Angioma Serpiginosum in a Patchy and Blaschkoid Distribution: A Rare Condition with an Unconventional Presentation

Karan Sancheti, Anupam Das, Indrashis Podder, Ramesh Chandra Gharami

From the Department of Dermatology, Medical College and Hospital, Kolkata, West Bengal, India.
E-mail: anupamdasdr@gmail.com

Indian J Dermatol 2016;61(5):570-2

Sir,

Angioma serpiginosum, one of the rare causes of telangiectasia, is a rare nevoid disorder affecting the small vessels of the upper dermis, with a characteristic clinical appearance. We report a young female, who presented with multiple progressive lesions scattered over her left side of the chest and upper extremity in a patchy and blaschkoid distribution, which was diagnosed as angioma serpiginosum by histology. A 21-year-old female presented with asymptomatic, progressive, multiple red-colored lesions distributed unilaterally over the left upper extremity and left breast in a patchy and blaschkoid pattern. The lesions started at the age of 10 years over the left breast and rapidly progressed during puberty to involve the left upper extremity. There was no significant history and other family members were not affected. Routine blood biochemistry was within normal limits.
Testosterone and estrogen levels were within normal limits and antibodies to hepatitis B and C were absent.

General examination was unremarkable. On cutaneous examination, we found multiple nonblanchable, grouped, pinpoint, red macules on an erythematous base distributed in a blashkoid pattern over left breast, patchy over medial side of arms and the volar aspect of forearm. Besides, there were patchy, scattered as well as serpiginous lesions over the extensor aspect of arms and forearms [Figure 1]. There was no similar lesion elsewhere in the body. The hair, nail, and mucosa were normal.

In the absence of dermoscopy (which could not be done because of unavailability at our center), a skin biopsy was taken from a representative lesion. Histolopathological examination (HPE) showed dilated, thin-walled capillaries in some of the dermal papillae and the superficial reticular dermis [Figures 2 and 3]. There were neither epidermal changes nor extravasation of red blood cells. Based on clinical appearance and histopathological findings, a diagnosis of angioma serpiginosum was made. The patient was counseled about the benign nature of the disease and referred for pulse dye laser (PDJ).

Angioma serpiginosum, an unusual vascular tumor, was first described by Hutchinson in 1889 as a peculiar form of serpiginous and later coined as “infective” nevus disease in 1894.[1] Subsequently, this disorder was attributed to a loss-of-function mutation involving the PORCN gene on X-chromosome (mild variant of focal dermal hypoplasia) by Hogue et al. in 2008.[6] However, this view has been refuted by Happle.[3] Lesions usually consist of multiple small, asymptomatic, nonpalpable, deep-red to purple puncta occurring in small clusters and sheets. The arrangement and extension of the lesions may produce a serpiginous pattern. The extremities are most commonly affected, usually with a unilateral distribution, though there are reports of disseminated distribution.[3] Truncal involvement is uncommon. The palms, soles, and mucous membranes are usually spared though plantar involvement has been reported.[5] In our case, the lesions were unilateral, distributed over the left chest and left upper limb.

On HPE, dilated capillaries can be seen on the backdrop of erythema, the latter occurring due to dilatation of the subpapillary venous plexus. Epiluminescence microscopy or dermoscopy can be helpful in diagnosis by demonstrating “red lagoons.”[6-7] Histologically, dilated and tortuous capillaries are observed in the papillary dermis but without any inflammation or red cell extravasation. Similar HPE findings were obtained in our case too.

If further observed under an electron microscope, it would be apparent that the thickening of the capillary walls is caused by a heavy precipitate of basement membrane-like material mixed with thin collagen fibers and an increased number of concentrically arranged pericytes. In addition, some of the dilated capillaries may show slit-like protrusions of their lumina and endothelial lining into the surrounding thickened vessel walls. These findings indicate that angioma serpiginosum is not just a simple telangiectasia but represents a vascular malformation. Some authors believe it to be a mosaic skin condition.[8]

Partial or complete spontaneous regression of the lesions may occur. No topical medications have been reported to be effective. PDJ has shown encouraging result in some cases.[9] There have also been reports of successful treatment of the condition with novel 532 nm potassium-titanyl phosphate laser.[10] We have referred our patient to a higher center for management using PDJ.

The differentials considered were unilateral nevoid telangietasia syndrome (clinically follows a dermatomal distribution, particularly those associated with the trigeminal nerve and cranial nerves III and IV), angiokeratoma (epidermis shows variable degrees of acanthosis with elongation of the rete ridges and hyperkeratosis), acquired port-wine nevus (there is usually capillary proliferation along with dilatation), and pigmented purpuric dermatoses (absence of dilated capillaries in the dermis). On the basis of clinical and HPE picture, these differentials were ruled out.

Angioma serpiginosum is a rare vascular anomaly and chest involvement has only been reported recently.[11] Our patient presented with lesions starting over the left side of the chest and extending in a patchy and blashkoid pattern to left arm and forearm. There are only few reports of such case in English literature.[12-14] The rarity of the present case has prompted the current report.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. Hunt SJ, Santa Cruz DJ. Acquired benign and “borderline” vascular lesions. Dermatol Clin 1992;10:97-115.
2. Hogue G, Oeffner F, Grzeschik KH. An Xp11.23 deletion containing PORCN may also cause angioma serpiginosum, a cosmetic skin disease associated with extreme skewing of X-inactivation. Eur J Hum Genet 2008;16:1027-8.
3. Happle R. Angioma serpiginosum is not caused by PORCN mutations. Eur J Hum Genet 2009;17:881-2.
4. Sandhu K, Gupta S. Angioma serpiginosum: Report of two unusual cases. J Eur Acad Dermatol Venereol 2005;19:127-8.
A rare condition with multiple, widespread, nonpruritic, painless skin lesions and systemic involvement (malignant atrophic papulosis and parvovirus B19-induced catastrophic endothelitis). 

Degos Disease: A Murderous Menace

Vishalakshi Viswanath, Jinal Lakhamshi Gada, Ronak Jagdeep Shah

Department of Dermatology, Rajiv Gandhi Medical College and Grant Government Medical College and Sir JJ Group of Hospitals, Chhatrapati Shivaji Maharaj Hospital, Thane, Mumbai, Maharashtra, India.

doi: 10.4103/0019-5154.190114

How to cite this article: Sancheti K, Das A, Podder I, Gharami RC. Angioma serpiginosum in a patchy and blaschkoid distribution: A rare condition with an unconventional presentation. Indian J Dermatol 2016;61:570-2.