Angiolymphoid hyperplasia involving large arteries

Frank Vandy, MD,[a] Luis Izquierdo, MD,[b] Jingxuan Liu, MD,[c] and **Enrique Criado, MD,[a]** *Ann Arbor, Mich; Madrid, Spain; and Stony Brook, NY*

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare vascular proliferative disorder, which most commonly involves the skin of the head and neck regions. Noncutaneous localization of this pathology is unusual, and its primary localization in large arteries presenting as a pulsatile mass is extremely rare. We report here two cases of ALHE manifested as masses of the occipital and brachial artery. ALHE should be considered in the differential diagnosis of localized peripheral arterial masses in young patients. (J Vasc Surg 2008;47:1086-9.)

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon proliferative benign lesion involving the skin microvasculature, manifested by cutaneous plaques and nodules in the head and neck area.[1] Histologically, it is often confused with the similar appearing Kimura's disease (KD).[1-3] ALHE affecting large muscular arteries is extremely rare and a review of the literature reveals only four reports of ALHE affecting arteries in the extremities.[4-7] In our literature search, we have not found any cases of ALHE with primary localization in the brachial or occipital arteries. We report here two cases of ALHE manifested as aneurysms of the occipital and brachial arteries.

CASE REPORTS

Case 1. A 26-year-old Caucasian woman presented with a painless mass in her left upper arm. On physical examination, she had a mobile, pulsatile mass of approximately 2 cm of maximal diameter in the proximal and medial aspect of the arm. There were no associated dermal or intradermal papules, plaques, or nodules. Past medical history was noncontributory. There was no history of trauma or previous surgery to the left arm. Initial laboratory values did not reveal a peripheral eosinophilia. An IgE level was not drawn. Arterial duplex imaging as well as magnetic resonance imaging demonstrated a 1.6 cm diameter by 2 cm long well defined mass surrounding the brachial artery (Fig 1). Fine-needle aspiration cytology was inconclusive. The patient underwent en bloc resection of the mass and brachial artery with interposition of a reversed saphenous vein graft. The patient tolerated the procedure well with no perioperative complications. Histology of the specimen described the lesion as an angiolymphoid hyperplasia with eosinophilia (Fig 2). Margins were negative for tumor growth. Eighteen months after excision of the tumor, the postoperative vascular follow-up was uneventful and no evidence of tumor recurrence was detected.

Case 2. A 43-year-old Caucasian male presented with a tender 1 to 2 cm mass overlying the left occipital artery in the posterolateral aspect of the scalp. The mass was not mobile; it was tender to palpation and had a pulsation. An ultrasound revealed a 1.5 × 1 × 1 cm mass surrounding the left occipital artery. The patient did not have any skin lesions and had no eosinophilia on peripheral blood differential. The mass was resected under local anesthesia with proximal and distal ligation of the occipital artery. It had an inflammatory appearance and encased a neighboring segment of the occipital nerve. The patient did well postoperatively, except for significant scalp pain that resolved over an 18-month period. At the 24-month follow-up, the patient was doing well without evidence of local recurrence of the mass. Histological examination revealed an angiolymphoid hyperplasia with eosinophilia (Fig 3).

DISCUSSION

Angiolymphoid hyperplasia with eosinophilia (ALHE) was first described by Wells and Whimster in 1969.[8] This terminology was later included under the umbrella classification of histiocytoid hemangiomas described by Rosai in 1979, which also included intravenous atypical vascular proliferation, inflammatory arteriovenous hemangioma, pseudopyogenic granuloma, and hemangioendothelioma. However, the term histiocytoid hemangioma is often used interchangeably with epithelioid hemangioma in the current literature.[1,9-11]

Typically, ALHE presents in young to middle-aged women, with no predilection for race. The clinical appearance is usually one of bright or dusky red papules or nodules in the dermis or subcutaneous tissue of the head and neck region.[3] However, ALHE presentation in medium and large vessels, bone, tongue, periostium, and even heart has been reported.[2,10,12] In an observational study of 96 patients by Fetsch and Weiss, 8% presented with pain.[13] However, ALHE has more commonly a benign and indolent course. ALHE distinguishes itself from other hemangiomas by its histologically unique appearance. Most striking is the histiocyte like endothelium with its cuboidal to dome shaped endothelial cells lining a florid vascular proliferation. Additionally, there may be heavy inflammatory infiltrates of lymphocytes,

From the Cardiovascular Center, University of Michigan School of Medicine, Ann Arbor,[a] Instituto Vascular, Madrid,[b] and Stony Brook University Medical Center, Stony Brook.[c]

Competition of interest: none.

Correspondence: Enrique Criado, MD, FACS, Department of Surgery, Vascular Surgery Section, Cardiovascular Center, University of Michigan School of Medicine, 1500 E Medical Center Drive, Ann Arbor, MI 48109-5867 (e-mail: ecriado@umich.edu).

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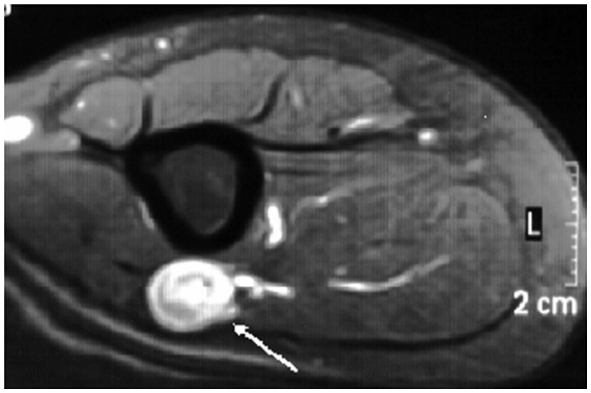

Fig 1. MRA of the left upper extremity demonstrating a well circumscribed mass surrounding the brachial artery (*arrow*). There is brachial artery dilatation and luminal defects.

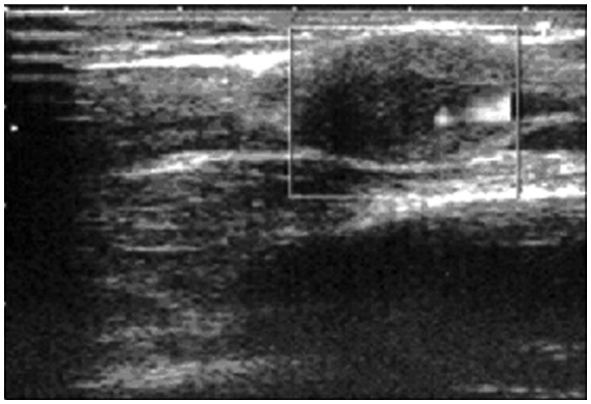

Fig 2. B-mode ultrasound of brachial artery. This longitudinal view demonstrates a dilated lumen with a surrounding mass giving the appearance of an aneurysm. Angiolymphoid hyperplasia with eosinophilia arising from brachial artery.

eosinophils, and the presence of lymphoid germinal centers.[1,3] Occasionally, ALHE courses with peripheral blood eosinophilia and elevated serum IgE.[14]

The diagnosis of ALHE arising from midsized to large arteries is exceedingly rare. In our literature review, we have not found any reports of AHLE involving the occipital or brachial arteries. Furthermore, we only encountered three cases of ALHE involving large vessels, including the radial artery, the popliteal artery, and the heart.[4-7,10] Our patients did not have the dermal or intradermal lesions characteristic of ALHE, but the histopathology confirmed the diagnosis. As shown in Fig 2, *b*, the "endothelioid cell" demonstrates the typical appearance of large, round, vesicular nuclei within a domed or cuboidal shaped cell. Additionally, an intense vascular proliferation surrounded by an abundance of lymphocytes and eosinophils is shown in Fig 2, *a*. Interestingly, Fig 2, *c* shows a lymphoid follicle. Although the presence of a lymphoid follicle does not weigh against a diagnosis of ALHE, their presence may make difficult the differentiation with Kimura's disease.

Kimura's disease (KD) was originally described in 1937 by Kimm and Szeto.[15] The classic presentation is in young Asian males with a discrete nodule in the head or neck area, commonly in the subcutaneous periauricular area or the salivary glands. Clinically, there is often associated regional lymphadenopathy accompanied by peripheral blood eosinophilia and elevated serum IgE.[15] Histologically, KD differs from ALHE in that the former lacks the dome-shaped endothelioid cell, but rather demonstrates a flattened endothelial cell.[14,16] Additionally, KD is characterized by the presence of multiple lymphoid follicles combined with massive eosinophilic infiltration and eosinophilic abscesses.[15] Similar to ALHE is evidence of vascular proliferation. Early literature described ALHE and KD as the same entity parts of a spectrum of disease, although recent literature supports a clear separation of these two disease processes.[14,16]

Although the presence of the cuboidal epithelioid cell supports the diagnosis of AHLE, it is necessary to point out atypical features in our cases that could suggest Kimura's disease. Clinically, the patient initially presented with a discrete proximal upper extremity mass, however, did not present with typical dermal lesions. Additionally, histology did show evidence of abundant eosinophils and presence of a germinal center.

The exact pathogenesis of ALHE has yet to be discovered. It has been considered to be a true vascular neoplasm of unknown etiology, while others believe that the proliferation is reactive to arterial or venous traumatic insults.[12,17] In the study by Fetsch and Weiss, the majority of cases were associated with vascular damage such as disruption of the muscular wall or fragmentation of the elastic lamina.[13] Additional evidence supports a role for infection, angiotensin II, arteriovenous malformations, and high estrogen states.[14] Like ALHE, the pathogenesis of Kimura's disease is unknown; it is thought to be a chronic immune-inflammatory disease. The etiology of Kimura's disease has been attributed to autoimmune disorder, allergic or neoplastic reactions, insect bites, parasites, and infection.[14,15]

Treatment for ALHE is ill defined given the rare nature of the lesion. A review of the literature illustrates both medical and surgical treatments.[18] However, surgical excision appears to be the treatment of choice. Although the disease is benign, surgical excision is still recommended because a histological analysis is necessary for diagnosis. Fine needle aspiration for initial diagnosis is often inconclusive.[19] Despite its benign etiology, ALHE still carries a 33% local recurrence rate.[12]

In summary, ALHE is a rare vascular proliferative disease, which to our knowledge has not been described presenting as aneurysms in the occipital or brachial artery. ALHE has considerable similarities with Kimura's disease, although the current literature supports that these lesions are different entities. Regardless, both processes carry an indolent course. Definitive diagnosis can

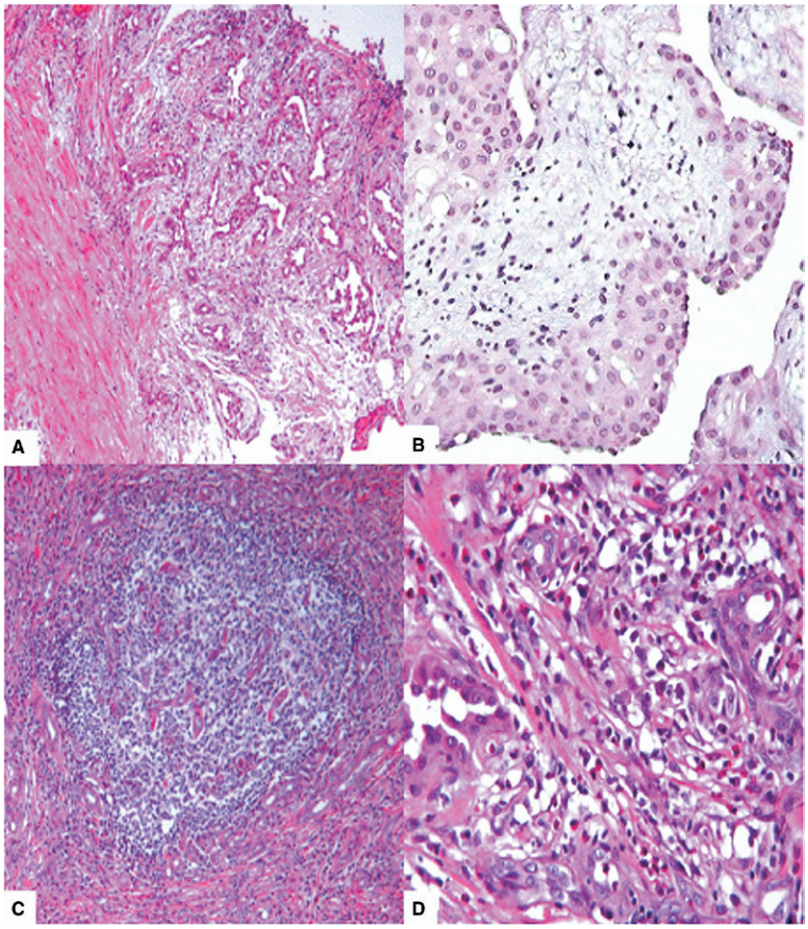

Fig 3. Microscopic images of specimen from angiolymphoid hyperplasia with eosinophilia arising from brachial artery. **A,** Low power (×100) power shows multiple proliferating small vessels in the wall of a larger vessel. **B,** High power (×400) shows small vascular spaces lined by epithelioid endothelial cells with abundant eosinophilic cytoplasm and round or ovoid vesicular nuclei. **C,** Intermediate power (×200) shows extensive lymphocytic and eosinophilic cell reaction with a lymphoid follicle. **D,** High power (×400) shows proliferating small vessels in a background of lymphocytes and eosinophils.

only be effectively made by the pathologist after surgical resection. Because of its tendency for local recurrence, wide local resection of these lesions appears to be the best treatment option.

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