Case 20

A 29-year-old woman from Suratthani

Chief complaint: Asymptomatic reddish rashes on both arms since childhood

Present illness: The patient gradually developed asymptomatic reddish rashes on both arms since childhood. The rashes were slowly progressive during the first few years with no regression. She had no systemic symptoms, no history of cold exposure, and external hormonal use.

Past history: She denies of any underlying disease.

Personal history: She denies history of smoking and drinking.

Family history: No member of her family had similar lesions.

Physical examination:

Skin examination:

Bilaterally multiple groups of punctate erythematous macules confluent to reticulate pattern on the upper arms. Diascopy was negative.

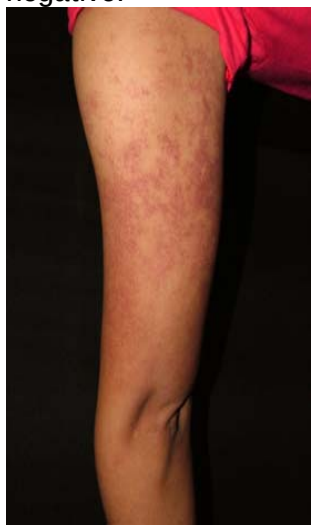

Fig. 20.1

Fig. 20.2

Histopathology (S08-17783) (Fig 20.**3**, 20.4)

- A proliferation of small blood vessels lined by oval shaped endothelial cells, in the upper dermis
- Normal epidermis

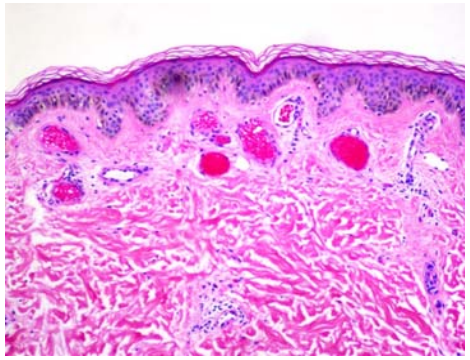

Fig. 20.3

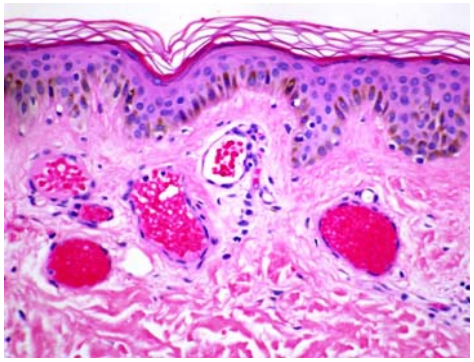

Fig. 20.4

Diagnosis: Angioma serpiginosum

Presenter: Piyavadee Singvahanont

Consultant: Vasanop Vachiramon

Discussion:

Angioma serpiginosum (AS) is a rare, benign, acquired vascular disorder. It classically begins in childhood and is more common in females (90%). Most cases are sporadic, but autosomal dominant inheritance has also been reported.[1]

The pathogenesis is unknown. Increased levels of estrogen may play a role in the development of the lesions because most cases arise in girls under 16 years old and progress rapidly during pregnancy. This progression may be attributed to the ability of estrogen to enhance the proliferation of hemangioma vascular endothelial cells.[2] In addition, Neumann suggested that the lesions

may represent an abnormal vascular response to cold. Dermal vessel damage due to cold exposure, possible in association with other yet unknown factors, may cause the formation of new capillaries that subsequently aggregate to form larger ecstatic vessels.[3,4]

The lesions are characterized by asymptomatic, nonblanchable, minute, punctate, purple-colored to bright-red macules usually arranged in a gyrate or serpiginous pattern. Linear lesions and distribution along Blaschko's lines have been reported very rarely.[5,6] The lesions may affect any region of the body, often unilaterally on the lower limbs and buttocks, but it may be more extensive involvement of the trunk and extremities[7], which can be associated with capillary abnormalities of the retina and spinal nerve root.[8]

Histopathologically, AS shows dilated, thin-walled capillaries are seen in some of the dermal papillae and the superficial reticular dermis. Epidermal changes and extravasation of red blood cells does not occur. Epiluminescence microscopy can be helpful in diagnosis by demonstrating "red lagoons".[9]

Differential diagnosis includes angiokeratoma corporis diffusum, unilateral nevoid telangiectasia, port-wine stain and linear pigmented purpura. All these diseases, in addition to AS, have dilated capillaries in the dermis, except pigmented
purpura. However, there are a few clinical and pathological features that can differentiate these entities from each other. Angiokeratoma corporis diffusum and pigmented purpura have characteristic pathological findings in terms of the presence of papillomatosis in the former, and hemosiderin deposition, extravasated red cells, and inflammation in the latter. Port-wine stain and unilateral nevoid telangiectasia can be differentiated clinically.

Spontaneous resolution of individual puncta is known to occur; rarely partial or complete regression of the whole lesion has been reported. Effective treatment options include pulsed dye laser [9-11], argon laser [12], long-pulsed KTP-Nd:YAG laser, and the IPL system.[6]

References:

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