Verrucous lesions on right lower limb

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CLINICAL FINDINGS

A 38-year-old male presented to the dermatology out patient department with the complaint of occasionally itchy, irregularly shaped, reddish plaques with multiple warty lesions arranged in a linear pattern affecting the back of right thigh. The main plaque started almost 30 years ago, and then progressively enlarged to the present size and also developed hard warty grayish lesions over many years. He was diagnosed clinically as a case of psoriasis many years ago, but had not responded to topical and systemic anti-psoriatic medications. Cryotherapy was tried but with minimal and transient improvement. The patient did not complain of any systemic illness and there was no family history of similar lesions.

Physical examination showed, a large, well-demarcated, erythematous and edematous plaque measuring approximately 25x10 cm on the posterior aspect of the right lower limb encroaching on its anterior region (Fig. 1, 2). The lesion was studded with multiple verrucous lesions along with some small discrete plaques noticed around the main lesion in a linear arrangement. Surface of the lesion was firm and verrucous with no ulceration, bleeding, or atrophy. There was no tenderness or pain on palpation. Nails and mucous membranes were not affected and showed no significant abnormalities. No regional lymphadenopathy was present. The review of other systems was unremarkable. Routine laboratory investigations including complete blood count, blood sugar, hepatic and renal profile revealed no abnormal findings. The X-ray examination of the lower limb showed no extension of the lesion to underlying bones.

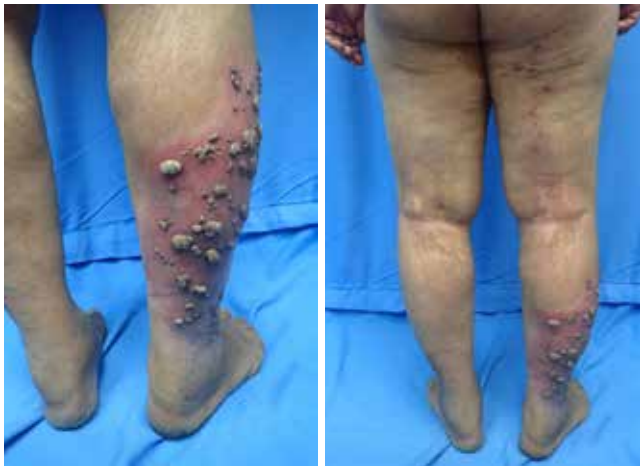

Fig. 1 & 2 A large, well-demarcated, erythematous and edematous plaque measuring approximately 25 x 10 cm studded with multiple verrucous lesions along with some small discrete plaques around the main lesion in a linear pattern

What is your clinical differential diagnosis?

Verrucous hemangioma
Angiokeratoma
Linear epidermal verrucous nevus
Inflammatory Linear Verrucous Epidermal Nevus
Klipple Trenauny syndrome

A skin biopsy revealed hyperkeratosis, parakeratosis, acanthosis, and papillomatosis of the epidermis. Underlying papillary dermis

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showed multiple thin-walled and ectatic blood filled spaces with mild perivascular mononuclear cell inflammatory infiltrate. Similar vascular channels were also seen in the mid dermis and subcutaneous tissue (Fig. 3, 4).

Magnetic resonance imaging (MRI) of the left lower limb showed cutaneous and subcutaneous angiomatosis representing low-flow angiomas.

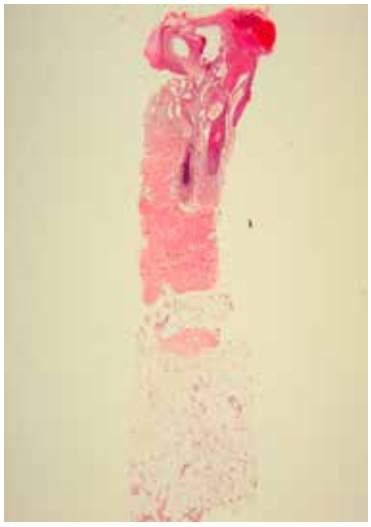

Fig. 3 Hyperkeratosis, acanthosis and elongated curved rete ridges. The dermis and subcutis show dilated vascular spaces

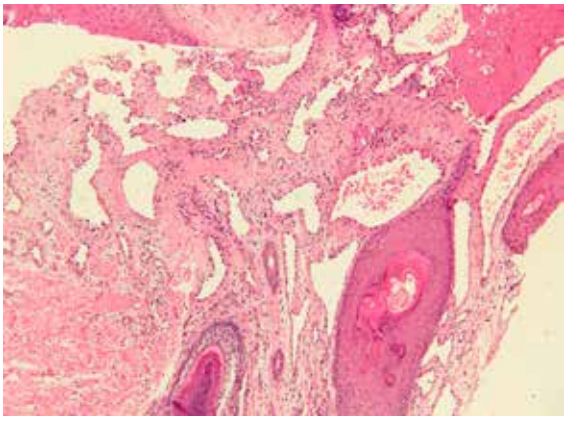

Fig. 4 Dilated vascular spaces lined with plumb endothelial cells

The bones and the muscles appeared to be uninvolved

FINAL DIAGNOSIS

Linear Verrucous Hemangioma

COMMENT

Verrucous hemangioma is an uncommon, congenital vascular malformation associated with reactive skin changes such as hyperkeratosis and papillomatosis, first denominated by Imperial and Helwig in 1967.[1] The International Society for the Study of Vascular Anomalies (ISSVA) differentiates vascular anomalies into vascular tumors and vascular malformations. Vascular tumors, such as infantile hemangiomas, result from exacerbated cell proliferation, are more prevalent in females, tend to regress with the child's growth, and present positive immunohistochemistry for WT1 (Wilms tumor 1 protein) and GLUT1 (glucose transporter-1 protein). On the other hand, vascular malformations consist of errors in vessel morphogenesis, with equal male/female prevalence, that grow proportionally with the child, and that do not display involution or positive IHC for WT1 or GLUT1.[2]

The exact incidence is difficult to determine as it has been referred to by many different names in the past.[3] They can be present since childhood or appear later in life during adulthood and are usually found on the lower extremities, being unilateral in approximately 95% of cases.[4] Bilateral presentation was seen in one case only.[5] A few atypical clinical presentations of verrucous hemangioma have been reported in the literature, Wentscher and Happle reported a 16-year-old male patient having multiple angiokeratotic lesions arranged in a linear pattern on his left arm.[6] Akyol et al. reported a 5-year-old boy with verrucous hemangioma in the glans penis and discussed briefly the hyperkeratotic vascular stains, which constitute a heterogenous group.[7]

Verrucous hemangiomas are often detected by histopathology, although they can also have a

distinct physical appearance. They might begin as a non-keratotic, bluish-red lesion resembling port-wine stain that progressively enlarge and become more hyperkeratotic and verrucous, giving the configuration of warty papules, plaques or nodules arranged in linear or serpiginous pattern, particularly after infection or subsequent trauma. The variation in size is widely documented, and they frequently form satellite lesions.[8]

In our case, the lesions involved the left lower limb and showed a linear arrangement of erythematous plaques with a characteristic verrucous feature. It is not confirmed whether this distribution corresponds to the lines of Blaschko, because the number of reported cases of linear verrucous hemangioma has been rather small.[9] Some authors prefer the idea of a dermatomal arrangement of vascular lesions such as angiomas or telangiectatic nevi.[10]

Histologically, verrucous hemangioma appears initially as a capillary hemangioma concentrated in the dermis and the hypodermis that later, after an intense proliferative reaction of the epidermis, adopts a verruciform pattern with hyperkeratosis, papillomatosis, irregular acanthosis, and an underlying capillary, cavernous, or mixed hemangioma in the dermis and subcutaneous tissue. The hemangiomatous component is greatly composed of dilated capillary and larger cavernous, endothelial-lined, blood-filled spaces. Inflammatory cells, hemosiderin, and fibrosis may be present in the upper dermis.[11] Verrucous hemangiomas must be differentiated from angiokeratomas that do not extend as deep into the reticular dermis and subcutaneous adipose tissue as verrucous hemangiomas. This deeper extension has implications in the surgical resection of the lesions, which must extend deeper than when excising angiokeratomas.[12] Also, in angiokeratoma circumscriptum neviforme, the epidermal findings of acanthosis with hyperkeratosis, often even hypergranulosis are very evident and the vascular alterations are limited to the papillary dermis.[13]

Recently identified genetic mutation in the Ang1-TIE2 (angiopoietin 1-tunica internal endothelial cell kinase) pathway confirms that verrucous hemangioma has genetic features consistent with a venous malformation, despite having an immunoprofile similar to vascular neoplasms.[14] Another especially important complementary test is the scanogram, which measures the limbs comparatively. In the presence of asymmetry, Klippel-Trenaunay syndrome (KTS) should be considered.[13]

Early diagnosis and intervention are very important in patients with verrucous hemangiomas, for a better cosmetic outcome. VH's do not resolve spontaneously and have a tendency to relapse. Surgery is the mainstay of treatment; resection should encompass the deep portions of the lesion with usually a 1-cm margin of excision to avoid the risk of recurrence. If the lesion is small (<2 cm), cryosurgery, electrocautery, sclerotherapy or laser therapy can be tried. These additional therapies can be used in combination with resection for extensive lesions to further assist in reducing the risk of recurrence.[15] Yang and Ohara reported that surgical excision in combination with pulse dye laser, CO_2 laser, or argon laser is the preferred treatment for verrucous hemangioma and is necessary for large and extensive lesions.[16] In our case, we referred him to the Plastic Surgery Department. The lesion was totally excised. No recurrence has been observed during 6 months′ follow-up period after excision.

The prognosis for verrucous hemangioma is

Table 1: Clinicopathological challenges of verrucous hemangioma.

Diagnosis	Clinical	Pathological
Linear epidermal verrucous nevus	• It appears since birth as linear brownish verrucous lesion commonly on limbs and trunk. It may be unilateral or bilateral	• Hyper keratatosis, acanthosis and papaillomatosis with increased melanin pigmentation
ILVEN	• It looks like linear epidermal nevus with background of erythema, sometimes itchy	• Like pathology of linear epidermal verrucous nevus in addition to parakeratosis and mild to dense superficial perivascular inflammatory infiltrate formed of lymphohistiocytic cells
Klipple Trenauny syndrome	• Formed of enlarged involved limb, nevus flameus, varicose vein	• Dilated vascular channels lined with bland endothelial cells in upper dermis
Chromoblastomycosis	• It appear as keratotic verrucous lesions mainly on lower limb	• Epdermal hyperplasia and neutrophilic suppration containing medlar bodies.
Angiokeratoma	• Keratotic and verrucous purplish nodule in pubic region and lower limb	• Epdermal hyperplasia and ectactatic vascular spaces in papiilary dermis

favorable, with recurrence being minimal when proper surgical margins are utilized. Recurrence rates might exceed 30% if a wide excision is not completed properly.[12]

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