

## Case report

# Acroangiokeratosis (Pseudo-Kaposi's sarcoma); A challenging vascular condition

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### Abstract

Acroangiokeratosis (synonym pseudo-Kaposi's sarcoma) is an unusual, benign condition which clinically presents as erythematous, violaceous, brown, or purple-colored patches, plaques or nodules, mostly on the extensor surfaces of lower extremities. Acroangiokeratosis is often associated with different disorders including chronic venous insufficiency, arteriovenous malformations like Klippel-Trenaunay syndrome, iatrogenic arteriovenous shunts like fistula in patients under hemolysis (lesions are developing distally to arteriovenous shunts), and in paralysed limbs. It has also been reported above amputation stump of the limbs (especially in those with poorly fitting suction-type devices). It resembles aggressive conditions like Kaposi's sarcoma and requires histopathological examination for its diagnosis. We report a case of a 47-years old man with lesions of acroangiokeratosis in both lower legs and feet, secondary to chronic venous insufficiency. Histopathology showed dilated capillaries in the dermis with extravasated red blood cells, hemosiderin deposits and granulation tissue formation. In conclusion we would like to emphasize the importance of histopathology and immunohistochemistry in distinguishing this benign condition from similar looking malignant conditions especially Kaposi's sarcoma.

**Keywords:** Acroangiokeratosis, pseudo-Kaposi's sarcoma.

### Introduction

Acroangiokeratosis is a reactive angiodysplasia of cutaneous blood vessels associated with venous insufficiency or with vascular anomalies such as Klippel-Trenaunay syndrome or stump dermatosis in amputees. Exaggerated stasis dermatitis begins as violaceous macules and patches that gradually develop into papules, nodules, or indurated plaques often bilateral, and usually located on the lower extremities with edema. Although benign, it may mimic malignant conditions like Kaposi's sarcoma and therefore requires histopathological examination for its diagnosis and differentiation.<sup>1</sup> We describe a case of acroangiokeratosis which is a quite uncommon condition if not rare; and to our knowledge it's the first case in Hawler city to be diagnosed clinically, and to be confirmed later on

by histopathological examination and immunohistochemical study.

### Case Report

On May 2010, a 47 years old male patient presented to the dermatology unit in Rizgary Teaching Hospital in Hawler city with multiple erythematous to violaceous macules, patches, and plaques. The surface of some of these lesions had developed superficial ulcerations and crusting. They were distributed on the medial side of both lower legs and ankle joint area. The lesions gradually increased in number, some of them developed post-inflammatory hyperpigmentation. The lesions were slightly pruritic, but they were not painful (Figures 1 and 2). On examination, there were multiple small varicose veins on lower legs and feet, bilateral pitting leg edema, peripheral pulses were intact, and skin over the legs

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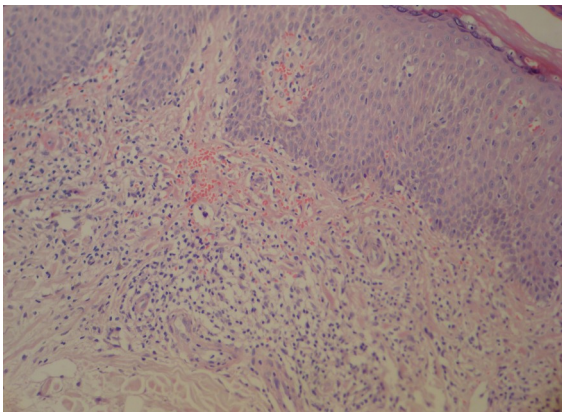
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was warm. A biopsy was taken from the lesion which showed hyperkeratotic stratum corneum, dilated capillaries with plump endothelial cells, extravasated red blood cells (RBCs), and hemosiderin, surrounded by granulation tissue with perivascular mononuclear cells infiltrate in the upper dermis. No vascular slits were seen (Figures 3 and 4). Immunohistochemical staining with CD34 was performed to confirm the diagnosis; it

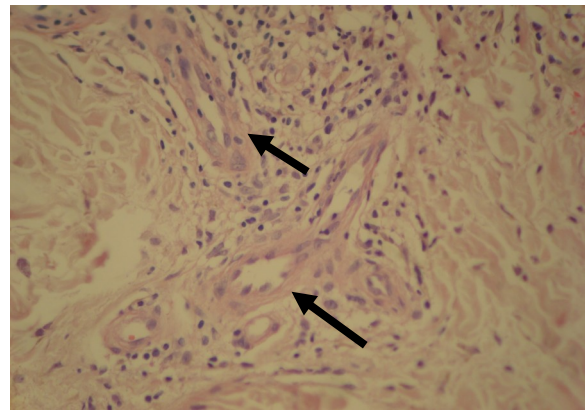
showed strong staining of the endothelial cells of proliferating vascular channels and an absence of perivascular CD34 staining is noted (Figures 5 and 6). The patient was treated conservatively with compression stockings and limb elevation during night hours, and later on referred to a cardiothoracic surgeon for a Doppler ultrasound test and further assessment of venous insufficiency of the lower extremities, but he disappeared on later follow up.



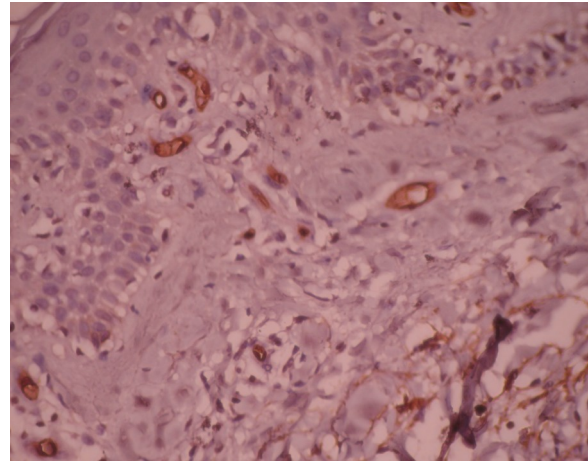
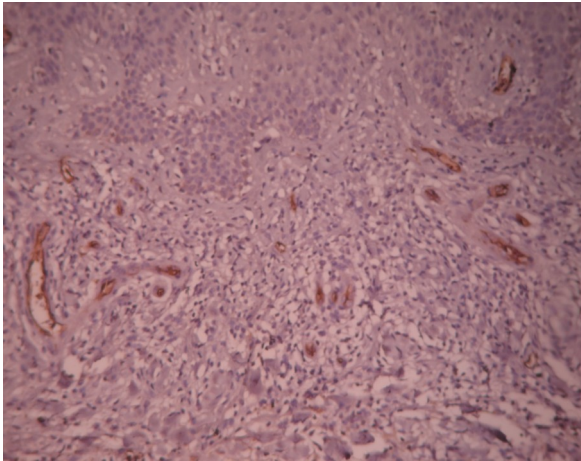
**Figure 1 & 2:** Multiple purple, reddish-brown patches and plaques on both lower limbs.



**Figure 3:** Histopathology, showing thick-walled capillaries, extravasated red blood cells, perivascular lymphocytic infiltrate (hematoxylin and eosin stain, X200)



**Figure 4:** Histopathology showing perivascular lymphocytic infiltrate and thick-walled capillaries (arrows) (hematoxylin and eosin stain, X400)



**Figure 5 & 6:** Immunohistochemistry: CD34 stained, with a strong positivity of the endothelial cells of proliferating vascular channels (arrows) and no staining of perivascular cells (IHC: CD34: X200 Figure 5, and X400 Figure 6)

### Discussion

Since Kopf and Gonzale, first described this condition as congenital dysplastic angiopathy in 1964,<sup>2</sup> a variety of terms have been applied to similar cases of this condition. In 1965, Mali *et al*, studied 18 cases of angiokeratosis associated with chronic venous insufficiency and first proposed the denomination "acroangiokeratosis", which is currently the most often used term.<sup>3</sup> In this same line of investigation, Bluefarb and Adams described this condition as "arteriovenous malformation with angiokeratosis".<sup>4</sup> In 1974, Earhart *et al*, studying a case in a 24-year-old man, noted the similarity, as much from the clinical as from the histopathological point of view; between this condition and Kaposi's sarcoma. Thus, they proposed the designation of "pseudo-Kaposi's sarcoma".<sup>5</sup> In 1982, Brenner *et al* published a study entitled "Kaposi-like arteriovenous malformation and Angiokeratosis".<sup>6</sup> Strutton and Weedon applied the designation of "Acroangiokeratosis".<sup>7</sup> Rüdinger performed a meticulous study on the subject, demonstrating the similarities of the syndromes of Mali and of Stewart-Bluefarb with Kaposi's sarcoma, however; they concluded that they were different

above all in relation to the prognosis and proposed the expression "Kaposi-like angiokeratosis".<sup>8</sup> Acroangiokeratosis can simulate a number of different clinical conditions such as Kaposi's sarcoma, pigmented purpura, lichen aureus, vasculitis, lichen simplex chronicus, actinic keratosis, basal cell carcinoma, stasis dermatitis, hemangioma, lymphangioma and lymphangiosarcoma.<sup>9</sup> Histopathologic examination shows proliferation of endothelial cells, newly-formed vessels with thick walls, often in a lobular pattern and surrounded by pericytes in the dermis. Extravasation of RBCs, hemosiderin pigment deposition, dermal fibrosis, small thrombi in the lumen and superficial perivascular infiltrate of lymphocytes, histiocytes and occasional plasma cells are also found, and may resemble Kaposi's sarcoma. This, however, has vascular slits, proliferation of fusiform cells and atypical cells, and the vascular hyperplasia is independent of pre-existing vasculature.<sup>9,10</sup> New techniques developed to avoid misdiagnosis include immunohistochemical studies with CD34 antigen expression to distinguish lesions of pseudo-Kaposi's sarcoma from those of authentic Kaposi's sarcoma. CD34 positivity is detected in both endothelial

cells and perivascular spindle cells of Kaposi's sarcoma, where there is a complete absence of CD34 perivascular cells of pseudo-Kaposi's sarcoma.<sup>11-13</sup> But more recently, the immunohistochemical demonstration of genomic material of HHV-8 in the nuclei of the proliferating cells in lesional skin has become the gold standard technique for histopathologic differential diagnosis between authentic Kaposi's sarcoma and all other vascular proliferations that may simulate Kaposi's sarcoma. HHV-8 may be immunohistochemically shown in the nuclei of the proliferating cells of lesions of Kaposi's sarcoma, both in all stages of evolution and in all epidemiologic variants of the process. In contrast, immunohistochemical studies in proliferating endothelial cells of all vascular proliferations that may be considered as histopathologic simulators of Kaposi's sarcoma have resulted negative for HHV-8.<sup>14</sup> Treatment tends to be conservative. Most of the treatment reports are of anecdotal nature. Compression therapy has been shown to heal ulcers secondary to pseudo-Kaposi's sarcoma and leads to regression of the lesions.<sup>15</sup> Erythromycin was found to be effective.<sup>3</sup> Therapeutic options consist of corrective arteriovenous shunts through embolization or surgery, surgical ligation of vessels or elastic compression therapy in chronic venous insufficiency. Anticoagulant therapy is useful in patients with thrombophilic alterations.<sup>16</sup>

### Conclusion

We would like to emphasize the importance of histopathology with immunohistochemical study in distinguishing this benign condition from similar looking malignant conditions, especially Kaposi's sarcoma.

### Conflicts of interest

The authors report no conflicts of interest

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