Case Report

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대한영상의학회지

# Diffuse-Type Tenosynovial Giant Cell Tumor of the Tendon Sheath in Both Wrists

## 양측 손목의 힘줄에 생긴 미만형의 건활막거대세포종

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Diffuse-type tenosynovial giant cell tumor (D-TSGCT), previously known as pigmented villonodular synovitis, is a locally aggressive neoplasm that may arise from the synovium, bursa, or tendon sheath. D-TSGCT is usually monoarticular and can be classified into intra- and extra-articular forms, the latter of which is rarer. Here, we report a case of D-TSGCT in a 64-year-old female that involved the entire flexor and extensor tendon sheaths of both wrists. We describe the ultrasonography and MRI findings, as well as review the relevant literature.

**Index terms** Giant Cell Tumor; Magnetic Resonance Imaging; Tendon Sheath

## INTRODUCTION

Diffuse-type tenosynovial giant cell tumor (D-TSGCT) is a rare benign tumor that causes the synovium, bursa, or tendon sheath to thicken and overgrow. D-TSGCT is a destructive villonodular hyperplastic lesion consisting of synovial mononuclear cells mixed with multinucleated giant cells, foam cells, siderophages, with hemosiderin deposition (1). The lesions are typically monoarticular and affects large joints (1). In the cases where D-TSGCT presents in an unusual location or with atypical appearance, the imaging diagnosis may be difficult. Here, we describe a case of extra-articular D-TSGCT of tendon sheaths in both wrist joints, which is difficult to differentiate from other inflammatory or infectious diseases.

## CASE REPORT

A 64-year-old woman presented with a 5-year history of insidious swelling in both wrist joints, accompanied with pain since 2–3 months ago (Fig. 1A). The physical exam-

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ination revealed approximately a 3 cm mass in medial and dorsal aspect of both wrist joints.

Simple radiographs of the wrist joints demonstrated soft tissue swelling without bony abnormality (Fig. 1B). Ultrasonography revealed heterogeneously hypoechoic masses with increased vascularity in multiple extensor & flexor tendon sheaths of both wrist joints (Fig. 1C). MRI of the right wrist joint (Fig. 1D, upper row) showed a diffuse infiltrative soft tissue mass-like lesion and synovial thickening in multiple extensor & flexor tendon sheaths. The lesion showed low signal intensity on T1-, T2-weighted images (T1, T2WIs) and fat-saturated T2WIs and faint contrast enhancement. And there were internal darker signal intensity foci in this lesion on both T1-, T2-and fat-saturated T2WIs. The MRI of the contralateral wrist joint (Fig. 1D, lower row) also demonstrated symmetric tenosynovitis, containing low signal intensity foci on T1-, T2- and fat-saturated T2WIs involving multiple tendon sheaths of flexor and extensor tendons. No significant joint effusion nor bone erosion was noted in both wrists. The clinician had initially suspected rheumatoid arthritis, considering the patient's age and symptom. The result of a serologic test including rheumatoid factor, anti-CCP, and FANA was negative. white blood cell count was normal and C-reactive protein was 7.73 mg/L, which is almost within the normal range.

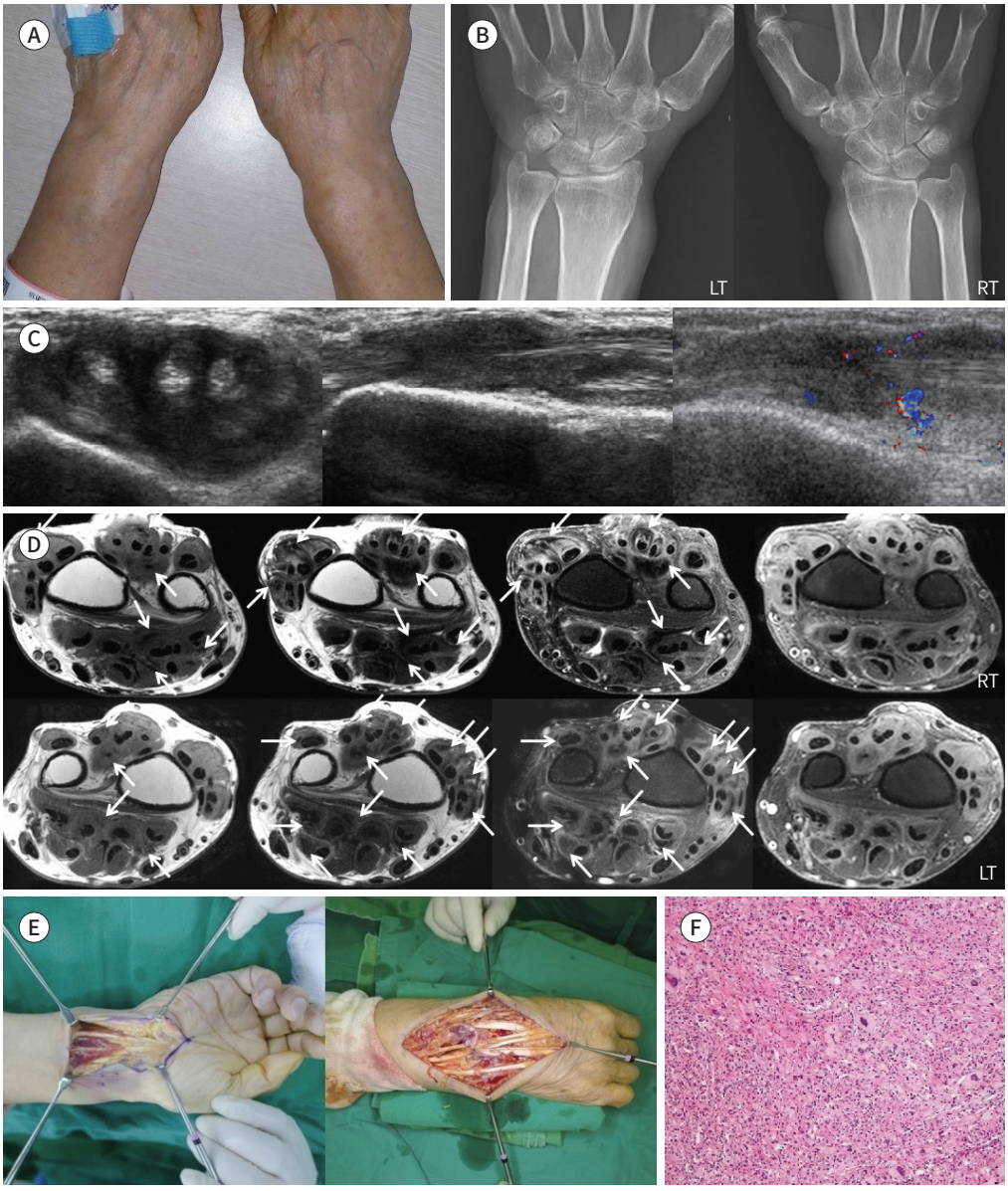
Open mass excisions were performed on both wrist joints. Operative findings showed diffuse yellowish mass-like lesions along the extensor and flexor tendon sheath of both wrists (Fig. 1E). Histologic sections revealed multiple papillary projections with diffuse infiltration of foamy histiocytes and multinucleated giant cells (Fig. 1F). The result of an excisional biopsy confirmed the diagnosis of tenosynovial giant cell tumor (previously termed as pigmented villonodular synovitis).

## DISCUSSION

TSGCT is classified into localized or diffuse by its appearance and biological behavior, and as intra-articular or extra-articular by its location (2). Localized-type TSGCT (L-TSGCT) represents the second most common soft-tissue mass of the hand and wrist, and is exceeded in frequency only by ganglion (1). On imaging, L-TSGCT is a solitary nodular mass arising from tendon sheath of volar aspect of index and long fingers (3). On the other hand, D-TSGCT is less common in the wrist, and mainly occurs in intra-articular form of the knee joint (75% of cases), followed by hip and ankle, exhibited by infiltrative mass-like lesion throughout the affected area beyond the joint and into the surrounding tissues (2). D-TSGCT of the tendon sheath as an extra-articular form is rare, and symmetric involvement of both wrists is extremely rare.

Most cases of extra-articular D-TSGCT are believed to represent extra-articular extensions of primary intra-articular disease. Extra-articular disease without intra-articular communication is the least common form of the disease-in bursa, tendon sheath or muscle and subcutaneous layer (3, 4). To our knowledge, only a few cases of extra-articular disease of tendon sheath without intra-articular involvement have been reported in the elbow and foot (3, 5). Furthermore, multifocal bilaterally symmetrical or asymmetrical D-TSGCT is also rare (1). There are few reports of intra-articular D-TSGCT involving bilateral knees, hips, shoulders, wrists, ankle and foot (6), and only one case of extra-articular D-TSGCT in both ankles. How-

Fig. 1. A 64-year-old woman with D-TSGCT of tendon sheath in both wrists.
A. Phtography shows swelling of both wrist joinsts.
B. Simple radiographs of the wrist joints demonstrate soft tissue swelling without bony abnormality.
C. US images reveal heterogeneously hypoechoic masses and increased vascularity in multiple extensor & flexor tendon sheaths of the wrist (transverse view, longitudinal view, and Doppler US from the left side).
D. MRI of the right (upper row) and left (lower row) wrists (T1WI, T2WI, fat-saturated T2WI, and contrast-enhanced T1WI from the left side). Diffuse infiltrative soft tissue mass-like lesion and synovial thickening with T1 and T2 low signal intensity and faint enhancement in the entire extensor & flexor tendon sheaths. The lesion showed internal darker signal intensity foci (arrows) on T1-, T2- and fat-saturated T2WIs.
E. Operative findings show diffuse yellowish mass-like lesions along the extensor and flexor tendon sheath of both wrists.
F. Histological sections reveals multiple papillary projections with diffuse infiltration of foamy histiocytes and multinucleated giant cells (hematoxylin and eosin, × 100).
D-TSGCT = diffuse-type tenosynovial giant cell tumor, T1WI = T1-weighted image, T2WI = T2-weighted image, US = ultrasonography



ever, they are mainly intra-artiular and temporally independent (7).

Histologically, D-TSGCT is destructive villonodular hyperplasia with synovial mononuclear cells mixed with multinucleated giant cells, foam cells, and siderophages with hemosiderin deposition (1). On MRI, the hemosiderin component of TSGCT causes preferential shortening of T2 relaxation time and yield predominant low signal intensity of the lesion on T2WI, which is a characteristic finding of the disease (1)-widely known, but important clue in our case. We could include TSGCT as a differential diagnosis in our case, due to diffuse predominant low signal intensity of the lesion on T1 and T2-weighted MRI and low echogenicity on the US, although unusual in other respects.

MRI is the optimal modality to diagnose and define the extent of intra- and extra-articular D-TSGCT (1). MRI of D-TSGCT reveals heterogeneous, diffuse, mass-like thickening of synovium, which widely infiltrates and entraps adjacent soft tissue, and occasionally accompanied with bone erosion (1, 3). Extra-articular D-TSGCT shows infiltrative mass in bursa, tendon sheath or muscle and subcutaneous layer with or without involvement of joint or bone (5). Importantly, it typically shows low signal intensity on T1, T2WIs and blooming artifact on gradient echo images due to hemosiderin, as mentioned above (1). Enhancement of PVNS is variable, but common after intravenous administration of gadolinium (1). And the majority of the lesions are monoarticular or solitary (1). Sonographic features may be nonspecific, and showed markedly thickened synovium or hypoechoic mass-like lesion with increased vascularity (5).

In our study, bilateral involvement of the disease was the challenging diagnostic feature. The following differential diagnoses should be considered in our case. The most likely differential diagnosis in this presentation is rheumatoid arthritis. Tenosynovitis is commonly seen in early rheumatoid arthritis of the wrists and hands, and usually presents bilaterally (8). Small areas of fibrous pannus can show intermediate to low signal intensity on T2WI and hypovascularity after enhancement, but majority of them are associated with synovitis and bone erosion (8). In this case, there was no evidence of synovitis, bone erosion or deformity, usually seen in chronic stage of rheumatoid arthritis, even the 5-year history of wrist swelling without medication. The next possible diagnosis is chronic infection such as nontuberculous mycobacterium (NTM). MRI findings of NTM revealed exuberant tenosynovitis, showing increased fluid within tendon sheath with surrounding soft tissue swelling, and rarely involves the underlying muscles and bony structures, with preserved joint space and without tendon tear even in the long-standing phase (9). Furthermore, the most common infection site of NTM in the musculoskeletal system is the hand and wrist (9). However, in our case, no significant fluid along the tendon sheath on T2WIs. And NTM usually has a solitary site of infection, because it is known to be acquired by direct inoculation due to trauma, surgery or corticosteroid injection (9).

In summary, we have reported a very rare case of extra-articular D-TSGCT of tendon sheath involving both wrist joints symmetrically. Although the diagnosis may be challenging especially in the case of atypical presentation, the typical characteristics of the disease-low signal intensity on T1WI/T2WI or blooming artifact on gradient echo images due to hemosiderin deposition, can be helpful to include it as one of the differential diagnoses.

## Author Contributions

Conceptualization, P.S., K.S.H., L.I.J; investigation, H.S., P.S.; project administration, P.S.; resources, all authors; supervision, P.S.; visualization, H.S., P.S., S.J.; writing—original draft, H.S.; and writing—review & editing, P.S., K.S.H., S.J., L.I.J.

## Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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# 양측 손목의 힘줄에 생긴 미만형의 건활막거대세포종

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건활막거대세포종은 이전에 색소성융모결절성 활막염으로 불리던 질환으로, 활막, 점액낭, 힘줄 등에 생기는 드문 양성 종양이다. 미만형의 건활막거대세포종은 국소형에 비해 드물고 그중에서도 관절 외에 발생하는 것은 더 드물다. 또한 대부분은 하나의 관절을 침범하기 때문에 양측을 대칭적으로 침범한 것도 매우 드물다. 이에 저자들은 64세 여자 환자에서 양측 손목의 신전건과 굴곡건을 모두 침범한 관절 외 미만형의 건활막거대세포종의 증례에 대해 초음파와 자기공명영상검사 소견을 중심으로 보고하고자 한다.

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