Angioma sepiginosus: two cases in children and review of literature

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F.B and G.R. contributed to conceptualization, formal analysis and editing of the study.
O.A., M.E and D.B. contributed to the methodology, formal analysis, and review of the manuscript.
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Statement of Ethics
This research was conducted in compliance with the guidelines for human studies and ethically in accordance with the World Medical Association Declaration of Helsinki. Written informed consent was obtained from the parents prior to any study-related procedures.

Informed consent: Informed consent was obtained from all individual participants included in the study.
Abstract:

Angioma serpiginosum (AS) is a rare benign vascular lesion that typically arises in early childhood, with a prevalence in female, and then grow up over a period of months/years. It is characterized by small asymptomatic purple-red dots that cluster together and they do not disappear on diascopy. It is mainly localized on the arms but some cases on face and neck have been reported. The etiology of AS is unknown, dermoscopy may aid in the diagnosis but usually the biopsy is necessary. We report 2 cases: one male and one female with angioma serpiginosum, aged 13 and 8 years old.

Introduction

Angioma serpiginosum was first described in 1889 by Hutchinson as a particular type of angioma but in 1893 Radcliffe-Croker proposed the specific term of “angioma serpiginosum”. There is a sex ratio of 9:1 female versus male. In the 80% of cases it arises before 20 years old. Probably it develops from the proliferation of endothelial cells with development of new capillaries. Some authors agree that hormones are involved in the pathophysiology. Clinically it appears as multiple, minute, pinpoint, grouped, bright red, no-blanchable macules and irregular patches.

Case reports

We report 2 cases in children: a 13 years old female who had a history of asymptomatic red lesion on her left shoulder blade for 2 years (fig. 1a) without itching, vesiculation neither familial similar disorders and a 8 years old male with erythematous reticulated macules affecting his right arm since birth (fig. 2a). Dermoscopy and skin biopsy were performed to both the patients:

- Dermoscopy revealed well demarcated round red lagoons in relation to dilated vascular spaces within the papillary or superficial reticular dermis, hairpin like vessels scattered among red lagoons. The dermoscopy findings in AS have been described as “school of red fish in a pound” (fig. 1b, 2b).

- Histopathology showed proliferated and dilated capillaries in the superficial papillary dermis, without erythrocyte extravasation or hemosiderin deposits or inflammatory elements. Immunohistochemistry showed positive staining with CD31, CD34, and Wilms tumor-1 (WT-1) and negativity with D2–40 and Glut-1 (fig. 3).
Considering the clinical examination, the dermoscopic and histopathological features the diagnosis of Angioma Serpiginosum (AS) was made in all the four patients and because of the young age of the patients, no treatment has been performed.

**Discussion**

AS is a rare benign vascular lesion, usually sporadic but in some cases an autosomal dominant inheritance pattern has been reported \(^4^,^5\). It is characterized by unilateral, asymptomatic eruption
with multiple, minute, pinpoint, grouped, bright red, no-blanchable macules or figured lesion. Usually may occur anywhere but the most involved sites are the upper and lower extremities. Some authors have reported an association with retinal and spinal angioma.

The differential diagnosis includes:

- pigmented purpura with extravasation of erythrocytes and hemosiderin pigment;
- unilateral nevoid telangiectasia with unilateral distribution (frequently in C3-C4 or the trigeminal area);
- angiookeratoma lesion.

AS is considered a vascular tumor due to endothelial cell proliferation with formation of new capillaries. Another etiological hypothesis is related to an abnormal morphogenesis in the form of capillary walls due to a precipitation of fibrillar structure and collagen fibers. In literature a partial or complete spontaneous regression is described but usually the lesion is slowly progressive lifelong.

**Conclusion**

With our report we want to keep the attention on a rare and misdiagnosed vascular malformation in childhood that need the support of dermatologist and pathologist to confirm the diagnosis. Treatment is recommended only for cosmetic reasons; and the gold standard treatment is considered the pulsed dye laser (585 nm).
Figure Legend

Fig. 1a: A 13 years old female with history of asymptomatic red lesion on her left shoulder blade for 2 years.

Fig. 1b: Dermoscopy revealed well demarcated round red lagoons in relation to dilated vascular spaces within the papillary or superficial reticular dermis, hairpin like vessels scattered among red lagoons (aspect of “school of red fish in a pound”).

Fig. 2a: A 8 years old male had erythematous reticulated macules affecting right arm since birth.

Fig. 2b: Small purple-red dots that cluster together and they do not disappear on diascopy.

Fig. 3: EE - Clusters of dilated capillaries in the upper dermis, composed of flattened endothelial cells and pericytes.
References


