

Angiolymphoid hyperplasia with eosinophilia and entrapment of the ulnar nerve

Hambra Di Vitantonio, Danilo De Paulis¹, Alessandro Ricci¹, Soheila Dehcordi Raysi, Sara Marzi¹, Mattia Del Maestro, Renato Juan Galzio

Department of Life, Health and Environmental Sciences (MESVA), University of L'Aquila, ¹Department of Neurosurgery, San Salvatore city Hospital, L'Aquila, Italy

E-mail: Hambra Di Vitantonio - hambra.diviatntonio@gmail.com; *Danilo De Paulis - d.depaulis@alice.it; Alessandro Ricci - alexricci@email.it;

Soheila Dehcordi Raysi - soheila.raysi@alice.it; Sara Marzi - saramarzi@gmail.com; Mattia Del Maestro - mattiadelmastro@gmail.com;

Renato Juan Galzio - renato.galzio@cc.univaq.it

*Corresponding author

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Abstract

Background: The angiolymphoid hyperplasia with eosinophilia (ALHE) is a sporadic vasoproliferative lesion of uncertain etiology involving the skin and the subcutaneous tissue. Occasionally, it involves also the large arteries compressing the near nerves. ALHE is commonly confused with Kimura's disease because of their clinical and histological similarities.

Case Description: We report a case of a 52-year-old female suffering from a 6-month pain and paresthesias in the fourth and fifth finger of the right hand. The angiography showed a pseudoaneurysm in the proximal third of the right ulnar artery. A complete surgical excision of the vascular lesion was undertaken. The lesion forced the right ulnar nerve. The histopathological diagnosis deposed for ALHE.

Conclusion: Up to now, literature has described 8 cases of ALHE involving the arteries, and only one case originating from the ulnar nerve. The authors report a case of a female with ALHE involving the ulnar artery that compressed the ulnar nerve. Clinical aspects, radiological features, surgical treatment, and operative findings are discussed reviewing the pertinent literature.

Key Words: Angiolymphoid hyperplasia with eosinophilia, entrapment, pseudoaneurysm, ulnar nerve

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INTRODUCTION

The angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare benign inflammatory vascular lesion of unknown etiology.^[1] Usually, the tumor-like lesion involves the skin and the subcutaneous tissue^[9] of head and neck with a predilection for the retroauricular area.^[11] ALHE less frequently involves other sites including oral mucosa, tongue, parapharyngeal space and breast, as well as upper extremities within skeletal muscles.^[11] The intra-arterial development of the lesion is an exceptional occurrence.^[8]

The authors report a case of a female with ALHE involving the ulnar artery that compressed the ulnar

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nerve. Clinical aspects, radiological features, surgical treatment, and operative findings are discussed reviewing the pertinent literature.

CASE REPORT

A 52-year-old Caucasian woman presented with a 6-month history of pain in the ulnar side and paresthesias in the fourth and fifth fingers of the right hand, mainly during the night hours. There was no history of trauma or pathological antecedents. The symptomatology had increased during the last month. There was no external deformity. The palpation of the right elbow was extremely painful, with positive Tinel-Hoffmann sign at this level. The Froment sign was positive. The clinical examination revealed a pulsatile swelling between the flexor carpi ulnaris and the medial part of the flexor digitorum profundus on the right. A tactile dysesthesia was in the region of innervation of the right ulnar nerve, from the elbow to the hand. There was no dorsal interosseous muscle atrophy, no hypothenar atrophy, and no motor deficit. The previous electrophysiological study had been normal.

The blood test was normal with no peripheral eosinophilia and no elevation in serum immunoglobulin E (IgE) levels.

The Doppler color flow showed no alteration of the blood flow in the ulnar artery.

The preoperative magnetic resonance image showed a lesion with high signal intensity on T1- and T2-weighted images in the proximal third of the right forearm [Figure 1a and b].

The angiography showed a pseudoaneurysm in the proximal third of the right ulnar artery. The lesion measured about 28 mm [Figure 1c and d]. The complete surgical excision of the vascular lesion was undertaken. The lesion forced the right ulnar nerve along its course without trapping the nerve fibers. The histopathological diagnosis deposited for ALHE since the vessel wall was characterized by a lymphoid and eosinophilic infiltrate, as well as by plump or “hobnail-like” endothelial cells [Figure 2].

The postoperative angiography showed the complete removal of the pseudoaneurysm [Figure 3].

At 1-year follow-up, the patient was asymptomatic and feeling well.

DISCUSSION

ALHE is an unusual vasoproliferative disease^[9] described for the 1st time in 1969 by Well and Whimster.^[14] The alternative label of “epithelioid hemangioma” reflects the conceptual uncertainty about the reactive versus

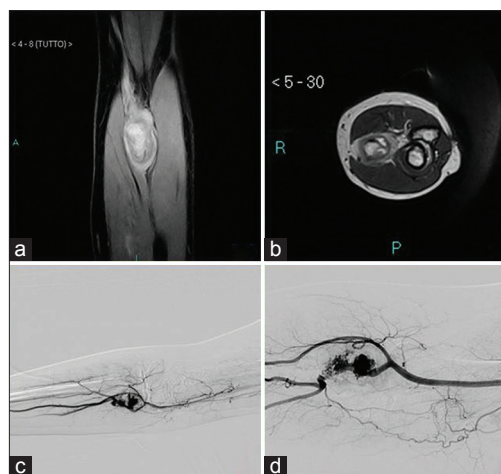


Figure 1: Preoperative magnetic resonance image in coronal and axial view showing a lesion in proximal third of the right ulnar artery (a and b). It is possible to see the relationship with the right ulnar nerve (b). The Angiography showed a pseudoaneurysm in proximal third of the right ulnar artery (c and d)

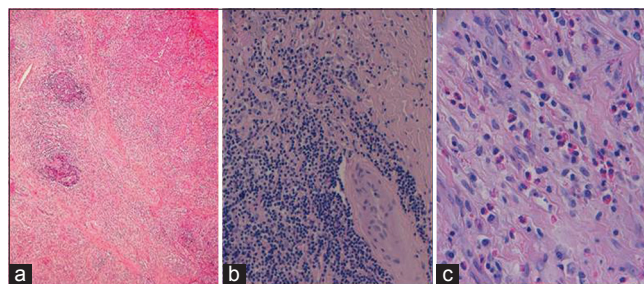


Figure 2: In hematoxilina and eosina and original magnification, $\times 40$, in the background, the wall of the pseudoaneurysm presented the lymphoid and eosinophilic infiltrate (a); the lymphoid infiltrate showed both a diffuse and a nodular pattern, with formation of follicles with prominent germinal centers (b); the endothelial cells often showed plump or “hobnail” appearance, eosinophilic cytoplasm, and round nuclei with granular chromatin (c)

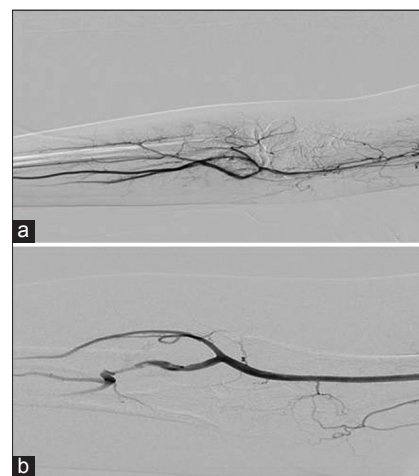


Figure 3: Postoperative angiography showed complete removal of the pseudoaneurysm (a and b)

neoplastic nature of ALHE.^[16] Patients develop nodules on the skin and in the subcutaneous tissue of head and

Table 1: Reviewing of literature on angiolymphoid hyperplasia with eosinophilia involving the arteries and nerves

Authors and year	Patient age (year)	Sex	Race	Artery	Nerve	Disease	Treatment
Morton <i>et al.</i> 1987	Not reported	Not reported	Not reported	Radial	-	ALHE	Not reported
Kimura <i>et al.</i> 2003	Not reported	Not reported	Not reported	Facial	-	ALHE	Not reported
Martorell <i>et al.</i> 2004	34	Male	Not reported	-	Ulnar	ALHE	Surgery
Arnander <i>et al.</i> 2006	Not reported	Female	Not reported	Temporal	-	ALHE	Not reported
Vandy <i>et al.</i> 2007 (2 cases)	26	Female	Caucasian	Brachial	-	ALHE	Surgery
	43	Male	Caucasian	Occipital	-	ALHE	Surgery
Bhat <i>et al.</i> 2010	30	Male	Not reported	Axillary	-	ALHE	Surgery
Koubaa <i>et al.</i> 2008	20	Male	Not reported	Temporal	-	ALHE	Surgery
Kukreja <i>et al.</i> 2011	34	Male	Not reported	Axillary	-	ALHE	Surgery

ALHE: Angiolymphoid hyperplasia with eosinophilia

neck.^[11] The noncutaneous localization of this pathology in the large arteries presenting as a pulsatile mass is extremely rare.^[3]

Up to now, the current literature has described 210+ cases.^[2,4,7,8,10-13,15] In most of them, the lesions were localized in the superficial and deep soft tissue of the head-neck region and were characterized by single or multiple smooth-top papules, or plaques of varying color.^[4,13] In 8 cases, ALHE involved the peripheral large arteries, and in only one case, it originated from the ulnar nerve.^[2,3,7,8,10-12,15] The average age of the reported patients was 30 years, with a male prevalence [Table 1]. However, the gender-related distribution of ALHE is currently uncertain, with some meta-analyses suggesting a predilection for females.^[5] The most frequent localization was in the arteries of the head and of the upper limbs, while there were no cases involving the arteries of the lower limbs. The treatment of choice was surgery without any other therapeutic options [Table 1].

The pathogenesis of ALHE is still a cause of debate.^[8] Various ideas have been proposed including the benign vascular neoplasm^[14] and the reactive inflammatory lesion.^[11] Ye *et al.*^[17] have proposed the pathogenetic relevance of the occasional detection of mutation in the endothelial cell tyrosine kinase receptor Tie-2. Instead, Kempf *et al.*^[6] have proposed the monoclonal rearrangement of the T-cell receptor gene in ALHE, and along the same line Andreae *et al.*^[1] have revealed the association with the peripheral T-cell lymphoma.^[1] Other reports have proposed a possible viral etiology, with involvement of the human herpes virus 8.^[11]

The differential diagnosis of this lesion, based on clinical and/or histopathological findings, includes pyogenic granuloma, reaction to insect bite, hemangioma, glomus tumor, hemangioendothelioma, angiosarcoma, and Kaposi sarcoma.^[3] The condition that is most commonly confused with ALHE is Kimura’s disease, which is a rare chronic inflammatory disease of unknown cause,^[14] more common in Oriental young men who present a tumor-like nodule and lymphadenopathy.^[11]

Clinically, ALHE rarely presents peripheral eosinophilia, elevated serum IgE levels, proteinuria, and nephritic syndrome, in contrast to Kimura’s disease,^[11] which is frequently associated with renal disease.^[14]

Histopathologically, ALHE is characterized by epithelioid endothelial cell changes, with rare lymphoid follicles [Figure 2], while in Kimura’s disease there are well-formed lymphoid follicles and a marked eosinophil infiltration.^[11]

ALHE rarely presents a spontaneous regression; however, usually a malignant transformation does not occur.^[10] Although ALHE is benign, recurrences are common, particularly after an incomplete removal.^[3] Other therapeutic options have been evaluated including intralesional corticosteroid injections, interferon- α 2b, cytotoxic agents, cryotherapy, electrodesiccation, pulse-dye laser, and carbon dioxide laser,^[10] but with limited success.^[11] Therefore, the surgical removal is still to be considered the most efficient option.^[8]

CONCLUSIONS

We believe that the pseudoaneurysm of the ulnar artery has determined the ulnar nerve entrapment at the cubital tunnel, without causing a permanent damage.

Although ALHE is a benign disease affecting the skin and the subcutaneous tissues of head and neck, its occurrence should be always suspected in the presence of a pulsatile mass along the course of the peripheral arteries and nerves, especially in the upper limbs. The surgical resection should be always radical, to prevent frequent recurrences and other systemic complications.

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Conflicts of interest

There are no conflicts of interest.

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