A case report and brief review of the literature of an unusual vascular malformation: linear verrucous hemangioma

Shilpa Y Krishnegowda, MD
Sudhir Kumar, MD

Adichunchanagiri Institute of Medical Sciences, Affiliated to Rajiv Gandhi Institute of Medical Sciences, Karnataka, India

Corresponding Author:
Sudhir Kumar N, MD
Department of Dermatology, Adichunchanagiri Institute of Medical Sciences, B G Nagar, Karnataka – 571448, India
Email: 72sudhirkumar@gmail.com

Conflict of Interest: None to declare

Received: 30 November 2014
Accepted: 25 January 2015

Verrucous hemangiomas (VH) are uncommon, congenital vascular malformations of the cutaneous and subcutaneous tissues that spare the deeper subfacial structures. VH usually presents at birth and slowly grows with age. It usually presents as hyperkeratotic, scattered, bluish-red plaques or nodules of various sizes. Although the lesions mostly have a linear pattern, serpiginous and reticulate forms can occur that are unusual. We present a case of a 27-year-old male with linear angiomatous lesions over his right leg.

Keywords: verrucous hemangioma, vascular malformations, angiokeratoms, excision

Iran J Dermatol 2015; 18: 65-68

INTRODUCTION

Verrucous hemangioma (VH) is an uncommon, localized vascular malformation of the cutaneous and subcutaneous tissues that spare the deeper subfacial structures [1]. The lesions are usually seen as soft, compressible, bluish-red, partly confluent masses with a unilateral distribution but they can also infrequently occur bilaterally [2,3]. VH is more commonly misdiagnosed as angiokeratoma clinically but shows distinctive features on histopathology. Though linear and serpiginous can occur, these are unusual [4]. We report a case showing linear arrangement with multiple plaques and nodules.

CASE REPORT

A 27-year-old man presented with asymptomatic swellings over his right leg. Present since birth, these lesions slowly increased in size and number with age and became verrucous. Occasional episodes of pain and bleeding were seen. Physical examination revealed multiple, well circumscribed, soft to firm, purple-red to brownish black plaques, ranging from 0.5 to 5 centimetres present over the extensor aspect of the right leg and foot. The lesions had a linear configuration showing partial compressibility and no attachment to deeper structures. The lesions were non-blanchable and non-pulsatile. In the vicinity of large plaques, smaller satellite nodules were seen. On the right side, the inguinal group of lymph nodes were enlarged. No disparity was noted in size or length of both limbs. Systemic examination did not reveal any abnormalities, nor did laboratory investigations, Mantoux test and chest x ray. Clinically, angiokeratoma, verrucous hemangioma, chromoblastomycosis, sporotrichosis and lupus vulgaris were considered.

A deep biopsy showed hyperkeratosis, papillomatosis and acanthosis in the epidermis. The dermis had dilated blood vessels, with occasional blood lakes, extending to the reticular dermis and subcutaneous tissue. No thrombi were observed. A diagnosis of verrucous haemangioma was made.

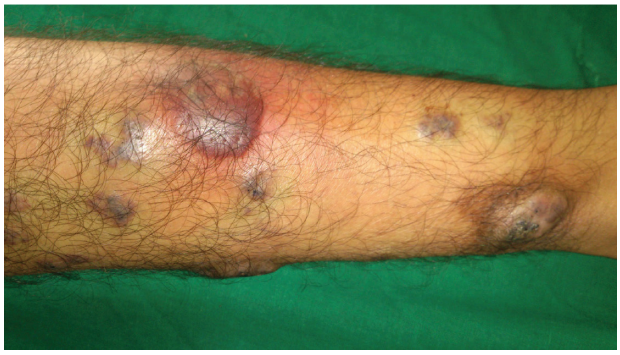

Figure 1. Multiple reddish- brown plaques arranged linearly over the right leg. Note the satellite lesions surrounding the erythematous plaque.

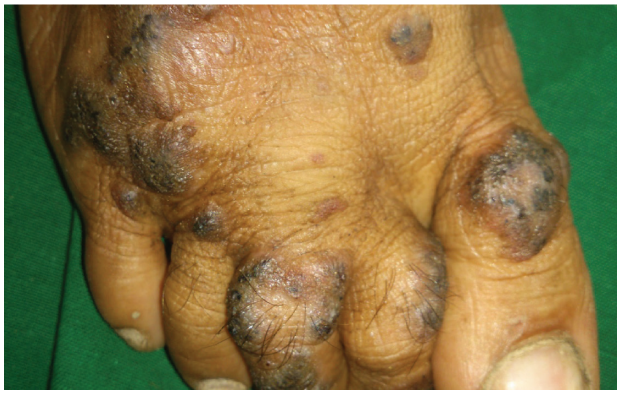

Figure 2. Multiple brownish black, hyperkeratotic plaques present over the right foot.

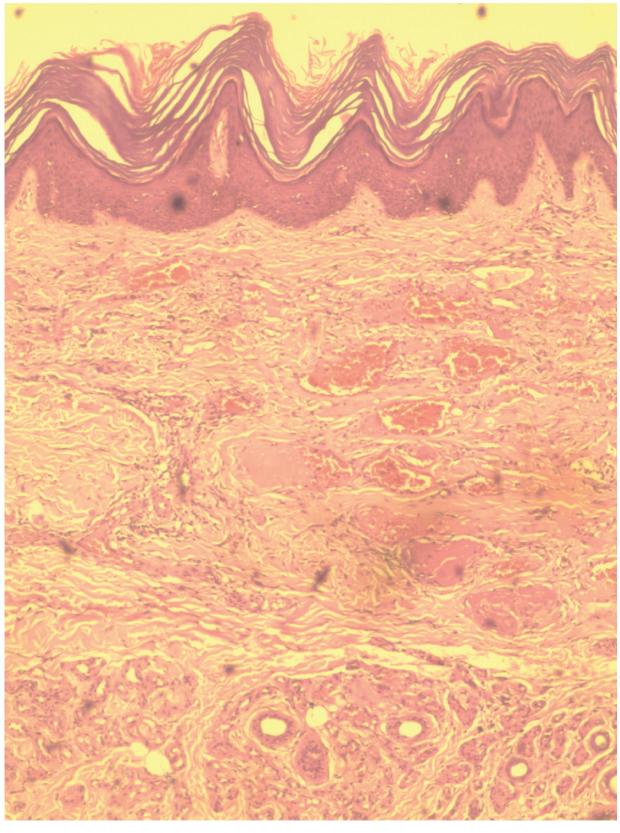

Figure 3. Histopathology of the lesions shows hyperkeratosis, papillomatosis, acanthosis, and dilated blood vessels in the dermis extending into the subcutaneous tissue (H & E, × 100).

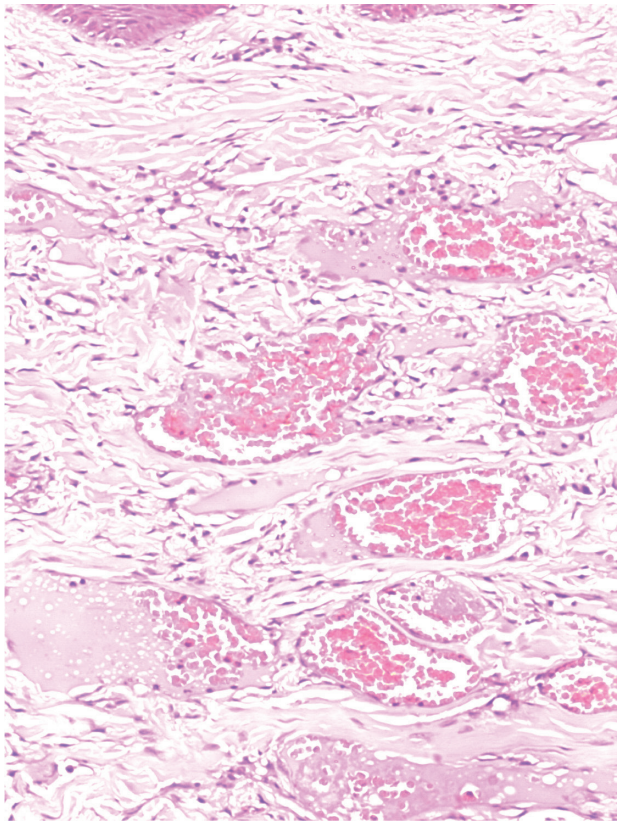

Figure 4. Verrucous hemangioma: ectatic blood vessels seen in the upper and lower dermis. (H & E, × 400).

DISCUSSION

In the past, verrucous hemangioma was known by a variety of names like hemangioma unilateralis neviforme, angiokeratoma circumscriptum neviforme, unilateral verrucous hemangioma, nevus vascularis unius lateralis, nevus angiokeratoticus, nevus keratoangiomatosus, keratotic hemangioma and papulous angiokeratoma [2]. Although Halter (1937) was the first to use the term "verrucous hemangioma", it was described as a separate entity by distinguishing from its mimic, angiokeratoma, and its variants by Imperial and Helwig in 1967 [1].

VH is usually present at birth or early childhood and increases in size with age. The lesions appear as bluish-red confluent nodules of various sizes with typical satellite lesions. With time, complications like secondary bleeding and infection occur,

giving a verrucous, hyperkeratotic surface and a characteristic bluish black colour to the lesions [5]. The lesions are usually scattered but may also be arranged in a linear, serpiginous, or reticular pattern [6]. The linear arrangement of some lesions may reflect genetic mosaicism [1] or dermatomal distribution [6]. VH is typically seen over the lower extremities. However, unusual anatomical locations include abdomen [6], upper limb [1], scalp [7] and glans penis [8]. Two variants have been identified, a disseminated variant with no evidence of systemic lesions and a digital variant, digital verrucous fibroangioma, characterized by dome shaped nodules on the dorsal aspect of the fingers [1,2].

Histologically, VH consists of a non-hemangiomatous and a hemangiomatous component. While the former characterizes the epidermis with hyperkeratosis, papillomatosis, irregular acanthosis and elongated rete pegs, the latter occupies the entire dermis and subcutaneous tissue with dilated capillary and cavernous blood-filled spaces with occasional thrombosis [2] This warrants for a deeper excision to prevent persistence of the lesion.

Differential diagnoses include angiokeratoma, angiokeratoma circumscriptum, hemangioma, Cobb syndrome, angioma serpiginosum, sporotrichosis, lupus vulgaris, chromoblastomycosis, cutaneous keratotic hemangioma, blue rubber bleb nevus, verrucae and vascular tumors. Angiokeratoma is telangiectatic without the deep angiomatous component of verrucous hemangioma [2]. Angiokeratoma circumscriptum is an acquired condition with small 1 to 5 mm papules unlike VH which is a true congenital vascular malformation, which presents with larger plaques, nodules and satellite lesions [6]. Cobb syndrome, a congenital vascular nevus, has a dermatomal distribution mainly over the trunk and is associated with meningospinal angioma which may lead to distal flaccid paraplegia [2]. Angioma serpiginosum lacks the hyperkeratotic surface on serpiginously distributed small violaceous papules. The localized lymphatic variant of sporotrichosis, a fungal infection, usually prefers the upper limbs and histology shows a granulomatous reaction with neutrophilic foci and asteroid bodies. Lupus vulgaris, a chronic paucibacillary form of tuberculosis, usually presents as a solitary lesion with apple jelly resemblance on diascopy. Chromoblastomycosis, a chronic infection caused by pigmented fungi, usually presents as a solitary plaque over exposed parts of the body. Cutaneous keratotic hemangioma, although histologically similar to VH, is an acquired vascular tumour mainly seen on the volar side of the fingers. Blue rubber bleb nevus is a venous malformation which is not only limited to the skin but also involves the gastrointestinal tract and mucous membranes.

Diagnosis is mainly by histopathology. Clinically, a slow spread with age, no tendency for spontaneous resolution, satellite lesions, and frequent recurrences should arouse a high suspicion for VH. Currently, no specific immunohistochemical markers are available for VH. In one study, positivity for glucose transporter protein 1(GLUT1), a determinant expressed by infantile hemangioma, was seen in 7 of 11 VH lesions [9]. Immunotyping for lymphatic specific endothelial markers like vascular endothelial growth factor -3, podoplanin and Prox1 may be used for categorizing vascular anomalies and aiding in classification and diagnosis [10]. These markers are expressed in lymphatic endothelial cells under normal conditions and may also be ectopically expressed in vascular tumors. Wilms tumor 1 (WT1), a transcription factor, may help to distinguish vascular tumors from malformations [10]. Magnetic resonance imaging shows low-flow angiomas in the skin and sub-cutaneous tissue [1].

VH neither regresses spontaneously nor responds to usual modes of physical therapy like electrocoagulation, cryotherapy, and laser. Smaller lesions can be treated with excision and electrocautery while larger lesions, greater than 2cm, require wide deep excision. Symptomatic measures should be undertaken for lesions which are surgically non–accessible [2].

REFERENCES

1. Wentscher U, Happle R. Linear verrucous hemangioma. J Am Acad Dermatol 2000; 42: 516.
2. Calduch L, Ortega C, Navarro V, Martinez E, Molina I, Jorda E. Verrucous hemangioma: report of two cases and review of the literature. Pediatr Dermatol 2000; 17: 213–7.
3. Cruces MJ, de la Torre C. Multiple eruptive verrucous hemangiomas: a variant of multiple hemangiomatosis. Dermatologica 1985;171:106-11.
4. Klein JA, Barr RJ. Verrucous hemangioma. Pediatr Dermatol 1985;2:191-3.
5. Wang G, Li C, Gao T. Verrucous hemangioma. Int J

Dermatol 2004;43:745-6.

6. Hayashi H, Shimizu T, Nakamura H, Shimizu H. Linear verrucous haemangioma on the abdomen. Acta Dermatol-Venereol 2004; 84: 79.
7. Kulkarni AA, Abhyankar SV, Singh RR, Bhatia SH. A rare case of large verrucous hemangioma on the scalp. Med J DY Patil Univ 2012;5:60-1.
8. Akyol I, Jayanthi VR, Luquette MH. Verrucous hemangioma of the glans penis. Urology 2008; 72: 230. e15.
9. Tennant LB, Mulliken JB, Perez-Atayde AR, Kozakewich HP. Verrucous hemangioma revisited. Pediatr Dermatol 2006; 23: 208.
10. Clairwood MQ, Bruckner AL, Dadras SS. Verrucous hemangioma: a report of two cases and review of the literature. J Cutan Pathol 2011;38:740-6.

