A Teenage Girl with a Spreading Violaceous Birthmark

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**History**

- A 17-year-old healthy Latina female presented with an asymptomatic slowly enlarging birthmark. A purplish patch had been present over her left chest since birth and over time had gradually spread to involve her left shoulder and arm. She noted that sometimes the lesions will become darker red with paler colored skin at the periphery.
- Her mother stated that she had a normal pregnancy and vaginal delivery with no complications.
- There was no family history of similar findings.

**Examination**

- Over the left chest, shoulder, and ventral arm in a segmental distribution were numerous violaceous, vascular appearing macules coalescing into patches. Some lesions exhibited mild pallor at the periphery (see Figure 2).
- Dermoscopy revealed numerous small, relatively well-demarcated, round to oval red lagoons. The lesions were partially blanchable (see Figure 3).

**Histopathology**

Figure 1. Microscopic exam of a representative lesion from the left arm, displaying dilated and engorged thin-walled vessels in the papillary dermis, along with a mild perivascular lympho-histiocytic infiltrate with rare neutrophils.

Figure 2. Serpiginous pattern of coalescing red macules on the left chest and upper extremity. Area circled in photo at right represents site of biopsy specimen pictured in Figure 1.

Figure 3. Dermoscopic view revealing numerous, small, red, vascular lagoons

**Course and Therapy**

- The initial differential diagnosis included angioma serpiginosum, unilateral nevoid telangiectasia, and acquired port wine stain.
- Given the history, distribution, clinical appearance, and pathology she was given a diagnosis of angioma serpiginosum.
- The patient opted not to pursue treatment as the lesions were asymptomatic and not cosmetically bothersome to her.

**Discussion**

- Angioma serpiginosum (AS) is a rare vascular nevoid disorder of unknown etiology.
- Strong female predominance (9:1) with onset during the first two decades of life. Most cases are sporadic, but autosomal dominant inheritance has been reported.
- Exam characterized by asymptomatic, pinpoint, violaceous to erythematous macules which may become papular. Slow progression in a serpiginous pattern can occur.
- Any area can be affected, but buttocks and extremities are most common. The distribution tends to be unilateral, sparing the palms, soles, and mucous membranes.
- Histologically, AS is characterized by increased numbers of dilated capillaries in the upper dermis.
- In comparison to AS, acquired port wine stain has a smaller degree of vessel proliferation and usually follows a trigeminal nerve distribution.
- Unilateral nevoid telangiectasia (UNT) is difficult to distinguish from AS both clinically and histopathologically. Features favoring UNT include a characteristic dermatomal or Blaschkoid distribution and anemic halos of vasoconstriction around individual telangiectasias. In contrast, the presence of lesions which are only partially blanchable favors AS.
- Management of AS consists primarily of cosmetic treatment of the vascular ectasias with pulsed dye laser.

**References and Acknowledgements**

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