Angioma Serpiginosum

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Abstract

Here we describes a 13-year-old girl with asymmetric, vivid red, punctuate eruptions on her left inner thigh for 3 years. Histopathological examination showed multiple dilated capillaries in the papillary dermis and superficial reticular dermis. According to the clinical and pathological findings, we established a diagnosis of Angioma Serpiginosum (AS) a rare, benign, cutaneous vascular disorder usually beginning in childhood.

Keywords: Angioma serpiginosum

Case Report

A 13-year-old girl was referred to our out-patient clinic with a history of asymmetric punctuate eruptions involving her left inner thigh for 3 years. Her condition started as a small, nummular, faintly erythematous patch when she was 10 years old and gradually increased in size. The patient denied any pain, itching, or abnormal sensation related to the lesions. Her past medical history was not remarkable and her family history was negative for the presence of such lesions. Dermatological examination revealed multiple, minute, bright red, punctuate macules on the left inner thigh. Diascopy revealed partial emptying (Figure 1). Skin biopsy was performed and revealed dilated capillaries with endothelial proliferation and a thickening of the capillary walls in the dermal papilla and subpapillary regions of the dermis. No inflammatory changes, hemorrhage, or hemosiderin depositions were present (Figure 2). Correlation of the clinical and histological findings resulted in the diagnosis of angioma serpiginosum.

Discussion

Angioma serpiginosum, a rare vascular disorder, was first described by Hutchinson in 1889 as a peculiar form of serpiginous and "infective" nevoid disease and was named by Crocker in 1894 [1]. Currently, there is a lack of consensus as to whether angioma serpiginosum reflects a type of capillary nevus, a vascular malformation or a vascular neoplasm. Chen et al. [2] recently described a 15-year-old girl with AS presenting in an asymmetric, systematized segmental pattern, reflecting cutaneous mosaicism, and suggested that AS might be best categorized as a vascular nevus. Some authors consider the condition a malformation because of the abnormal morphogenesis in the form of thickened capillary walls [3]. Other authors consider angioma serpiginosum a neoplasm because of endothelial cell proliferation with formation of new capillaries [4].

AS is characterized clinically by minute, punctate, reddish-purple to bright red macules that may be as large as 1 mm, and have a tendency to become papules. They occur in groups, which enlarge through the formation of new lesions at the periphery, while those at the center fade. In this way, small rings or serpiginous and gyrate patterns are formed over the period of months to years [5-6]. No purpura is present, but a netlike or diffuse erythema forms the background [7]. After an initial period of growth before puberty, the lesions usually remain stable throughout adulthood. Spontaneous resolution, although rare and incomplete, may result in partial regression of the lesions. The lesions can affect both genders at all ages, but 90 percent of cases occur in girls under age 16 [7]. The disorder is often arranged in an asymmetric or otherwise segmental pattern [1,5] that may also involve the trunk [6]. The lesions can be located anywhere on the body and have been reported in all areas except the palms and mucocutaneous junctions [5-7]. The areas of predilection are the extremities, especially the lower extremities.

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The pathogenesis of angioma serpiginosum is still unclear yet. A role for estrogen has been implicated [8,9], however, the onset of lesions during childhood, the uniformity of lesions throughout the menstrual cycle, the presence of normal serum sexen levels in affected patients and finally, the absence of estrogen and progesterone receptors around the dilated vessels contradict this theory [10]. In 2007, angioma serpiginosum with esophageal papillomatosis in a 4-generation family was described as a X-linked dominant condition that is mapped to a gene locus on Xp11.3-Xq12 [11]. Subsequently, Houge et al [12] documented that a Xp11.23 deletion containing PORCN may also cause angioma serpiginosum. In addition, an abnormal vascular response to cold has also been proposed in the pathogenesis of angioma serpiginosum [8].

The typical histopathological features of angioma serpiginosum include clusters of dilated capillaries in the papilla and subpapillary regions of the dermis with the absence of inflammatory cell infiltration, hemorrhage and hemosiderin deposition [1]. The overlying epidermis is normal.

Angioma serpiginosum must be differentiated from diagnoses of pigmented purpuric dermatoses, angiokeratoma, unilateral nevoid telangiectasia and acquired port-wine stain. All of these diseases have dilated capillaries in the dermis, except for pigmented purpuric dermatoses. Inflammatory infiltrates around capillaries, extravasation of red cells and hemosiderin deposition can be found in pigmented purpuric dermatoses, unilateral nevoid telangiectasia and acquired port wine stains but not in AS and angiokeratomas. Angiokeratoma is a X-linked disorder that is characterized histopathologically by hyperkeratosis, papillomatosis, acanthosis, and dilated vasculature in the papillary dermis. Acquired port-wine stain occurs unilaterally on the face and neck, with nearly half of all cases found in the papillary dermis, usually smaller in numbers than that seen in angioma serpiginosum.

The lesions of angioma serpiginosum are cosmetically unsightly; laser therapy is considered an acceptable and effective mode of treatment [13]. Good cosmetic results can be achieved with a tunable pulse dye laser by selective photothermolysis to the vascular lesion. No other topical or systemic treatment options have been reported for angioma serpiginosum.

References