

Late Onset Angioma Serpiginosum: Report of Unusual Acral and Blaschkoid Cases

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Abstract

Observation: Angioma serpiginosum (AS) is a rare vascular nevoid disorder with dilatation and proliferation of capillaries in the papillary dermis. Clinical manifestation of AS is characterized by asymptomatic, serpiginous, grouped, red to violaceous punctate macules. The onset of this asymptomatic disorder is usually under the age of 20, but it may also start in adulthood. AS with the involvement of foot and sole is infrequent. Also, a linear clinical distribution of AS following Blaschko lines has rarely been reported. Herein we present 2 cases diagnosed with rarely involvement of AS. One of our patients had sole involvement and the other had Blaschkoid involvement in the leg. Our patient with sole involvement had suffered intense sorrow before the disease. Therefore, we think that the disorder in this patient might occur following stress and distress. Our patient with sole involvement showed complete regression after 3 months which has been rarely reported in the literature.

Introduction

Angioma serpiginosum (AS) is a rare vascular nevoid disorder with dilatation and proliferation of capillaries in the papillary dermis. Clinical manifestation of AS is characterized by asymptomatic, serpiginous, grouped, red to violaceous punctate macules [1, 2, 3, 4, 5, 6, 7, 8, 9, 10]. Clinical course and etiopathogenesis of this rare disorder are not fully understood. We suggest that the disorder may occur following stress and distress. Therefore, herein we present 2 cases diagnosed with rarely involvement of AS.

Case Report

Case 1: A 49-year-old female patient presented with reddish rash on the left leg. Her medical his-

tory revealed that rash had started on her left thigh 20 years ago and progressed over time extending from the left thigh down to the knee. Her medical history was not remarkable. There was no family history of a similar lesion. Dermatological examination revealed lesions consisted of violaceous red punctate macules with blurred borders, beginning from lateral part of the left thigh, extending toward medial and ending on the medial of down the knee following Blaschko lines (Figure 1). The lesions were non-blanchable on press. Her mucous membranes, hair and nails were normal. Her physical examination was normal except the dermatological finding. The patient had presented to another center with the same complaint 15 years ago, and underwent a skin biopsy with the provisional diagnosis of vasculitis; however the diagnosis could not be confirmed. Biopsy was taken



Figure 1. There were punctate maculae on the left thigh extending toward medial and ending on the medial of down the knee following Blaschko lines

again from the patient. In histopathological examination, there were vascular structures filled with erythrocytes and dilated within one focus in the papillary dermis (**Figure 2**). Laser therapy (Nd:YAG) was initiated and partial response to treatment was obtained, but the patient discontinued the therapy.

Case 2: A 32-year-old female patient presented with the complaint of erythema on her both soles. Her rash had started two months ago following an intense sorrow. Dermatologic examination revealed lesions consisted of violaceous red punctate macules (**Figure 3**). Her mucous membranes, hair and nails were normal. In histopathological examination, there were vascular structures filled with erythrocytes and dilated within one focus in the papillary dermis (**Figure 4**). Complaints of the



Figure 3. There are punctate maculae on the right and left sole

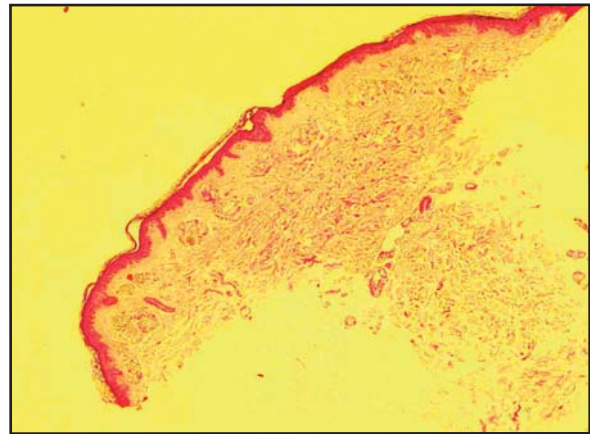


Figure 2. There were vascular structures filled with erythrocytes and dilated within one focus in the papillary dermis HEx100

patient were completely resolved about 3 months after the stressful period was over.

Discussion

Angioma serpiginosum is a rare benign vascular disorder characterized by red to violaceous, non-blanchable lesions with serpiginous distribution[1, 2, 3]. The pathogenesis of AS is not fully known. Since most of the cases have onset in the adolescence period, progression during pregnancy and association with hormone replacement therapy, increased levels of estrogens have been considered in the pathogenesis [1, 2, 3, 4, 5, 6]. However, immunohistochemical examination in some cases revealed no estrogen or progesterone receptors within the involved blood vessels. In addition, normal hormonal profiles of these patients support the opinion of hormonal stimuli have

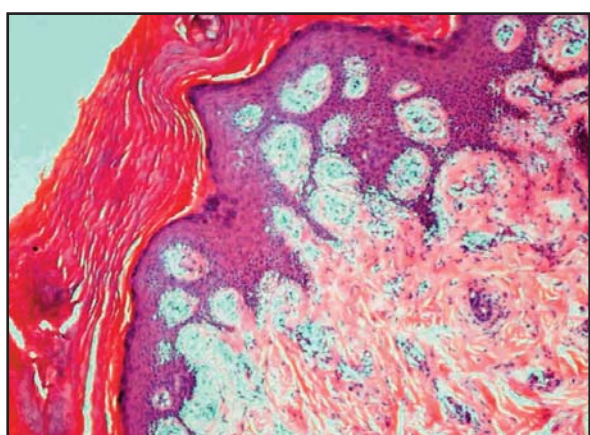


Figure 4. There were vascular structures filled with erythrocytes and dilated within one focus in the papillary dermis HEx100

no role in the pathogenesis of AS [4,5]. An abnormal vascular response to prolonged cold exposure has been proposed to play a role in the pathogenesis of AS [1, 2, 3]. In a Norwegian family, AS has been demonstrated to follow X-linked dominant inheritance with a distribution of the lesions that corresponded to Blaschko lines [1,3,7]. This finding suggested that AS may be developed due to ionization (random inactivation of one of each X chromosomes in all the somatic cells during embryological development) [3,8]. However, since there are some segmental and localized AS cases without familial involvement, it is thought that the disorder may also occur with the development of cutaneous mosaicism [3,8].

AS is usually localized in the lower extremities and hips in women and progresses asymptomatic with the patches that have erythematous background, consisting of red to purple-colored non-blanchable macules of needle tip in size. AS may be seen in any age and in both genders. Majority of the cases start under the age of 20, but it may also have onset in adulthood. The disorder is mostly sporadic, but autosomal dominant inheritance has been reported in two families [3,8]. AS may be confined to any body area such as the upper extremities, trunk, breast, feet and soles, while diffuse involvement may be observed. AS is often unilateral, but bilateral localization has also been reported [1, 2, 3, 4, 5, 6, 7, 8, 9, 10]. So far, only 3 cases with sole involvement have been reported. To our knowledge, our case is the fourth published in the literature. Linear distribution following Blaschko lines has also been reported [3,7]. New lesions spread from the center to the periphery within months or years in the early periods of life and the progression usually stops in the adulthood period [1, 2, 3, 4, 5, 6, 7, 8, 9, 10].

The lesions do not disappear with diascopy performed [2,9]. Another method which can be used in diagnosis is dermatoscopy which provides important information. In dermatoscopy, red lacunae that are primarily seen in eruptive hemangiomas and angiokeratomas may be seen in AS as well. These red lacunae consist of dilated capillaries in the papillary dermis which seem red because of the superficial capillaries influenced by AS [2,9]. The dermatoscopic view is different in cases with sole involvement. The dermoscopy reveals an eryt-

hematous parallel ridge pattern with an irregular globular component in acral AS. The red bands are composed of red rows of dots and globules arranged in a double line, with no involvement of acrosyringia openings. This appearance has been described by Freitas-Martinez [2]. We also performed dermatoscopy, but could not view. In the histopathology of AS, single or diffuse dilated capillaries are observed in the papillary dermis. There is no inflammatory accumulation, diapedesis of erythrocytes or pigmentation of hemosiderin. The red spots seen in clinic correspond to microaneurysms in the histopathologic examination [1, 2, 3, 4, 5, 6, 7, 8, 9, 10]. Because punctate macules mimic purpura, patients undergo unnecessary hematological tests. Other diseases that should be considered in the differential diagnosis include pigmented purpuric dermatosis, Henoch-Schönlein vasculitis, nevus flammeus, angiokeratoma corporis diffusum, angiokeratoma circumscriptum neviriforme, generalized or progressive essential telangiectasia and Kaposi's sarcoma. These disorders can be differentiated by dermoscopy and histopathological examination [1, 2, 3, 4, 5, 6, 7, 8, 9, 10].

The disorder is often not accompanied by systemic anomalies, but several capillary anomalies may rarely be seen including angioma in the retinal and spinal nerve roots [6].

Spontaneous regression may occur in individual punctate macules, but partial or complete regression is rare [2]. 532 nm potassium-titanyl phosphate (KTP) laser, pulse dye laser and IPL therapies are successful treatment methods [9,10].

Our patients have no history of spontaneous bleeding or by trauma. They have no remarkable medical and family past histories. Our patient with Blaschkoid involvement had been diagnosed with AS 20 years after occurrence of the disease. We could not identify any reason for the occurrence of AS. We initiated laser therapy and obtained partial benefit, but the patient did not attend to the control visits. Our 32 year old patient with sole involvement had suffered an intense sorrow before the disease. After suffering of the patient was improved, her lesions were spontaneously resolved after about 3 months. Complete regression has been rarely reported in the literature. Our case is noteworthy also by this aspect. Therefore,

we suggest that the disease might occur following the stress and distress.

Conclusion: AS is an important vascular dermatosis, because it is rarely seen, may be confused with vasculitis and may lead to unnecessary laboratory investigation. Acral skin involvement and blaschkoid arrangement should be included in the clinical spectrum of AS. Herein, we presented two rare cases of AS with one of them has sole and the other Blaschkoid involvement and late onset. We reviewed the relevant literature.

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