Angioma serpiginosum: Two cases in children and review of literature

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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 discapropy. 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 angiomma but in 1893 Radcliffe-Croker proposed the specific term of “angioma serpiginosum”.¹ There is a sex ratio of 9:1 female versus male. In the 80% of cases it arises before 20 years old.³,⁵ 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 Report

We report 2 cases in children: a 13-year-old female who had a history of asymptomatic red lesion on her left shoulder blade for 2 years (Figure 1a) without itching, vesication neither familial similar disorder and a 8 years old male with erythematous reticulated macules affecting his right arm since birth (Figure 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” (Figure 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 (Figure 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.⁶ The differential diagnosis includes:⁷,⁸

- 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.

Conclusions

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).

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