soft tissue (B) and benign appearing cartilaginous cells in lacunae (C) consistent with tenosynovial chondromatosis. Original magnifications of panels: b x40; c, x100.

Figure 4: A 59-year-old male with tenosynovial chondromatosis of the right fifth finger.

Intraoperative photograph of complete excisional biopsy (A) at the palmar aspect of the fifth finger shows calcified cartilaginous soft tissue mass (as denoted by *) and magnified image (B) of the mass attached to the flexor digitorum profundus tendon sheath.
**Definition**
Extra-articular counterpart of synovial chondromatosis.

**Etiology**
- Hyaline cartilage nodules that form in the subsynovial tissue of a tendon sheath.
- The hands, feet, and wrists are most commonly affected. In the predilection of hand lesions, majority appear to be located in the finger and more frequently involve the flexor surfaces.

**Incidence**
Not clearly defined due to low incidence.

**Gender ratio**
Varies in predominance as follows:
- Males affected more than females in the third through fifth decades
- Female: Male>2:1 after the fifth decade

**Age predilection**
Peak predilection in the fifth decade of life.

**Risk factors**
Unknown. No association with trauma, infection, mechanical stress or over usage.

**Prognosis**
High local recurrence rate that ranges between 24% to 88%.

**Findings on imaging**
- Plain radiographs are usually nonspecific, showing the presence of a soft tissue mass, scattered calcifications, or a radiopaque tumor, which depends on the existence calcification or ossification of cartilaginous nodules within the lesion. Erosion of the bone surface is also a possible finding.
- Diagnostic imaging studies using ultrasound, computed tomography and magnetic resonance imaging have also been reported.
- MRIs have improved sensitivity to reveal the location and involvement of the lesion. Although signal intensity varied by case, reflecting the histological variation within the lesions that contained chondroid material, calcification, and ossification, osseous erosion, and remodeling. Peripheral and septal pattern enhancement with contrast administration.

**Histopathology**
Circumscribed regions of hyaline cartilage nodules adjoining or implanted within subsynovial connective tissue or tenosynovium.

**Diagnosis**
Histopathological analysis correlation with radiology studies.

**Treatment**
Surgical exploration and complete excision.

**Table 1:** Summary table of Tenosynovial chondromatosis.
<table>
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<tr>
<th>X-Ray</th>
<th>CT</th>
<th>MRI</th>
<th>Histology</th>
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| Tenosynovial Chondromatosis | Nonspecific. Soft tissue mass, scattered calcifications, or a radiopaque tumor. | Helpful to detect non-calciﬁed and calcified loose bodies and cortical erosions. | MRI T1W: Low to intermediate signal intensity  
MRI T2W: Hyperintense  
MRI C+: Peripheral and septal pattern |
| Circumscribed regions of hyaline cartilage nodules adjoining or implanted within subsynovial connective tissue or tenosynovium. |
| Synovial chondromatosis | Multiple round bodies of similar size and variable degree of calcifications. | Low attenuation of non-mineralized areas of synovial thickening with associated joint ﬂuid and the high-water content of the hyaline cartilage. | MRI T1W: Low to intermediate signal intensity  
MRI T2W: Hyperintense  
MRI C+: Peripheral and septal pattern |
| Lobular hyaline cartilaginous nodules surrounded within synovial lining.  
Nodules are frequently hypercellular, nuclear hyperchromasia and enlarged chondrocytes. |
| Chondroma of soft parts | • Well-circumscribed extraskeletal soft tissue mass.  
• Often with rings and arcs calcifications and possible remodeling to the adjacent bone structure. | Extraskeletal soft tissue mass, often with internal calcifications and can present with remodeling to the adjacent bone structure. | MRI T1W: Low to intermediate signal intensity  
MRI T2W: Hyperintense  
MRI C+: Variable patterns of enhancement |
| Chondrocytes with mild or moderate atypia in a cartilaginous matrix. |
| Periosteal chondromas | • Solitary periosteal mass with matrix calcifications.  
• Cortical erosion and remodeling of underlying bony cortex. | Periosteal mass with internal calcifications, cortical erosion and scalloping of the underlying osseous cortex. | MRI T1W: Low to intermediate signal intensity  
MRI T2W: Hyperintense  
MRI C+: Peripheral or ring-and-arc |
| Small chondrocytes in cartilaginous matrices arising from the periosteum with insignificant mild focal cellular atypia. |
| Giant cell tumor of the tendon sheath | • Multinodular masses that may cause pressure erosions on the underlying bone.  
• Highly rare dystrophic calcifications. | Multinodular lesion with possible pressure erosions on the underlying bone. | MRI T1W: Hypointense  
MRI T2W: Hypointense  
MRI C+: Diffuse patterns of enhancement |
| Polymorphous population of cells with osteoclast-like giant cells and smaller mononuclear stromal cells composed of eccentric nuclei, and eosinophilic cytoplasm. |
| Synovial Sarcoma | • Round to oval soft tissue lesions adjacent to the joint.  
• Calcifications are usually peripheral and present in up to 30% of cases.  
• Possible indolent bone erosions. | Heterogenous deep mass with a signal attenuation that is slightly lower or isointense compared to muscle. | MRI T1W: Isointense or slightly higher signal than muscle  
MRI T2W: High heterogeneity with a predominant hyperintensity  
MRI C+: Variable patterns of enhancement (Heterogenous>Homogenous) |
| Monophasic type (most common): spindle cells with ovoid/pale nuclei predominate with mild-moderate mitotic activity.  
Biphasic type: contains both epithelial cells forming glands and spindle cells. Poorly differentiated type: epithelioid morphology with high mitotic activity. |

Table 2: Differential diagnosis table for Tenosynovial chondromatosis.
Extra-articular tenosynovial chondromatosis of the right fifth digit in a 59-year-old man: A case report and literature review.

**ABBREVIATIONS**

CT = Computed Tomography  
DIP = Distal Interphalangeal Joint  
MRI = Magnetic Resonance Imaging  
PIP = Proximal Interphalangeal Joint  
PVNS = Pigmented villonodular synovitis  
T1W = T1-weighted images  
T2W = T2-weighted images  
TC = Tenosynovial Chondromatosis

**KEYWORDS**

Tenosynovial chondromatosis; Hand-Finger; Extra-articular; Synovial chondromatosis; Surgical excision

**ACKNOWLEDGEMENTS**

The authors thank Dr. Hunter Benvenuti, MD, Dr. Feras Yamin, MD, and Dr. Michael Christy, MD from the Department of Plastic & Reconstructive Surgery, Nassau University Medical Center for providing intraoperative photographs. Also thank Dr. Qi Tao from the Department of Pathology for providing histology images.

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