

Precalcaneal congenital soft-tissue lesions in children: A case report of fibrous hamartoma of infancy and an approach to differential diagnosis

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Precalcaneal soft-tissue lesions are uncommon in the pediatric population and can present a diagnostic challenge. Fibrous hamartoma of infancy (FHI) is relatively rare in this location. We report an interesting case of FHI in a 3-years-and-10-months-old boy in the precalcaneal location that was present since birth. We describe the imaging findings of FHI on X-ray, ultrasound, and MRI and discuss the differential diagnoses. It is important to consider FHI during differential diagnosis and be aware of the imaging features of other common possible diagnoses in the precalcaneal region for appropriate management.

Introduction

Subdermal fibromatous tumor of infancy, described by Reye in 1956, was later renamed fibrous hamartoma of infancy (FHI) by Enzinger in 1965 after a systematic review of 30 cases with similar histopathology. Several case series have been reported since then, the largest by Dickey and Sotelo-Avila in 1998. In this clinicopathologic review of 197 cases, only two cases (1%) were reported in the feet (1).

We describe here a third case and the specific imaging findings of FHI in the foot. Our aim is twofold: 1) Precalcaneal soft-tissue masses present since birth are relatively uncommon in young children. It is important to consider FHI in the differential diagnosis of precalcaneal congenital soft-tissue lesions in young children. 2) We describe the imaging features of FHI and how to differentiate it from “do

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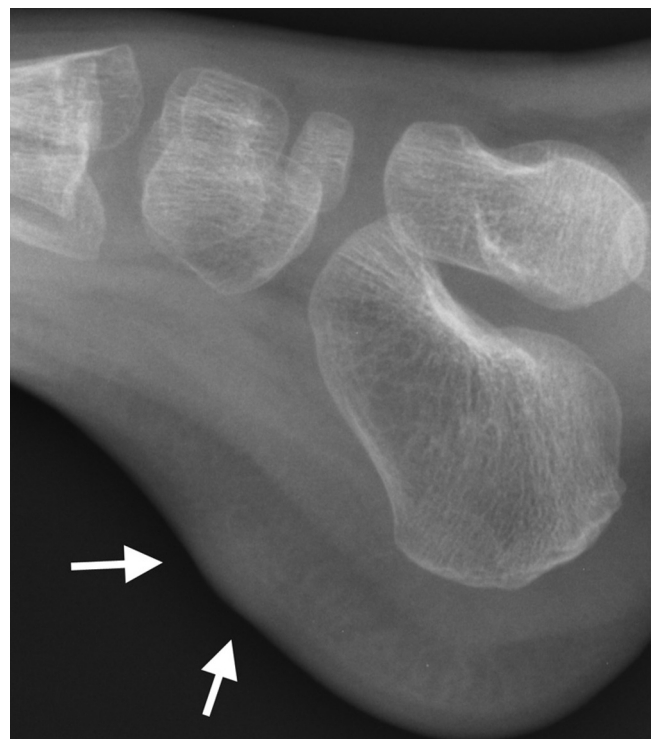


Figure 1. Nearly 4-year-old boy with FHI. Lateral X-ray of the foot showing subcutaneous mass in the precalcaneal region (arrows).

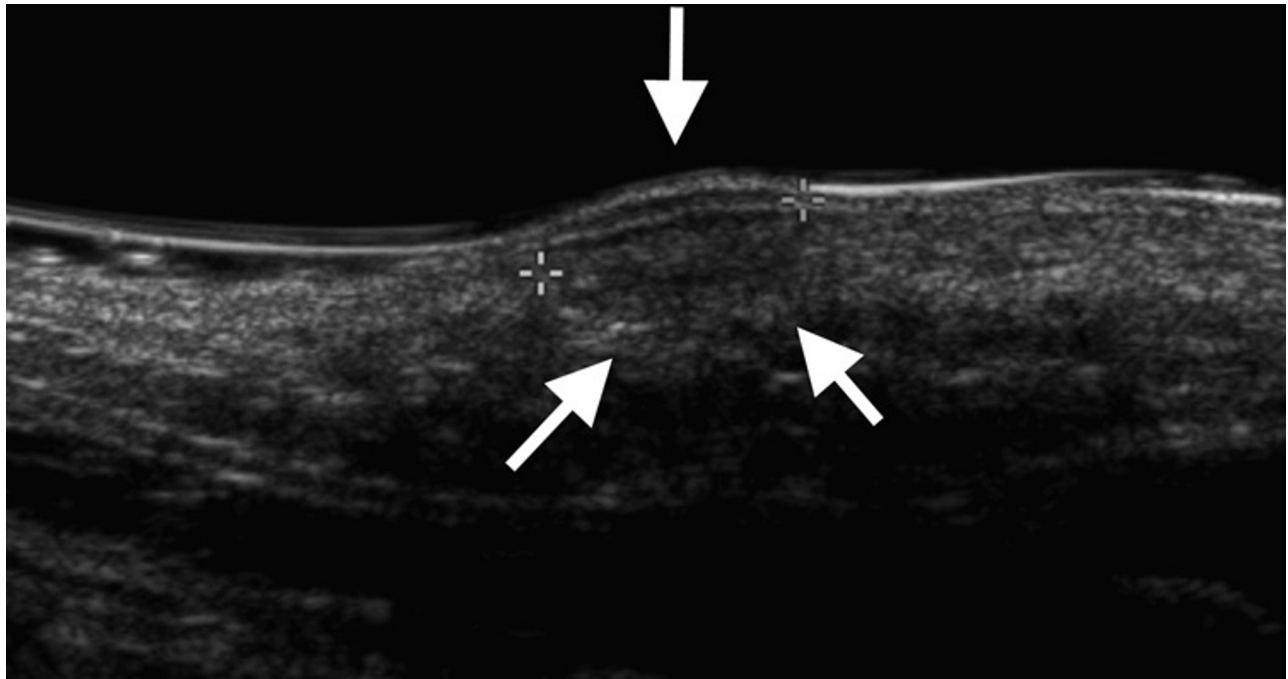


Figure 2. Nearly 4-year-old boy with FHI. Ultrasound demonstrating ill-defined hypoechoic mass in the precalcaneal region (arrows).

not touch lesions” such as congenital fibrolipomatous hamartoma (PCFH) and other soft-tissue masses in the precalcaneal region in young children (2).

Case report

A previously healthy male child, age 3 years and 10 months, presented with recent-onset pain in a soft, skin-colored lesion present since birth in the anteromedial plantar aspect of the right foot. The lesion was slowly growing with age. He walked on his forefoot and refused to bear weight on his right heel due to pain. There was no history of trauma, infection, skin-color changes, or any other cutaneous abnormalities. There was no significant medical or family history. Physical examination revealed mild antalgic gait with an ill-defined, approximately 1.5-cm soft, non-tender, freely mobile mass within the subcutaneous tissue in the anteromedial plantar aspect of the right heel. The overlying skin was unaltered. General physical and systemic examinations were unremarkable.

Radiographs showed an ill-defined, approximately 9.3 x 9.1-mm soft-tissue opacity within the superficial soft tissues in the anteromedial plantar aspect of the right heel (Fig. 1). There was no bony erosion, demineralization of bones, fracture, dislocation, foreign body, or soft-tissue calcification. Ultrasound revealed an ill-defined, approximately 1cm x 1cm, heterogeneously hypoechoic subcutaneous mass with no calcification, necrosis, or invasion of the plantar fascia (Fig. 2). MRI revealed an ill-defined mass in the subcutaneous fat of the right heel that was hypointense compared to muscle on T1W images (Figs. 3A and 3B) and that

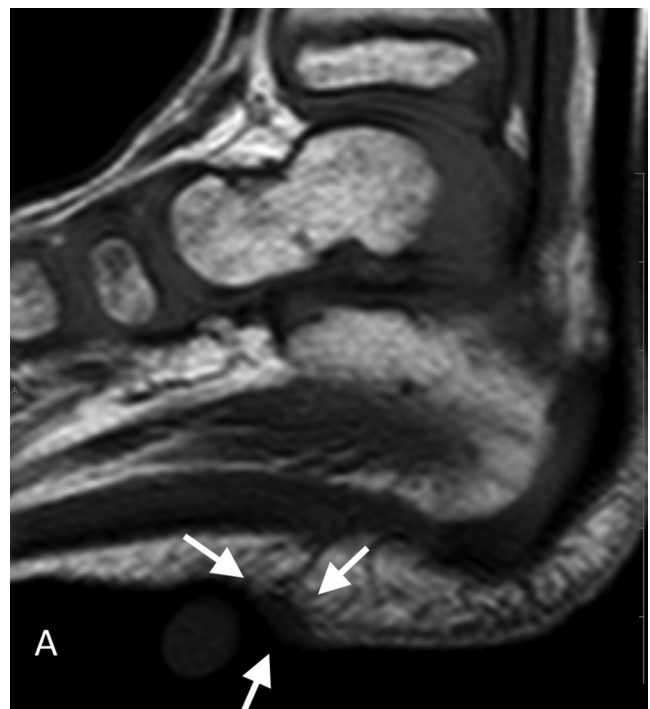


Figure 3A. Nearly 4-year-old boy with FHI. Sagittal T1 sequence reveals an ill-defined, hypointense, subcutaneous mass in the precalcaneal region (arrows). The skin marker slightly distorts the normal anatomy in this region.



Figure 3B. Nearly 4-year-old boy with FHI. Coronal T1 sequence reveals an ill-defined, hypointense, subcutaneous mass in the precalcaneal region (arrows). The skin marker slightly distorts the normal anatomy in this region.

had heterogeneous signal with mild hyperintensity on sagittal STIR sequence (Fig. 3C) without significant contrast enhancement after administration of gadolinium. The child underwent complete excision of the lesion. Grossly, the tumor was unencapsulated with pink-tan-yellow, soft, lobulated tissue. Histopathology confirmed the diagnosis of FHI with the presence of mature adipose tissue and bundles of fibrous tissue interspersed with myxoid tissue. There were no mitoses, calcifications, hemorrhages, or necroses within the tumor. Postoperatively, the child recovered well. There was some evidence of scar contracture, but he walked well without any complaints.

Discussion

FHI is a rare benign soft-tissue tumor, most common under the age of two years, that occasionally presents at birth (3). Most commonly, it involves the axilla, shoulder region, upper arm, upper trunk, inguinal region, and external genital area (3). FHI has a male preponderance with

no familial predisposition and no association with any other congenital malformations, neurocutaneous syndromes, or other hamartomas. The usual presentation is a solitary, soft,



Figure 3C. Nearly 4-year-old boy with FHI. Coronal STIR sequence showing an ill-defined, minimally hyperintense, subcutaneous mass in the precalcaneal region (arrows).

painless, poorly circumscribed, freely mobile subcutaneous mass, typically measuring 2cm to 5cm (3). The tumor is known to grow rapidly initially up to the age of 5 and slows down with age, but does not regress spontaneously (3). Grossly, FHI is located in the subcutaneous fat or lower dermis and is round or bosselated, with grayish-white tissue intermixed with irregular islands of mature fat. The pathognomonic histological feature of FHI on routine hematoxylin and eosin stain is the presence of an “organoid” pattern with 1) intertwining, well-defined, dense fibrocollagenous trabeculae, 2) varying amounts of mature fat, and 3) immature-appearing, well-defined myxoid areas, which are not normally present in the subdermis (3).

Superficial soft-tissue masses are most commonly diagnosed clinically. However, imaging studies (most commonly

plain radiographs, ultrasound, and MRI) are used if there are any confounding clinical features. They can be diagnostic, obviating surgical intervention. Ultrasound is useful to differentiate solid from cystic masses and to demonstrate vascularity and the involvement of deeper tissue planes. Computed tomography is not usually performed due to poor soft-tissue differentiation and risk of radiation exposure in children. MRI is useful in the differential diagnosis of precalcaneal soft-tissue lesions because of multiplanar localization and excellent soft-tissue discrimination. It can be specific in identifying soft-tissue tumors based on the amount of fat and fibrous tissue (4). Fibrous tissue is hypointense compared to muscle on T1W and T2W images, myxoid tissue has low signal on T1W images and high signal on T2W images, and fat is hyperintense on T1W and T2W images (5). Characteristically, FHI has low signal on T1W images and heterogeneous signal on T2W images on MRI, with fibrous trabeculae interspersing with fat in an organized pattern in the subcutaneous tissue--which is pathognomonic of FHI (4-6). Management of FHI includes complete local excision of the tumor, which is curative with rare recurrences. Prognosis for FHI is excellent with no malignant potential, even if recurrent (3).

PCFHs are relatively more common benign tumors in the precalcaneal region and were seen in 5.9% of newborns and 39.4% of infants in a survey of 269 newborns and 189 infants (2). Usually they manifest at birth or in the first few months as bilateral, symmetric, soft, nontender nodules growing slowly and paralleling the growth of the child. They are also known as congenital peizogenic-like pedal papules, bilateral congenital adipose plantar nodules, benign anteromedial plantar nodules of childhood, bilateral congenital fatty heel pads, or infantile pedal papules. The different etiologies for PCFH include hamartomatous formation of fat and connective tissue, hypertrophy secondary to incomplete involution of the normal fetal subcutaneous plantar tissue, and congenital herniation of fat tissues (2). They are managed conservatively without surgery (3). There are case reports of ultrasound findings that demonstrate herniation of fat through the dermis with loss of normal hyperechogenic signal between the dermis and the subcutaneous fat layer (2). There is no published case report of typical MRI findings of PCFH, but the features would be expected to be similar to ultrasound and to show fat herniation through the dermis.

Fibrolipomatous hamartoma (FLH) of medial plantar nerve, a rare and a separate entity, may also present at birth in the precalcaneal region. It has characteristic MRI findings of thickened nerve fascicles surrounded by fat (7). Tumors with fibrous proliferations like soft-tissue sarcoma, juvenile hyaline fibromatosis, fibrosarcoma, desmoid-type fibromatosis, aponeurotic (calcifying) fibroma, dermatofibromas, and neurofibromas also have to be ruled out based on their clinical history, imaging, and histological studies (3, 6, 8). These lesions are devoid of fat signal on imaging, most of them being hyperintense on T2W MRI images with variable contrast enhancement. Other differential diagnoses such as lymphatic malformation, venous malforma-

tion, neurofibroma, infection, and metastatic lesions from neuroblastoma also have T2 hyperintensity with characteristic enhancement patterns. Lymphatic malformations appear as multiloculated cystic lesions with peripheral and septal enhancement. Fluid-fluid levels may be seen. Hemangioma appears as an avidly enhancing soft-tissue lesion with high flow. Venous malformations appear as an enhancing soft-tissue mass with slow flow. This can be confused radiographically with other soft-tissue lesions such as rhabdomyosarcoma or metastatic lesions from neuroblastoma. Fat-containing lesions like lipoma, lipoblastoma, and involuting hemangioma can be easily differentiated, as they are devoid of fibrous tissue signal on imaging.

Conclusion

Precalcaneal soft-tissue masses are relatively uncommon in young children, and are often diagnosed clinically and not imaged. "Don't touch" lesions such as PCFHs are relatively more common among the precalcaneal soft-tissue lesions, do not require surgery, and are managed conservatively. FHI, on the other hand, is uncommon in the precalcaneal region, but treatment with complete local excision is usually curative with an excellent prognosis. It is important to be aware of the possibility of FHI in the precalcaneal region as well as of the differential diagnosis of common congenital soft-tissue masses in this region in young children. Imaging studies are useful to distinguish FHI, PCFH, FLH and other precalcaneal congenital pathologies for appropriate clinical management.

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