CASE REPORT

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Tenosynovial Giant Cell Tumor of the Ankle: A Case Report with an Unusual Location

Ayak Bileğinde Gelişen Tenosinovyal Dev Hücreli Tümör: Nadir Yerleşimli Olgu Sunumu

***Hatice KAPLANOĞLU*[1]** ***Veysel KAPLANOĞLU*[2]** ***Aynur TURAN*[1]** ***Rasime Pelin KAVAK*[1]** ***Tuba AKDAĞ*[1]**

ÖZ

Tenosinoviyal dev hücreli tümör (TSDHT), tendon kılıfından veya bursasından kaynaklanan yavaş büyüyen benign bir tümördür. Ayak ve ayak bileğinin TSDHT' ü, el ve dizle karşılaştırıldığında çok daha az sıklıkta bildirilmiştir. Morfolojik olarak sıklıkla elde görülen lokalize tip ve büyük eklemlerde görülen diffüz tip olarak sınıflandırılır. TSDHT, ayak ve ayak bileğinin yumuşak doku tümörlerinin ayırıcı tanısında düşünülmelidir. Sunulan çalışmada, nadir lokalizasyon olan ayak bileğinde yerleşimli, intra-artiküler lokalize tip, TSDHT olgusunun klinik ve radyolojik bulguları sunulmuştur.

Anahtar Kelimeler: ayak bileği; lokalize tip; manyetik rezonans görüntüleme; tendon kılıfının dev hücreli tümörü

ABSTRACT

Tenosynovial giant cell tumor (TSDHT) is a slow-growing benign tumor arising from the tendon sheath or bursa. TSDHT of the foot and foot ankle has been reported much less frequently compared to the hand and knee. Morphologically it is classified as the localized form, often seen in the hand, and the diffuse form seen in large joints. Therefore, TSGCT should be considered in the differential diagnosis of the foot and ankle soft tissue tumors. Clinical and radiological findings of the intra-articular localized form of TSGCT in the ankle were presented in this study.

Keywords: ankle; localized type; magnetic resonance imaging; tenosynovial giant cell tumor

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[1] Health Sciences University Diskapi Yıldırım Beyazıt Training and Research Hospital, Department of Radiology, Ankara, Türkiye.
[2] Health Sciences University Keçiören Training and Research Hospital, Department of Radiology, Ankara, Türkiye.
Corresponding Author: Hatice Kaplanoğlu, Health Sciences University Diskapi Yıldırım Beyazıt Training and Research Hospital, Department of Radiology, Ankara, Türkiye. e-mail: hatice.altnkaynak@yahoo.com.tr

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INTRODUCTION

Tenosynovial giant cell tumor (TSGCT), or giant cell tumor of the tendon sheath, typically originates from the synovial cells of the tendon sheath.[1] Both inflammatory and neoplastic processes play a role in the pathogenesis of TSGCT.[1] Based on its localization, TSGCT is classified into two groups: intra-articular or extraarticular. The latter can be divided as localized and diffuse according to its growth pattern. The localized form usually affects the fingers and toes, is less aggressive than its diffuse form, and usually presents as a single nodular lesion.[2] Localized form of TSGCT is rarely seen intra-articular in large joints such as the hip, knee, and ankle.[3] Patients usually present with painless, single-solitary swelling. Magnetic resonance imaging (MRI) represents the most valuable imaging modality in the diagnosis and preoperative evaluation. The MRI signal characteristics of TSGCT vary depending on hemosiderin, foam cells, and hyalinized connective tissue.[3] Resection of the primary tumor with negative margins is the gold standard treatment for TSGCT.[2]

CASE REPORT

An eleven-year-old female patient complains of pain and swelling on her right ankle for about three months. There was no previous history of trauma. On physical examination, tenderness was present on the right side of her ankle with localized swelling. However, the mass shows deep tissue extension and no skin extension. Laboratory tests were within normal ranges. Radiographs of the ankle with lateral and anteroposterior views showed no bone damage. MRI showed a lobulated contoured mass lesion with dimensions of 40x20 mm arising from the posterior of the talus, located intra-articular region and extending through the peroneal tendons, which was hypointense in T1W images (Figure 1), heterogeneous intermediate-hyperintense in T2W images (Figure 2). Blooming artifacts of hemosiderin were observed in the gradient-echo (GRE) sequence (Figure 3). The enhancement pattern of the lesion after gadolinium injection was prominent (Figure 4). Diagnosis of pigmented villonodular synovitis or TSGCT was primarily considered, and total excision of the lesion was suggested. The informed consent form was taken from the patient. The patient was operated on under general anesthesia.

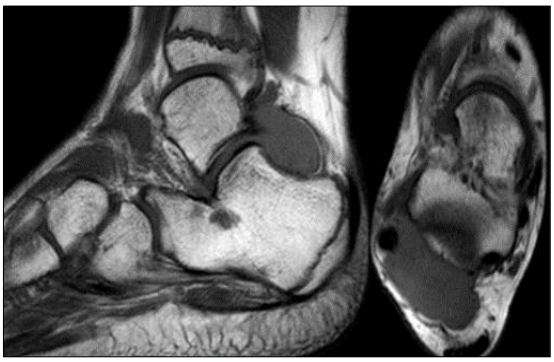

Figure 1. The sagittal and axial T1weighted images depict hypointense, lobulated contoured mass posterior of the talus.

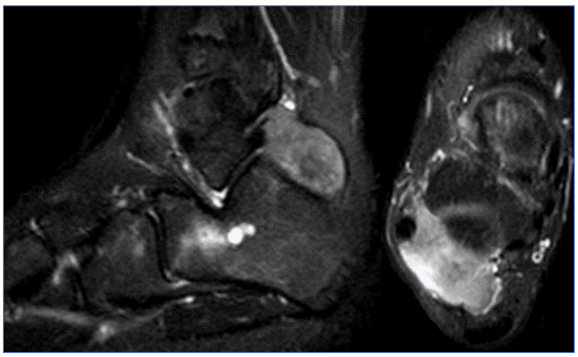

Figure 2. The sagittal and axial T2W images show heterogeneous intermediate-hyperintense, lobulated contoured mass posterior of the talus.

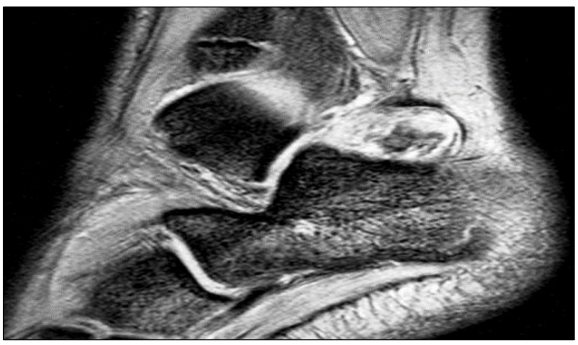

Figure 3. Blooming artifacts secondary to hemosiderin pigments were observed in the GRE sequence in the mass.

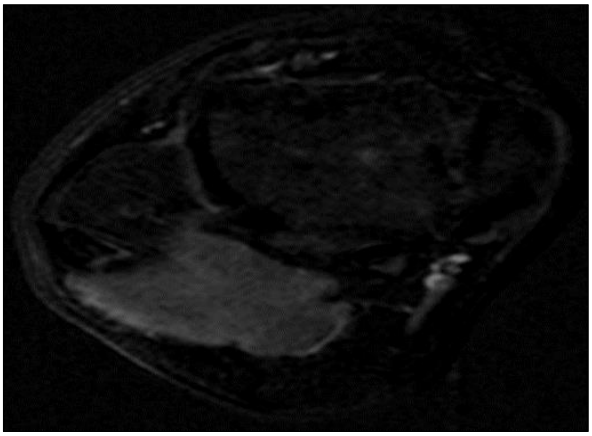

Figure 4. Subtracted postcontrast T1W image shows prominent enhancement of the mass.

Gross tissue evaluation of a pathology specimen showed cream to yellow colored, lobulated, partially encapsulated, 4x3.5x1.5 cm sized tissue with smooth surfaces. Microscopic examination revealed a round-oval mononuclear cells, hemosiderin-laden macrophages, and multinuclear giant cells in a fibrovascular stroma. Malignant components were not observed. Findings indicate that lesion was consistent with TSGCT.

DISCUSSION

TSGCT is a benign, slow-growing solid soft tissue tumor.[4] It is debatable whether the lesion is a true neoplasm or a reactive response to soft tissue trauma. Determination of cytogenetic abnormalities due to karyotype analysis, recurrence, and multifocality of lesion suggest that TSGCTs are true neoplasm.[5] On the other hand, the human androgen receptor (HUMARA) assay as TSGCTs have polyclonal proliferation indicates that the disease is reactive or hyperplastic.[5] Even generally believed their reactive nature; this association has not yet been fully determined.[6] It is twice as common in women and often occurs in the third to fifth decades.[7] The most common symptom is an asymptomatic mass. Additional symptoms such as pain and joint motion restriction may occur due to compression or invasion of adjacent tissues.[4]

TSGCTs are divided into two subtypes as intra-articular and extraarticular, based on location. Additionally, they can be classified as localized and diffuse forms according to their growth patterns. The localized type is the most common condition and is often seen in hands. The diffuse form usually occurs in large joints.[7]

Imaging modalities effectively distinguish TSGCT from other soft tissue tumors of the foot and ankle.[4] Ultrasonography is used to distinguish solid from cystic masses in which TSGCT is typically a homogeneous, solid, and hypoechoic mass.[8] MRI is proved to be the most useful non-invasive method for diagnosis and preoperative planning. MRI characteristics are associated with the contents of the hemosiderin of the tumor, and tumors are typically iso-to-hypointense in T1W images, and their signal is variable in T2W images, which is often seen as hypointense due to the hemosiderin pigments.[9]

Macroscopically, the tumor is usually gray-white colored, encapsulated, lobulated mass with yellow and brown spots. The amount and distribution of brown or yellow spots in macroscopic examination vary depending on the amount of hemosiderin-laden histiocytes.[7] Microscopic examination shows uniformly structured intra-extracellular hemosiderin-containing multinucleated giant cells, histiocyte-like cells, fibroblast-like cells, xanthoma cells, and vascular structures.[7]

In addition to TSGCT, the differential diagnosis of soft tissue tumors of the ankle consists of lipomas, synovial cysts, pigmented villonodular synovitis (PVNS), fibromatosis, undifferentiated pleomorphic sarcoma, desmoid tumor, leiomyosarcoma, and synovial sarcoma.[4] Even TSGCT rarely affects large joints such as the ankle and knee it should be kept in mind in the differential diagnosis of soft tissue tumors located in the foot-ankle.[4]

In the diffuse form of the disease, MRI features can be similar to those seen in PVNS.[10] In contrast to PVNS, TSGCT originated from the extraarticular synovium, and most of the lesions are located outside the ankle capsule.[10] A definitive diagnosis should be made before the operation because surgical treatment of the two tumors is different. While local resection of the affected tissue is effective in treating TSGCT, extensive excision with ankle fusion is recommended in the treatment of PVNS.[10] If left untreated, hypertrophic synovium and multiple soft tissue masses can lead to persistent pain, limitation of movement, joint destruction, and osteoarthritis.[11] The presented case had severe pain secondary to compression of the adjacent tissues. Treated cases have a high risk of recurrence and relapse between 4-44 %. In relapsing cases, re-excision is applied because the conversion to malignancy is not reported even after multiple relapses.[10]

Conclusion

It is important to keep TSGCT in mind in the differential diagnosis of a soft tissue tumor around the ankle or foot. MRI is the most appropriate method for diagnosing and preoperative evaluation of tumors and is valuable in determining adjacent tissue compression and extension. Although the definitive diagnosis is made by histopathological examination, MRI contributes to the diagnosis.

Conflict of Interests

The authors declare that there is not any conflict of interest regarding the publication of this manuscript.

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Ethics Committee Permission

Written informed consent was obtained from the patient.

Authors' Contributions

Concept/Design: HK, AT, VK. Data Collection and Processing: HK, RPK, TA. Data analysis and interpretation: AT, VK, TA. Literature Search: HK, TA, RPK. Drafting manuscript: HK, AT, VK. Critical revision of the manuscript: RPK, TA. Supervision: HK.

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