# **Case Report**

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# Angiolymphoid hyperplasia with eosinophilia as simultaneous cutaneous and vascular lesions with radial artery aneurysm

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#### **ABSTRACT**

Among the primary vascular neoformative processes is the angiolymphoid hyperplasia with eosinophilia (ALHE), rare and of endothelial origin with variable clinical presentation. We report an unusual presentation of ALHE, with both cutaneous and vascular lesions in the upper extremity, vascular lesion associated with radial artery aneurysm. A Male, 33 year-old with 6 months history of painless, erythematous and violaceous papule in upper extremity. No history of trauma or infection. Eosinophilia of 1.12 x 10<sup>3</sup>/uL. Negative serology of HHV8. Radiological imaging showed three similar solid lesions in forearm. One lesion was in contact with the radial artery, which presented aneurysmal dilation. Arteriography demonstrates radial artery aneurysm. The lesion biopsy was performed and exhibited morphological and immunocytochemical features compatible with ALHE. Surgical resection and bypass between brachial artery and radial artery distal to the lesion with inverted saphenous vein was performed. Histopathological and immunocytochemical study of the lesions confirmed the diagnosis. Our most interesting finding was the presence of multiple lesions in the upper extremity, which appeared as cutaneous and vascular lesions. The latter appeared as a vascular tumor with aneurismal dilatation of the radial artery. Our case highlights attention reporting simultaneous presentation of skin lesions and vascular lesions in the extremities, showing a spectrum of ALHE may be broader than known so far. Its etiology and long-term outcomes is still unknown. This puts the ALHE as differential diagnosis in cases of pulsatile cutaneous lesions and highlights the utility of radiological imaging and histopathological and immunocytochemical study.

Keywords: Angiolymphoid hyperplasia with eosinophilia, ALHE, Vascular tumors

## INTRODUCTION

Primary vascular neoformative processes involve a heterogeneous group of tumors classified according to their cellular content. As they are of rare occurrence, they require a multidisciplinary approach.

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare disorder of endothelial origin. It is known by different names, even sometimes confused with Kimura's disease (KD).  $^{1-3}$ 

The clinical presentation is variable, usually single lesions located in the head or the neck, and less frequently in the trunk, groin or limbs for which it has always been described as a single injury. It often affects skin and subcutaneous tissue, but vascular lesions have been described.<sup>1,5-7</sup>

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The behavior is unpredictable. There is no histological evidence of malignancy. ALHE is surgically resectable. The overall 5-year mortality is low and chemotherapy or radiotherapy usefulness has not been demonstrated. <sup>5,7,8</sup>

We present a case of ALHE presenting skin and multiple vascular lesions in the upper extremity. One of these lesions was associated with an aneurysm of the radial artery, which is an unusual presentation.

#### CASE REPORT

A 33 year old patient with a history of personality disorder was referred from the dermatology to the vascular surgery service, he was presented with purple lesions on the fingers, the palm and the right forearm that were prone to bleed, the biopsy's cytology was positive for ALHE. The initial treatment was  $CO_2$  laser to reduce the tendency to bleed.

Clinical history of the patient revealed that he presented, for about six months, erythematous papules and painless bruising on the hand and on the right forearm, without traumatic or infectious history.

The physical examination revealed purple papules on the previously described locations without bleeding tendencies and purplish coloration of the right thinner region. Upon palpation, a 2 cm diameter subfascial tumor was found in the right antecubital fossa without pulsatility when the limb is extended, but with thrill and pulsatility when bended. Right distal pulses were present.

Biochemical and hematological analysis were normal, except for the presence of eosinophilia of  $1.12 \times 103/uL$ . HHV8 serology was negative.

The Doppler ultrasound and CT Scan of the upper limb revealed lobulated mass, well-defined borders, richly vascularized, which included a proximal segment of the radial artery near to the brachial artery bifurcation, and with proximal and anterior expansion, associating arterial dilatation with Doppler signal, 80 x 30 x 24 mm, Lateral, Antero-Posterior and Transversal (LxAPxT) respectively.

MRI showed three similar solid lesions. The first one in front of the brachial muscle, between the brachioradialis and pronator muscles, of 76 x 27 x 34 mm corresponding to the lesion described in the Doppler Ultrasonography. The second lesion located in distal third of right arm, 26 x 13 x 7 mm. Second and third lesion seemed to be connected.

Arteriography demonstrated radial artery aneurysm within 1 cm of the humeral bifurcation and signs of hypervascularized tumor in the distal third of right forearm. The lesion biopsy was performed (11mm deep puncture with 18G needle) and exhibited morphological and inmunocytochemical features compatibles with ALHE.

Finally, the surgical resection of the lesion was decided. Resection of the 3 tumor masses was performed under general anesthesia. To repair the arterial defect, a bypass between the brachial and the radial artery distal to the lesion with a reversed intern saphenous vein was performed. No postoperative complications appeared and the extension study was negative.

Histopathology analysis of the lesion confirmed the diagnosis of ALHE, presenting a follicular and paracortical hyperplasia. Microscopic analysis showed proliferation of capillary and venular vessels that were regularly arranged in the connective tissue where the extensive foci of lymphocytic infiltrate were found presenting occasional activated lymphoid follicles and germinal centers and polymorphonuclear leukocytes mainly composed of eosinophils.

The immunohistochemical study demonstrated that the vascular elements of the lesions expressed CD31, CD34 and factor VIII. The lymphocytic component demonstrated expression of CD45, with a B cell (CD20) and another T cell (CD3) population arranged harmoniously. Histiocytes (CD68) and leukocytes (CD15) were also observed. Ki67 index was less than 2%. No expression of HHV8 was observed. Hematological analysis at 7 and 60 days confirmed decrease and disappearance of eosinophilia (0.69 y 0.19 x 10^3/uL, respectively).

The 3-year follow-up revealed a permeable bypass, without signs of stenosis, distal pulses were present, and lack of late complications or signs of tumor recurrence.

#### **DISCUSSION**

ALHE is a rare benign disorder, first published by Wells and Whimster. With the emergence of immunohistochemical techniques, it is possible to detect the expression of CD34 on the endothelial cells, which allow strengthening the diagnosis. In our case, in accordance with the findings of Aurello et al it has been possible to demonstrate the expression of CD34 in the resected specimen. Likewise; we also demonstrated the expression of CD31 and Factor VIII as demonstrated by Petrakis et al.

Our most interesting finding is the presence of multiple lesions in the upper extremity, which appear as cutaneous and vascular lesions. The latter appeared as a vascular tumor with aneurysmal dilatation of the radial artery. To our knowledge, this is the first described case of simultaneous presentation of skin and vascular lesions in extremities, extending the spectrum of clinical presentation of this rare disease.

Olsen and Helwig, 4 in their series of 116 cases, the largest reported to date, encountered lesions in upper extremity in only 6.8% of the cases, revealing how unusual this presentation is. Additionally, the author

reports that 21% ALHE presents multiple lesions, however, these has never been reported a multiple vascular lesions in the upper extremities neither was a simultaneous presentation with skin lesions. Moreover, previous articles only reported solitary lesions in this area. <sup>1,5-7</sup> Finally, few cases reported an associated aneurysmal dilatation.

Some authors describe laser therapy as an alternative treatment for multiple lesions, however in our case it showed little utility.<sup>2</sup>

#### CONCLUSION

Neoformative vascular processes and particularly ALHE represents a diagnostic and management challenge. However, thanks to a correct strategy for the clinical diagnosis, including radiological and novel pathology techniques and the increasing number of publications available on this disease, it is possible to correctly diagnose and treat this condition to avoid any medical diagnostic confusion as those that occurred in the past with other pathologies.

Our case extends the actual spectrum of clinical presentation of this disease. Its etiology and long-term outcome is still unknown. Our study also places the ALHE as a differential diagnosis in pulsatile skin lesions, regardless of their location and the number of lesions. It also highlights the utility of radiological imaging in their study. Early diagnosis can help decrease the morbidity associated with the disease.

#### Consent

Written informed consent was obtained from the patient for publication of this case report.

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