Intravascular Papillary Endothelial Hyperplasia (IPEH) of the Foot

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Section: Musculoskeletal system
Area of Interest: Soft tissues / Skin Extremities
Procedure: Imaging sequences
Procedure: Contrast agent-intravenous
Imaging Technique: Ultrasound
Imaging Technique: Ultrasound-Colour Doppler
Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 20 years, male

Clinical History:
A 20-year-old male patient with a palpable mass on the plantar aspect of the foot with mild tenderness was admitted to the hospital for further evaluation. He had no history of recent trauma or any systemic complaints.

Imaging Findings:
Conventional radiographs of the foot showed no calcification within the soft tissues. No accompanying osseous changes were observed. Ultrasonography (US) revealed a hypoechoic, well-defined multilobulated mass lesion. There was no vascularization within the lesion on Doppler US (Fig. 1). Magnetic resonance imaging (MRI) confirmed the presence of a well-defined multilobulated mass within the subcutaneous fat at the plantar aspect of the foot, extending along the fascial planes among the flexor muscles. The lesion was slightly hyperintense when compared to muscle on T1W images and peripherally hyperintense on fat saturated T2W images (Fig. 2). Marked peripheral enhancement was observed following the administration of gadolinium (Fig. 3). A wide excisional biopsy was performed.

Discussion:
IPEH is generally considered an unusual form of thrombus organisation with excessive papillary endothelial proliferation and frequently seen superficially in the head, neck, trunk and fingers. It has no age predisposition but a slight predominance in females. It represents approximately 2-4% of the vascular tumours of the skin and soft tissue. Surgical excision is the treatment of choice for both diagnostic and curative purposes. Recurrence is extremely rare if the lesion is completely excised [1, 2]. Histologically, three different types have been described. The primary or pure form accounts for 56% of cases and arises in a dilated vessel. The secondary or mixed form (40%) originates on a pre-existing vascular lesion such as a haemangioma, pyogenic granuloma or vascular malformation. The very uncommon third form (4%) is the extravascular form which presents clinically as a primary neoplasm and histologically may mimic angiosarcoma [3]. Except one case which is located very close to the mandibula [4], no accompanying osseous change was defined with IPEH [5, 6]. Internal calcification within the mass has been reported in the mixed form which originates on a haemangioma and calcifications may be considered as phleboliths. Magnetic resonance imaging (MRI) is important for diagnosis and preoperative planning. There are only a few small
case reports and series that describe the MR appearance of IPEH [2, 7, 8]. The signal intensity on conventional MR sequences depends on the age of the thrombosis. Therefore, the lesion is hyperintense to muscle in the acute and medium stage and demonstrates hypo- to isointense signal in the chronic stage on T1W images and hyperintense on T2W images in all stages [2]. There are several patterns such as diffuse or septal and peripheral enhancement on contrast-enhanced studies. Septal and peripheral enhancement pattern seems to be the more frequent type which is consistent with the histopathological appearance defined as central thrombi with peripheral endothelial proliferation [2, 7, 8].

Ultrasound (US) findings of IPEH were reported to be nonspecific especially in the pure form. The lesion is mostly homogeneously hypoechoic on gray scale US. Doppler US may demonstrate septal or peripheral vascularity, reflecting the findings on MRI [8, 9].

In conclusion, IPEH is a rare vascular soft tissue tumour which shows endothelial proliferation with thrombus formation arising in a dilated vessel. MRI signal intensity characteristics are related to the age of the thrombus occurring within the lesion. Peripheral enhancement pattern is the most frequent type among a few cases reported in the literature, likewise in our case. US findings described in this case are nonspecific as hypoechoic and homogeneous on gray-scale US without internal vascularity on Doppler US. IPEH should exist in the differential diagnosis of a soft tissue tumour with peripheral or septal enhancement.

**Differential Diagnosis List:** Intravascular papillary endothelial hyperplasia, Haemangioma, Vascular malformation

**Final Diagnosis:** Intravascular papillary endothelial hyperplasia

**References:**


Figure 1

Description: The lesion is hypoechoic and well-defined in a multilobulated appearance on gray-scale US (a). On Doppler US (b), there is no internal vascularity. Origin:
Description: On T1W (a) the lesion is isointense-slightly hyperintense (arrow). On fat-saturated T2W (b) the lesion is peripherally hyperintense containing low signal foci (arrow). The lesion also has a small intramuscular component (arrowhead). Origin:
Description: On contrast-enhanced fat-saturated T1W coronal image, the lesion demonstrates peripheral enhancement (arrow). Origin: