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Dermatology

Differentiating Between Verrucous Hemangioma and Angio Keratoma Circumscriptum Neviforme: Two Rare Vascular Malformations

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Abstract **Case Report**

Verrucous hemangioma is a rare congenital vascular skin disorder characterized by keratotic hemangiomatous lesions. Clinically presents as red macules since birth, generally over lower limbs which over the course of time transforms into hyperpigmented, hyperkeratotic verrucous plaques [1]. Its main differential diagnosis is angiokeratoma circumscriptum neviforme, with an almost similar clinical presentation and site predilection as verrucous hemangioma. Histopathology plays a crucial role in differentiating both the disorders. We hereby report a case of 18 year old boy, with angiokeratoma circumscriptum neviforme, diagnosed with the help of histopathology.

Keywords: Verrucous hemangioma, angiokeratoma circumscriptum, vascular malformations.

INTRODUCTION

First described by Imperial and Helwing in 1967, Verrucous Hemangiomas present as rare, congenital keratotic hemangiomatous disorder which present as a reddish macular area similar to the "port-wine" stain since birth or may appear later in life during adulthood and are usually found on the lower extremities[2]. With time macules transform into warty, bluish, vascular papules, plaques, or nodules which may be single or grouped, linear or serpiginous arrangement [3].

Angiokeratoma Circumscriptum is also a rare malformation of the blood vessels presents as congenital or acquired, multiple, red to violaceous, punctate macules ,which later on may become hyperkeratotic and appear black if thrombosed or traumatized, usually located in the lower extremities found more commonly in females. They may occur in clusters or they enlarge peripherally and fade at the center giving rise to a serpiginous pattern. The lesions are benign, and generally resolve spontaneously [4].

Since both the diseases are clinically indistinguishable, the onus of diagnosis lies on histopathology. A deep biopsy is necessary to confirm the clinical diagnosis by histopathological examination, so as to evaluate the precise vascular structure involved.

Histologically, verrucous hemangioma characterized by hyperkeratosis, papillomatosis, irregular acanthosis with the dilated capillaries and vessels in dermis and subcutis organized in a diffuse or lobular structure [5].

Angiokeratoma Circumscriptum is characterized by hyperkeratosis, papillomatosis, and irregular acanthosis with the clusters of ectatic capillaries in the papillary dermis [6].

CASE REPORT

An 18 year old boy presented to dermatology OPD with multiple linearly arranged, hyperpigmented verrucous plaques interspersed over an erythematous base present over right leg. The lesions were relatively asymptomatic with mild itching sometimes, and did not bleed on trauma.

Patient stated that the condition started at 3 years of age, where a red macule first appeared over his right great toe and over a course of 4 years it grew in size and transformed into verrucous lesion spreading up the leg in a linear fashion (fig. 1). At times the verrucous plaques get secondarily infected and ulcerate for which the patient takes course of oral and topical antibiotics, and gets temporary relief, however the verrucous plaques kept on growing with Age without any regression of previous lesions.

Patient took multiple sessions of radiofrequency ablation previously, following which the verrucous plaques shed off from the area treated temporarily, only to grow back. Systemic examinations, laboratory investigations and Color Doppler of right lower limb came out to be normal.

A deep excisional biopsy was taken from the verrucous plaque and sent for histopathology examination which revealed hyperkeratosis, acanthosis and extensive numerous dilated capillary spaces in epidermis and papillary dermis.

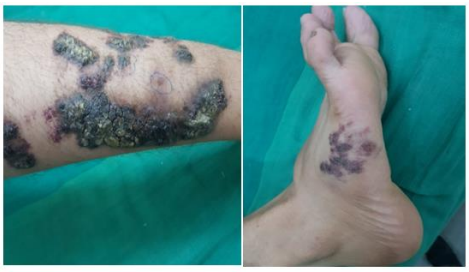

Fig-1: Images showing multiple well defined hyperpigmented verrucous plaque of size 1cm in smallest diameter upto 10 cm in largest diameter distributed over lateral side of right foot and shin of right leg.

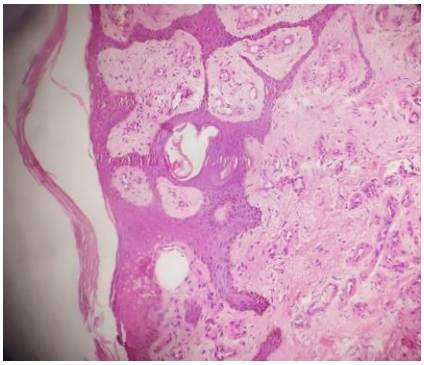

Fig-2: Showing hyperkeratosis, acanthosis and extensive numerous dilated capillary spaces in epidermis and papillary dermis

DISCUSSION

Verrucous hemangiomas are rare variety of angiomatous nevi generally involving lower limbs and in 95 % cases present unilaterally [7].

Verrucous Hemangioma presents at birth or in early infancy without any sex predilection, whereas Angiokeratoma circumscriptum has a female predilection. Both conditions Clinically present as erythematous patches that evolve to form violaceous plaques, becoming scaly and even verrucous, most commonly affecting the lower limbs , which in case of Verrucous Hemangioma progresses with growth of child while in Angiokeratoma circumscriptum spontaneous regression is noted in few cases[8].

On Histopathology examination- In Verrucous Hemangioma hyperkeratosis, irregular acanthosis, hypergranulosis, papillomatosis with vascular proliferation extending upto the deep dermis and subcutaneous tissue, In Angiokeratoma circumscriptum the epidermal findings of hyperkeratosis, acanthosis, hypergranulosis are present with papillomatosis and the vascular alterations limited to the papillary dermis only[5, 6].

On the basis of clinical examination and histopathological findings, a diagnosis of Angiokeratoma circumscriptum is made.

Moreover immunohistochemistry in Verrucous Hemangioma shows positivity to GLUT1 (glucose transporter-1) and WT1 (wilms tumor-1 protein), both of which are expressed in vascular tumors. WT1 gene is a tumor suppressor gene that affects angiogenesis by modulating vascular endothelial growth factor (VEGF), if defective [9].

Management include Cryotherapy, electrocoagulation, curettage, CO_2 laser (deep) because of the deep angiomatous proliferation, surgical resection with adequate margins, recently a combination of CO_2 and dual pulsed dye laser Nd:YAG has shown satisfactory response in some cases.

Here, we report a case of 18-year-old male, with Angiokeratoma circumscriptum which is linearly distributed over his foot and lower leg, since 3 years of age, presenting as the keratotic angiomatous lesions with histopathology findings of hyperkeratosis, hypergranulosis, and papillomatosis with vascular alterations limited to the papillary dermis.

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