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Verrucous venous malformation

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Abstract

Verrucous venous malformation, also known as verrucous hemangioma, is a superficial vascular malformation with a variable degree of hyperkeratosis that is composed of capillaries and veins in the dermis and sometimes subcutaneous tissue. We describe a 53-year-old man who presented with a large hyperkeratotic plaque of the left dorsal and plantar foot. Biopsy revealed verrucous acanthosis of the epidermis and a proliferation of thin-walled vessels in the dermis. We provide a brief review of the clinical and histopathologic presentation, differential diagnosis, and management of this rare entity.

Keywords: verrucous venous malformation, verrucous hemangioma, venous malformation, vascular malformation

Introduction

Verrucous venous malformation (VVM), also known as verrucous hemangioma, is an uncommon vascular malformation that is associated with reactive skin changes that include a variable degree of hyperkeratosis and papillomatosis. These skin changes tend to develop over time. Because of its similarity to other vascular malformations, it is frequently misdiagnosed clinically. We describe a 53-year-old man who presented with a large hyperkeratotic plaque of the left foot overlying a vascular stain. On clinical exam, the significant hyperkeratosis was reminiscent of an infection or neoplasm. However, routine histology confirmed the vascular origin, establishing a diagnosis of verrucous venous malformation.



Figure 1. A) Hyperkeratotic plaque of the left foot (dorsal view). The lateral flesh-colored subcutaneous mass represents an atypical feature of the verrucous venous malformation. **B)** Hyperkeratotic plaque of the left foot (medial view). **C)** Hyperkeratotic plaque of the left foot (plantar view). The vascular stain is most visible on the plantar aspect of the foot.

Case Synopsis

A 53-year-old man hospitalized for hematochezia was incidentally noted to have a large hyperkeratotic plaque on his left foot. He reported that he was born with a violaceous patch on the plantar foot that grew thicker over time. The lesion interfered with ambulation and occasionally drained serosanguinous fluid but was otherwise asymptomatic. He had immigrated to the U.S. from Mexico more than 10 years earlier and denied a history of agricultural work, contact with animals, or travel.

Physical examination revealed a thick, hyperkeratotic tan to brown plaque extending in a curvilinear fashion from the dorsal foot medially to the plantar foot, measuring 14cm in width and 6cm in height. There was a soft, compressible nodule present on the dorsolateral foot and an irregular violaceous patch on the plantar foot (**Figure 1**). The remainder of the skin examination was unremarkable.

Punch biopsy specimens (4.0mm) were obtained from the left dorsal foot for histologic analysis with hematoxylin-eosin (**Figure 2**) and tissue culture. Biopsy revealed irregular acanthosis and papillomatosis of the epidermis (**Figure 2A**). On higher power, there was a proliferation of vessels in the papillary and reticular dermis (**Figure 2B**). Tissue culture grew *Trichophyton spp*, consistent with an overlying tinea pedis infection.

Case Discussion

Verrucous venous malformation, also known as verrucous hemangioma, is a rare superficial vascular malformation that is frequently clinically misdiagnosed. It consists of variable degrees of hyperkeratosis overlying a malformation composed of capillaries and veins in the dermis and sometimes subcutaneous tissue. Recently, a somatic mutation of *MAP3K3* gene was found to be associated with this lesion [1].

Verrucous venous malformations typically present as reddish-brown solitary or multiple hyperkeratotic plaques or nodules with diameters ranging from

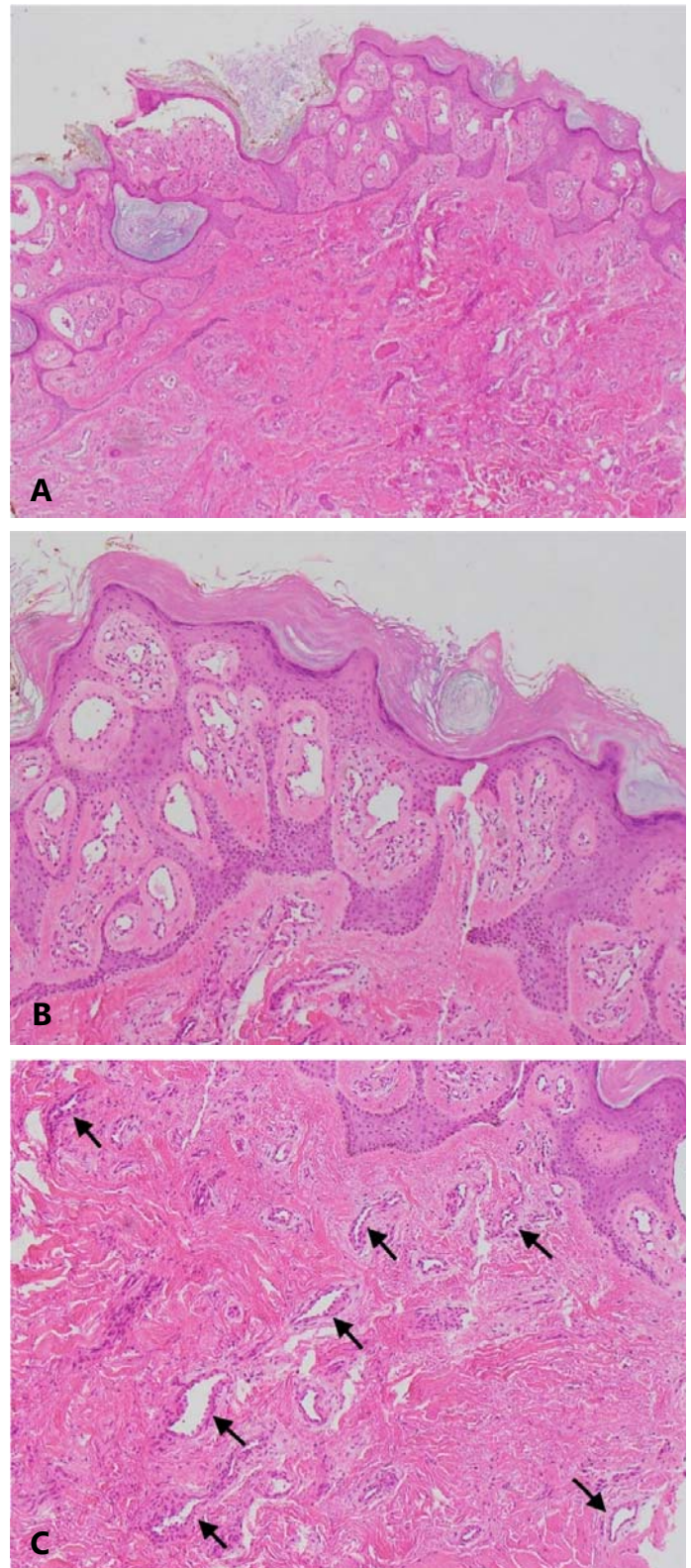


Figure 2. A) Punch biopsy specimen from the left foot shows prominent acanthosis and papillomatosis of the epidermis. H&E, 10 \times . **B)** An acanthotic and papillomatous epidermis overlies a proliferation of thin-walled vessels in the papillary dermis. H&E, 40 \times . **C)** There is a proliferation of thin-walled vessels of various sizes (marked by arrows) in the dermis. H&E, 40 \times .

5mm to 2cm [2]. They sometimes show a linear or Blaschkoid-like distribution. The lesions are most frequently seen on the extremities and buttocks; less common locations include the trunk and axilla [2]. The vascular component is usually present at birth as a violaceous stain, whereas the hyperkeratotic portion typically develops over time. The degree of hyperkeratosis is widely variable and may lead to pain, pruritus, or bleeding, as in our patient.

The typical presentation of VVM may be sufficient to confirm the diagnosis without a biopsy. The vascular portion of the lesion is frequently best visualized at the periphery (owing to the central hyperkeratosis) and is composed of an erythematous patch with small vascular red to violet dots [3]. The dots are accentuated with dermoscopy. However, unlike other vascular lesions such as capillary malformations, the vascular dots in verrucous venous malformations are also visible to the naked eye when viewed up close [3].

Despite its characteristic clinical presentation, VVM is under-recognized and often misdiagnosed. Its hyperkeratotic appearance leads to frequent misdiagnosis as a lymphatic malformation or angiokeratoma [2]. Apart from these entities, the differential diagnosis also includes venous malformation, capillary-lymphatic malformation, and verrucous epidermal nevus.

Histopathology of VVM shows acanthosis of the epidermis and a proliferation of small, often bizarre-appearing vessels in the dermis and subcutaneous tissue. The endothelial cells of the abnormal vessels stain positively with CD31.

There is limited data on the treatment of verrucous venous malformations. In the medical literature,

VVMs have been treated with laser therapy, radiotherapy, surgery, cryotherapy, topical corticosteroids, and sirolimus with variable success. These lesions tend not to respond to minimally invasive treatments such as cryo-ablation [4]. Several studies have suggested that complete excision is optimal, as they tend to enlarge and become more keratotic in response to injury, infection, or subtotal resection [5]. More recently, a retrospective analysis of 10 patients with a mean age of 17 months suggested that oral sirolimus may be a safe and effective treatment. In this study, oral sirolimus administered twice daily at a dose of 0.8mg/m² led to a volume reduction of at least 90 percent in all patients [6]. There are no available studies evaluating the safety and efficacy of oral sirolimus for the treatment of verrucous venous malformation in adults.

Conclusion

Verrucous venous malformation is a rare condition characterized by a proliferation of capillaries and veins in the dermis and sometimes subcutaneous tissue. Over time, the lesion may develop striking hyperkeratosis, as in our patient. This entity remains under-recognized and is frequently clinically misdiagnosed. It should be included in the differential diagnosis for angiokeratoma, lymphatic malformation, capillary-lymphatic malformation, and verrucous epidermal nevus.

Potential conflicts of interest

The authors declare no conflicts of interests/[the following potential conflicts].

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