

Angioma serpiginosum in a bilateral distribution with acral involvement: An uncommon presentation

Sir,

Angioma serpiginosum is a vascular nevus of superficial capillaries characterized by minute red to copper-colored macules in clusters or in a serpiginous pattern,¹ resulting from congenital hyperplasia or ectasia of preexisting superficial dermal capillaries. The condition usually arises before 20 years of age and thereafter it reaches a state of relative stability.² Herein, we report a case of angioma serpiginosum with characteristic clinical presentation involving both upper limbs, trunk, palms and nails.

An 18-year-old man, born out of nonconsanguineous parentage, second in order of birth, presented to our outpatient Department (OPD) with red-colored lesions on both upper limbs, chest and upper back for the last 8 years. Initially there was a single lesion on the left arm which subsequently progressed in size and number to involve both the upper limbs and chest. He reported a rapid progression in the last 3 years. There was no history of any systemic complaints. Dermatological examination revealed pin-point to pin-head-sized erythematous discrete to coalescent macules in a serpiginous pattern distributed over both upper limbs, chest [Figure 1a], upper trunk and palms [Figure 1b-d]. Erythema was also noticed on the middle finger nail of left hand [Figure 1e]. On diascopy, the lesions were nonblanchable. No other part of the body was involved including the lower limbs. Hair and mucosae were spared. Systemic examination was within normal limits. A differential diagnoses of angioma serpiginosum and bilateral nevoid telangiectasia was made. Dermoscopy of the lesions on the arm and chest showed characteristic oval to round red lagoons, consistent with angioma serpiginosum [Figure 2a]. Dermoscopy of the palmar lesions showed erythematous parallel ridged pattern

with red globules and dots arranged linearly without affecting the acrosyringial openings [Figure 2b]. Onychoscopy of the left-hand middle fingernail showed a diffuse erythema. Histopathology revealed presence of dilated and proliferating capillaries in the superficial papillary dermis with no evidence of extravasation of erythrocytes [Figure 2c]. Rest of the hematological, biochemical, radiological and systemic workup was noncontributory. On the basis of clinical, dermoscopic and histopathology features, a final diagnosis of angioma serpiginosum was made.

Angioma serpiginosum is an uncommon cutaneous vascular nevus first reported by Jonathan Hutchinson in 1890, while Frain-Bell distinguished this entity from pigmentary purpura and other disorders in 1957.¹ A female preponderance has been reported and it mainly affects lower extremities and buttocks, although involvement of upper extremities has been reported, as in our case.^{2,3} It predominantly has a unilateral distribution and usually involves the trunk, palms and mucosae.³

Bilateral nevoid telangiectasia, on the other hand, is an acquired condition usually seen in hyperestrogenic states and liver diseases that presents clinically with 'spider' telangiectasia.^{4,5} Distribution is bilaterally symmetrical and it can be associated with gastrointestinal telangiectasia, presenting as unexplained gastrointestinal bleeding.⁴ Association with Hepatitis C infection and cholangiocarcinoma has also been reported.⁴

Histopathology of angioma serpiginosum shows dilated and tortuous capillaries in the papillary dermis without any evidence of inflammation or red cell extravasation, whereas bilateral

nevus telangiectasia demonstrates superficial papillary dermal telangiectasia with minimal chronic inflammation.^{3,5} Dermoscopy of angioma serpiginosum shows characteristic well-demarcated oval red lagoons.⁶ Acral variant on dermoscopy demonstrates erythematous parallel ridge pattern with red globules and dots in a linear arrangement, with sparing of the acrosyringial openings.^{7,8} Dermoscopy of unilateral nevus telangiectasia, on the other hand, shows thin and tortuous linear vessels with absence of red lagoons.⁹

Both dermoscopic and histopathologic findings in our case were consistent with the diagnosis of angioma serpiginosum.

Etiology of this condition remains obscure. Role of estrogen has been suggested in the etiopathogenesis of both angioma serpiginosum and nevus telangiectasia.^{2,5} The proliferative effect of estrogen on the vascular endothelial cells support this hypothesis.² However, absence of estrogen and progesterone receptors on the involved blood vessels in angioma serpiginosum has contradicted this theory.¹⁰ Other than hormonal theory, abnormal response of capillaries to cold has also been proposed as a possible etiology.² This condition remains stable or partially regresses after puberty.

Treatment with pulse dye laser has shown promising results in some reports.^{2,6} Our patient has also been counselled regarding the therapeutic option of pulse dye laser.

Our case presented with an unusual bilateral asymmetrical distribution involving both upper extremities, trunk and palms, whereas this condition is predominantly unilateral. Another unique feature of our case was possible involvement of the nail bed that manifested as nonblanching erythema in contiguity with palmar lesions. Onychoscopy showed diffuse erythema of nail bed. The patient, however, refused nail biopsy. To the best of our knowledge,



Figure 1a: Pin-point to pin-head erythematous discrete to coalescent macules present on the right upper limb with involvement of the chest



Figure 1b: Pin-point to pin-head erythematous discrete to coalescent macules present on the left upper limb with involvement of the trunk



Figure 1c: Close-up view of the dusky erythematous macules



Figure 1d: Involvement of the palms with erythematous macules



Figure 1e: Erythematous macules extending to the dorsa of the hands. Note the erythema present in the middle finger nail bed

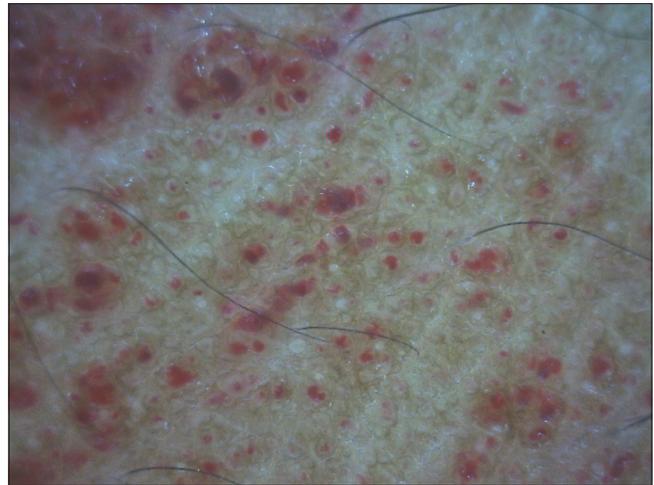


Figure 2a: Dermoscopy showing the characteristic red lagoons (Dinolite AM413ZT; ×150)

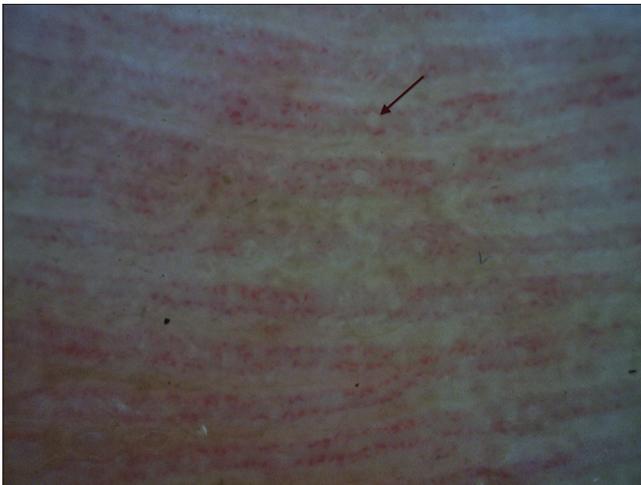


Figure 2b: Dermoscopy of palm showing red globules and dots spreading in a linear arrangement in a parallel ridge pattern without affecting the acrosyringeal openings as shown with red arrow (Dinolite AM413ZT; ×150)

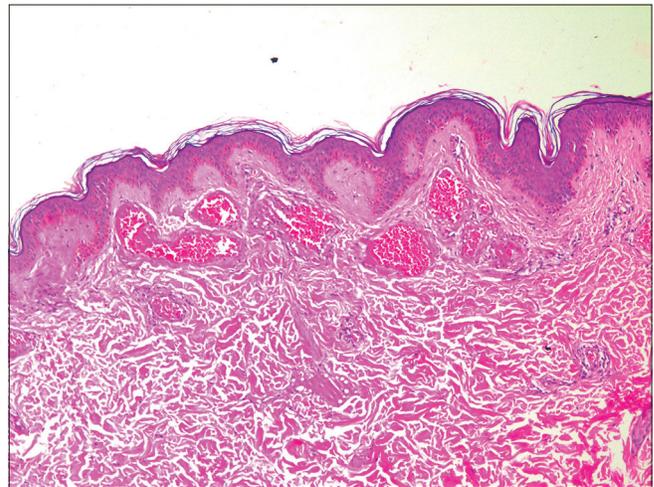


Figure 2c: Dilated and proliferating capillaries in the superficial papillary dermis without any evidence of inflammation (hematoxylin and eosin, ×400)

this unique distribution of angioma serpiginosum has not been reported yet in the literature.

The rarity of our case and the role of noninvasive dermoscopy in differentiating angioma serpiginosum from bilateral nevoid telangiectasia has prompted us to report this case. This distinction is especially pertinent as the latter is more likely to be associated with underlying pathologies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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