Angiolymphoid Hyperplasia with Eosinophilia : a Case Report and Review of the Literature

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Abstract

A case of angiolymphoid hyperplasia with eosinophilia (ALH) is reported in a 42-year-old woman who developed multiple nodules behind the ear. Angiolymphoid hyperplasia with eosinophilia usually occurs on the head and neck of young adults and is more common in women than in men. Characteristic histologic features of ALH present in this case included proliferation of thick-walled blood vessels lined by prominent endothelial cells, infiltration of the interstitium by chronic inflammatory cells (mainly eosinophils), and presence of lymphoid follicles with germinal centers. The patient referred for surgeon for complete excision. in this context , cases previously described in the literature, and the differential diagnosis of ALH are discussed.

Angiolymphoid hyperplasia with eosinophilia (ALH) is a rare benign vascular tumor characterized by solitary or multiple red to brown papules or nodules found mainly on the head and neck of young adults between 20 and 40 years of age,[1] with a mean age at onset of 30 to 33 years.[2] It has also been described in other tissues, including liver, orbit, spleen, palate, bone, heart, and blood vessels.[1,3] It is more common in women than in men[1] and is associated with peripheral blood eosinophilia of 6% to 34% in about 20% of patients.[2,4] The lesions are often pruritic or painful and may coalesce into confluent plaques that are chronic, with little propensity for spontaneous resolution. Itching is significantly greater when the tumor is larger than 2 cm in diameter.[1] A history of trauma is found in some patients (9% in the study by Olsen and Helwig[4]). The interval between trauma and the onset of the lesion ranges from 7 months to 20 years, with a median interval of 30 months.[4] Lesion growth, tumor pulsation, and bleeding are other common presentations.[2,4] Most intradermal lesions are small, with diameters of 0.5 to 2 cm; subcutaneous lesions may be much larger, up to 5 to 10 cm in diameter.[2,5] The tumor is generally intradermal or subcutaneous, but it may involve deep soft tissues and vessels.[1] Peripheral blood eosinophilia, enlargement of regional lymph nodes, and arteriovenous shunts have been reported but are variable features.[4] We report a case of ALH that presented as retroauricular nodules in 42-year-old woman.

Keywords: Angiolymphoid,ALH, eosinophilia

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Case Report

42-year-old woman presented in the dermatology and venereology department in AL-Kindy teaching hospital with history of pain in the auricle of her right ear of 4months duration associated with attacks of bleeding especially when the lesion get traumatized during bathing the pain was sever and prevent the patient from getting sleep on the right ear. On examination multiple nodules 13 in number with intact pink to red brown skin color distributed behind the right auricle the largest was 2 × 1.5-cm lesion forming cauliflower-shaped and a single nodule located over helices of the ear figure 1,2. Lesion was felt soft on palpation. Her complete blood count was normal, and there was no peripheral blood eosinophilia. An excisional biopsy for one of nodule under local anesthesia was done and sends for histopathology which reveals a vascular lesion thick and thin walled vessels lined by plump endothelial cells. The epithelioid endothelial cells have rounded nuclei abundant eosinophilic cytoplasm The inflammatory component consists of lymphocytes with many eosinophils and mast cells within the stroma, figure 3, 4,5,6,7. The diagnosis

was confirmed as angiolymphoid hyperplasia with eosinophilia (ALH) and the patient was referred to surgeon to complete excision of all remaining nodular lesions.

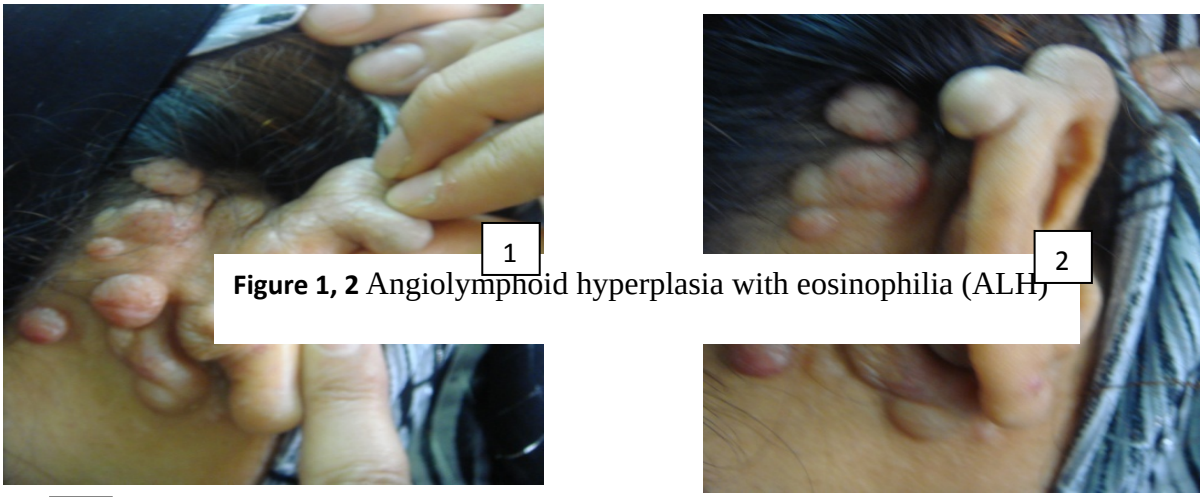


Figure 1, 2 Angiolymphoid hyperplasia with eosinophilia (ALH)

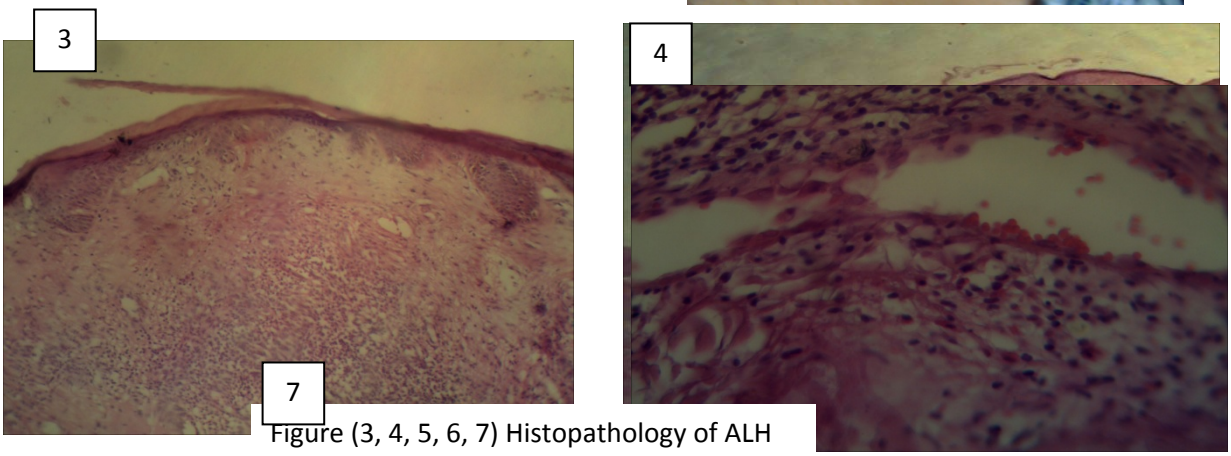
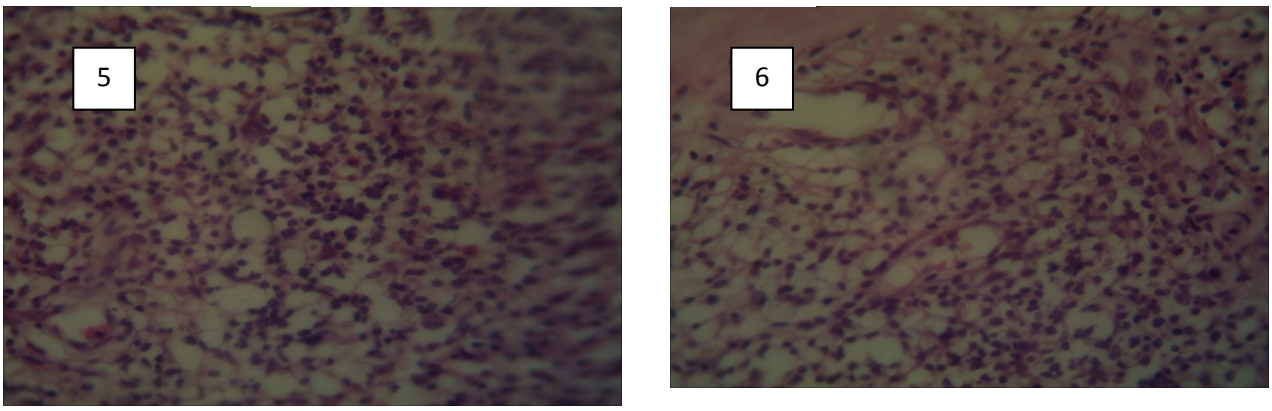


Figure (3, 4, 5, 6, 7) Histopathology of ALH



Comment

First described by Wells and Whimster[6] as "subcutaneous angiolymphoid hyperplasia with eosinophilia," ALH is a benign vascular tumor. It has also been called epithelioid hemangioma, inflammatory

angiomatous nodule, atypical or pseudopyogenic granuloma, and histiocytoid hemangioma. There has been confusion in differentiating between ALH and Kimura disease. Kimura et al[7] in 1948 in Japan indicated that Kimura disease was first described in the Chinese literature. They described the disease as an "unusual granulation combined with hyperplastic change of lymphatic tissue." Kimura disease is prevalent among young men of Asian descent. It presents as lymphadenopathy with or without an associated soft tissue mass and involvement of major salivary glands. Although the skin lesions in Kimura disease also commonly occur on the head and neck, they are located deeper than ALH lesions, usually in subcutaneous tissues. Peripheral eosinophilia is almost always present in Kimura disease, but it is only present in about 20% of ALH cases. Elevated serum IgE is usually present in Kimura disease but is rare in ALH.[1,8]

Microscopically, lesions in Kimura disease lack the epithelioid endothelial cells that are the morphologic hallmark of ALH.9Sclerosis, vascularization of the germinal centers, polykaryocytes, eosinophilic abscess, necrosis of the germinal centers, IgE-bearing dendritic cells in the germinal centers, and postcapillary venule proliferation are consistent features of Kimura disease. Kimura disease can also be distinguished from ALH by the presence of dense hyaline fibrosis in the affected lymph nodes.[1,8]

Angiolymphoid hyperplasia with eosinophilia has different presentations, resulting in various clinical impressions. In a study of 116 patients by Olsen and Helwig,[4] the most common clinical diagnoses were epidermal cysts and angiomas. Consistency, color, shape, size, and growth rate of the lesion are some of the factors that lead to prebiopsy diagnoses of scalp nodule or mass, pyogenic granuloma, lipoma, lymph node, and Kaposi sarcoma.4 Histologically, the differential diagnoses of ALH include benign and malignant vascular lesions of the skin, as well as various reactive conditions dominated by lymphocytes and eosinophils. If the clinical presentation of ALH is considered along with its typical microscopic features, then one can exclude other vascular and reactive lymphoid conditions, such as cavernous hemangioma, pyogenic granuloma, venous lake, capillary aneurysm, Kaposi sarcoma, angiomatous lymphoid hamartoma, granuloma faciale, eosinophilic granuloma, polyarteritis nodosa, pseudolymphoma (eg, insect bite, lymphocytic infiltration of Jessner, and lymphocytoma cutis), cutaneous angiosarcoma, and epithelioid hemangioendothelioma.[4]

Histologically, cavernous hemangioma has large cavernous vascular spaces separated by scant stroma, but prominent endothelial cells and inflammatory infiltrate are not present. Although proliferating capillaries and acute and chronic inflammatory infiltrates are present in pyogenic granuloma, its marked edema and absence of tombstone-like endothelial cells help differentiate it from ALH. Venous lake has a single large dilated space or several connecting dilated spaces that have thin walls and are lined by a single layer of flattened endothelium and scant stroma. Capillary aneurysm is considered a precursor of venous lake and has similar histologic features. In the early stage of Kaposi sarcoma, dilated and sometimes irregular and angulated blood vessels with interspersed infiltrate of lymphocytes, plasma cells, and macrophages are seen, but plump endothelial cells and eosinophilic infiltration are not present. The later stage of Kaposi sarcoma, which

is more cellular and may be composed of plump spindle cells, is rarely confused with ALH. Germinal center vascularization may be seen in the hyaline vascular type of angiomatous lymphoid hamartoma, but these germinal centers are generally atrophic and there are no eosinophilic infiltrates. Vasculitic changes, extravasated red blood cells, and hemosiderin deposition are often seen in granuloma faciale but not in ALH. Eosinophilic granuloma usually involves bone, and Langerhans histiocytosis is the characteristic feature of the lesion. In polyarteritis nodosa, inflammation occurs through the entire arterial wall, not just in the interstitium. Although mixed inflammation is present in pseudolymphoma, prominent endothelial cells of ALH are not seen. Cutaneous angiosarcoma is a malignant vascular neoplasm that affects older persons, presenting as slow-growing multiple red nodules on the face and scalp. It shows local invasion and distal metastatic spread. Microscopically, all degrees of differentiation of cutaneous angiosarcoma may be found, from mainly vascular channels with plump anaplastic but distinguishable endothelial cells to undifferentiated solid spindle cell tumors producing no blood vessels. Unlike ALH, cutaneous angiosarcoma does not show tissue eosinophilia.[10] Finally, epithelioid hemangioendothelioma presents as a soft tissue mass in adults, and histologically it has short cords and nests of slightly pleomorphic endothelial cells surrounded by a myxoid matrix. Unlike ALH, epithelioid hemangioendothelioma has no tissue eosinophilia.[11]

Rarely does ALH regress spontaneously. Therefore, treatment is generally necessary. Different treatment modalities have been used, but about 30% of ALH lesions recur, although none have been reported to metastasize, to our knowledge. Intralesional injections of glucocorticoids, interferon alfa-2a, and cytotoxic agents, as well as cryotherapy, electrodesiccation, and pulse-dye or long-pulse turntable dye laser, are among the modalities used. Deep surgical excision of ALH lesions has been reported to have the most favorable results, with no reported recurrence after 1-year follow-up.[12] Our patient was treated with surgical excision.

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