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Case Report

Histiocytic sarcoma mimicking localized tenosynovial giant cell tumor in the pediatric foot: A rare case report with MRI Findings ☆

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ABSTRACT

We present a rare case of histiocytic sarcoma (HS) occurring in the foot of a 12-year-old male, initially misdiagnosed as localized tenosynovial giant cell tumor (TSGCT). HS is an exceptionally uncommon hematologic malignant neoplasm, with its occurrence in children and extranodal sites being even rarer. To our knowledge, this is the first reported case of extranodal HS in the foot, emphasizing comprehensive MRI findings. Initially, the patient was diagnosed with TSGCT based on histological results following surgical resection. However, after recurrence and subsequent surgical resection, histological and immunochemical analyses led to a revised diagnosis of HS. This report focuses on the MRI findings of HS, highlighting the distinctions from localized TSGCT. While both conditions share histopathological similarities, immunohistochemical tests are crucial for accurate diagnosis. The report underscores the importance of differentiating HS for appropriate treatment.

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Introduction

Histiocytic sarcoma (HS) is an extremely rare and aggressive hematologic malignant neoplasm, accounting for less than 1% of all hematolymphoid neoplasms [1,2]. It exhibits morphologic and immunophenotypic features of histiocytic differentiation [1,2]. HS predominantly affects males across a wide age spectrum, with peak incidence in individuals aged 0-29 years and 50-69 years [1]. The lymph nodes are the most common primary site of involvement, although HS can also manifest in extranodal sites such as the skin and gastrointestinal tract [1–4]. Conversely, tenosynovial giant cell tumor (TSGCT) is a benign fibrous tissue tumor that primarily occurs in the hand and ranks second in prevalence after ganglion cysts [5]. Localized TSGCT is the most common form, and magnetic resonance imaging (MRI) is the preferred diagnostic modality [5,6]. In contrast, HS is diagnosed based on clinical features, immunophenotype, and genetic findings, with no specific MRI features reported [1] and there have been few reports of MRI findings in the extremities [2,4]. Therefore, diagnosing HS, especially in pediatric cases involving extremities, can be exceedingly challenging due to the absence of distinct known MRI findings. Moreover, although these two conditions share striking histopathological similarities, HS should be considered only after excluding other lymphoid lineage tumors through immunohistochemical tests, given its extremely rare occurrence. Additionally, the prognosis for HS varies significantly based on the site of involvement; HS invading the skin generally presents a relatively favorable prognosis. However, while TSGCTs are mostly benign, the malignant nature of HS necessitates a different approach to treatment. Hence, distinguishing HS is crucial for determining the appropriate course of treatment.

This case report aims to present the ultrasound (US) and MRI findings of HS in the foot and differentiate it from localized TSGCT, which exhibits similar imaging features.

Case report

A 12-year-old male without any underlying diseases presented at our orthopedic outpatient department with a plantar mass persisting for over a year. The mass, located in the plantar region of the first metatarsophalangeal (MTP) joint, was characterized by bulging skin coverage with firm consistency and erythema. However, there was an absence of pain, tenderness, and no signs of infection were evident (Fig. 1A). US revealed a multilobulated, heterogeneous hypo- and hyperechoic mass with increased internal vascular flow that closely contacted the flexor hallucis longus (FHL) tendon (Fig. 2). The initial differential diagnosis considered localized TSGCT and hemangioma, common conditions in this area. Surgical resection was performed, and the mass was histopathologically diagnosed as localized TSGCT without further immunohistochemical testing. Approximately 4 months later, the patient presented with recurrent mass in the same region, resembling the previous excised mass but larger in size, accompanied by throbbing pain and skin erosion (Fig. 1B). Hematological laboratory findings revealed no specific abnormalities. To obtain a detailed evaluation, contrast-enhanced foot MRI was performed (Fig. 3). MRI showed a lobulated mass with relatively iso- to high-signal intensity (SI) on T2/proton density (PD)-weighted images and marked contrast enhancement. While the mass enveloped the FHL tendon and exhibited nonenhancing regional dark SI on all sequences akin to TSGCT, it also displayed partial infiltration into adjacent muscle. A second surgical excision was conducted, followed by histopathological examination and additional immunohistochemical staining. The 3.7 x 2.3 x 2.3 cm soft tissue mass, adhering to the skin, exhibited monocytoid cell proliferation with nuclear atypia and increased mitotic counts, favoring malignancy. The immunohistochemical staining results were as follows (Fig. 4): positive for leukocyte common antigen (LCA) and CD68/CD4, negative for CD1a, equivocal for S100, high Ki-67, and negative for other T-cell or B-cell markers. Although CD68 can also appear positive in TSGCT, considering the other findings, HS was conclusively diagnosed. The patient underwent a follow-up period of approximately 9 months without specific treatment, except for conservative measures. The surgical wound healed well without complications, and no recurrence was observed during this period.

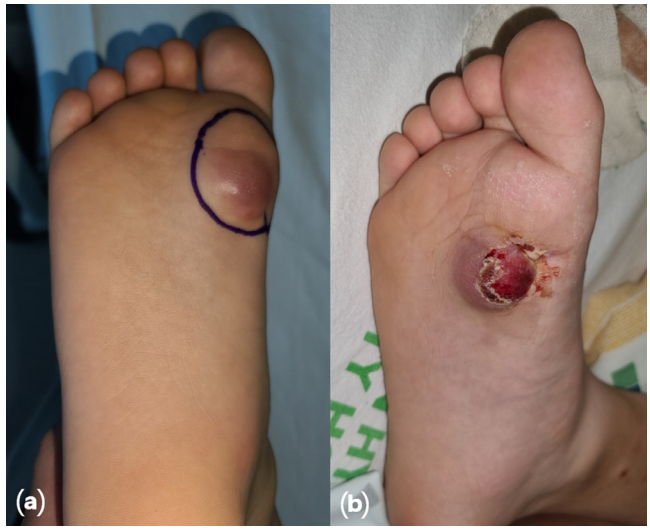

Fig. 1 – Photographs of the right foot at the first visit (A) and at the time of recurrence four months later (B).

Discussion

HS is an exceedingly rare tumor characterized by malignant cell proliferation that exhibits morphological and immunophenotypic characteristics of mature tissue histiocytes, according to the 2008 WHO classification [1]. Its rarity makes distinguishing HS solely via histopathological examination without immunophenotypic analysis challenging, as it shares similarities with various other conditions such as diffuse large B-cell lymphoma, high-grade lymphoma, melanoma,

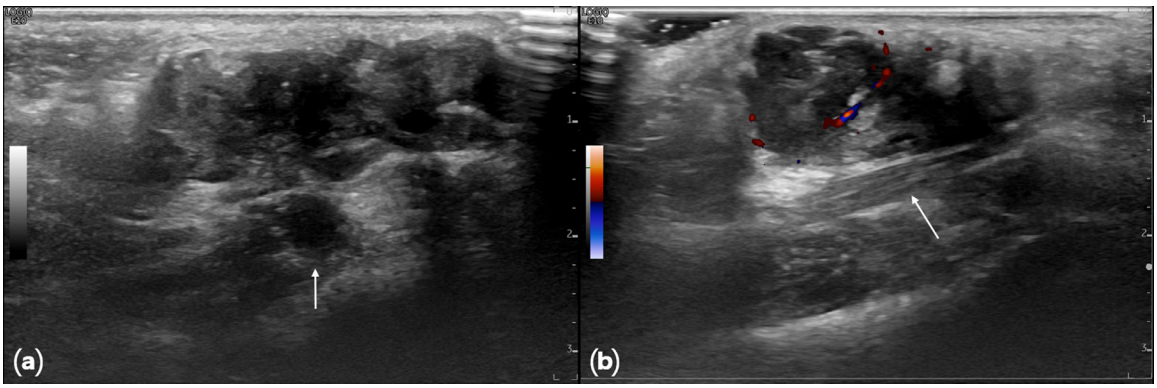

Fig. 2 – US images of the plantar mass on right foot. (A) transverse scan of B-mode and (B) sagittal scan of color Doppler image. A multilobulated mass measuring approximately 5 cm with heterogeneously low echogenicity and an inner focal hyperechoic portion is seen along the flexor hallucis longus tendon (arrow), which exhibits vascular flow on color Doppler imaging.

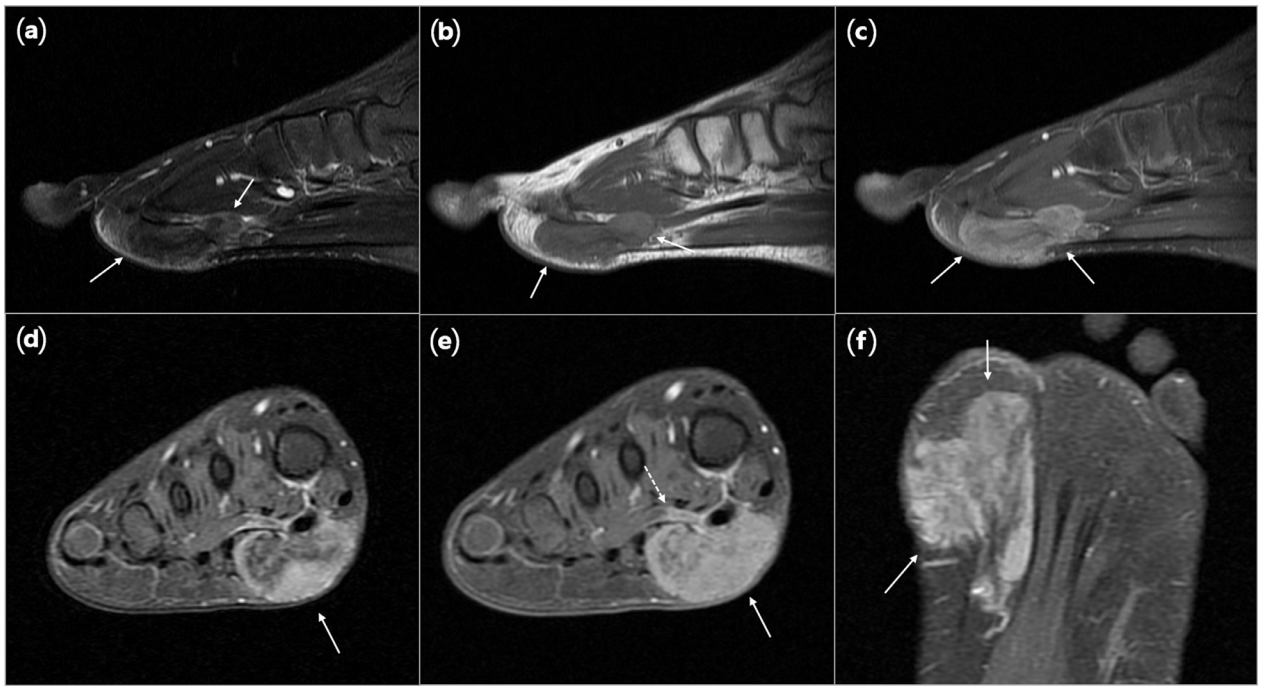

Fig. 3 – MRI of the mass in the right foot: sagittal (A) T2-weighted image (WI) with fat saturation, (B) T1-WI, (C) contrast-enhanced T1-WI, and (D) coronal proton density (PD)-WI with fat saturation, contrast enhanced T1-WI of coronal (E) and axial (F) scans. A multilobulated mass measuring approximately 3.1 x 2.1 x 5.0 cm (arrows) is observed, abutting the flexor hallucis longus tendon and partially infiltrating the adjacent muscle (dashed arrow). The mass shows high signal intensity on PD-WI and intralesional dark signal intensity content on PD-WI, T1-WI, and T2-WI, with mostly homogeneous enhancement. The adjacent bones and first metatarsophalangeal joint are well separated.

and sarcomas displaying epithelioid features (including epithelioid sarcoma and anaplastic carcinoma). Immunophenotypic studies are crucial before diagnosing HS to exclude other diseases [1], particularly because its infrequency often renders it elusive unless specifically suspected. Some cases of HS occur concurrently or sequentially with lymphoblastic lymphoma or leukemia, with a higher prevalence in children and young adults [1,7]. However, our case did not have any comorbidities. Although TSGCT can occur at any age, it is rare in children and most commonly affects individuals in their 30s and 40s [5,6,8]. Localized TSGCT predominantly occurs in the digits (85%) around synovial sheath or joints [6,8]. In our case, both entities did not entirely match, as the child had no underlying diseases, but the region around the flexor tendon of the MTP joint can occasionally be affected by TSGCT. Therefore, diagnosing TSGCT solely based on histopathological examination and radiological findings (MRI) during the initial surgery was a highly rational conclusion.

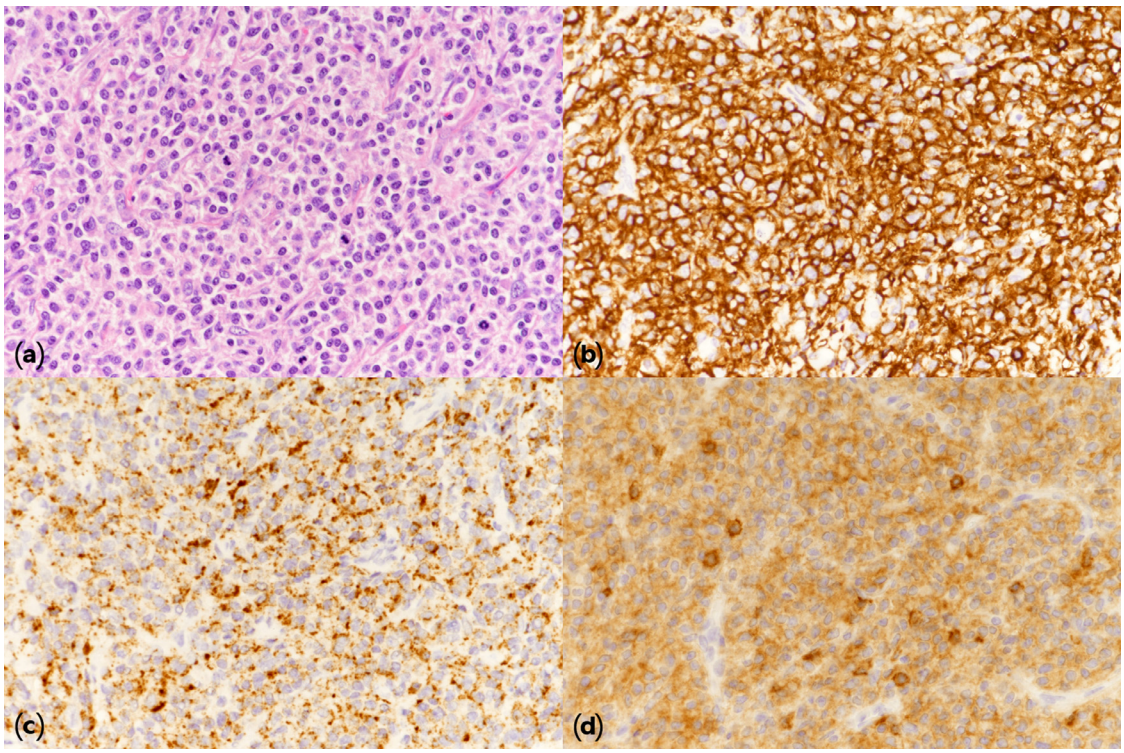

Fig. 4 – Histopathology and immunohistochemistry staining images. (A) The tumor cells have moderate to abundant eosinophilic cytoplasm, irregularly folded, and eccentrically placed nuclei, and variably prominent nucleoli (H&E, x400). The tumor cells are positive for leukocyte common antigen (LCA) (B), CD68 (C), and CD4 (D) (x200).

Differentiating between HS and TSGCT poses challenges in both morphological and histological assessments [8]. Morphologically, HS closely resembles TSGCT when it occurs rarely around joints or exhibits a multinodular pattern. HS can present with a wide range of symptoms depending on the location of occurrence, including involvement of the skin, appearing either as a solitary or multiple lesions. Commonly, both diseases manifest as slowly growing, painless, palpable masses, lacking specific clinical findings [1,2,6]. In this case, as well, it was initially discovered as a painless mass. Histologically, HS often displays abundant eosinophilic cytoplasm and significant inflammatory infiltration (neutrophils, lymphocytes, or eosinophils) with various variations [1,2]. Moreover, localized TSGCT may also demonstrate large cells locally, severe cellular pleomorphism, increased mitotic figures, and areas of necrosis, making it exceedingly difficult to differentiate between the 2 conditions based on histopathological findings only. Additionally, HS shares similarities with mature tissue histiocytes, making it difficult to differentiate from TSGCT which lacks multinucleated giant cells [8]. CD163 and CD68 are commonly used immunohistochemical markers for HS [4,8,9]. However, both markers can also yield positive reactions in TSGCT [8,9]. CD163, with its high specificity for histiocytic cells, is particularly valuable in identifying histiocytic malignancies [1,9]. Furthermore, as histiocytes originate from monocytes, it is essential to exclude B-cell or T-cell leukemias. In our case, the negative CD1a staining helped exclude Langerhans cell histiocytosis [2].

The typical imaging findings of TSGCT include its specific location, such as the periarticular region or around the tendon sheath, appearing to envelop these structures and sometimes exhibiting adjacent bone erosion [5,6]. TSGCT usually presents as an iso- to low-SI mass on T2-weighted images due to hemosiderin accumulation. Contrast-enhanced imaging depicts substantial enhancement throughout the mass, except in areas with hemosiderin. Increased vascularity can also be observed on color Doppler ultrasound [5,6]. In contrast, limited typical imaging findings for musculoskeletal extranodal HS have been reported. Prior case reports of HS involving the spine and orbit mentioned MRI usage without detailed image analyses [3,10–12]. In these cases, most of the masses showed homogeneous T2-iso to high SI and mostly homogeneous enhancement, infiltrated bone or adjacent muscle and other soft tissues, but partially showed a pattern of widening adjacent neural foramen and deviation of the dural sac, which resembled lymphoma, and the mass surrounding the FHL tendon and infiltrating adjacent muscles was consistent with our case. Another previous case involving HS in the orbit was similar to our case in that the mass infiltrated into the skin and appeared as a reddish palpable mass [12]. Although our case shared certain similarities with TSGCT concerning mass location, spread, and focal dark SI, a comprehensive analysis of previous cases revealed distinguishing features of HS: relatively high SI on T2-weighted images, mostly homogeneous enhancement, and the ability to invade the entire layer of soft tissue including the muscle layer.

Localized TSGCT has been reported to have a recurrence rate of less than 10% after surgical excision [6,13]. Although the overall prognosis of HS is known to be poor [1], the prognosis of surgical resection of localized, extranodal HS is relatively favorable [1,7], consistent with our case. However, advanced HS presents a high mortality rate, and while treatments such as chemotherapy are available, there is currently no established definitive treatment [1,4,7].

In this study, we described an exceedingly uncommon instance of extranodal HS located in the foot, mimicking localized TSGCT. Detailed US and MRI findings were provided, crucial in accurately diagnosing HS at an early stage and aiding in its differentiation from the more prevalent localized TSGCT. This will assist in establishing the patient's treatment plan and predicting their prognosis.

Patient consent

The IRB approval was obtained from Soonchunhyang University University Bucheon Hospital. Written informed consent was obtained from the patient's legal representative.

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