Angiolympoid Hyperplasia with Eosinophilia (ALHE)

**Patient:** R.T., a 31-year-old woman

**Duration:** 2 years

**Distribution:** Left periauricular area

**History:** Slightly pruritic lesions.

**Physical Exam:** Soft erythematous papules and nodules.

**Histopathology:**

**Skin:** Superficial and to a lesser extent deep predominantly perivascular nodular mixed lymphoehosinophilic infiltrate with scattered histiocytes and plasma cells in association with minimal vascular proliferation. Imzop studies including CD20 and CD45RO reveal focal staining of inflammatory cells by the B-cell marker and diffuse staining by the T-cell marker.

**Lymph node:** Proliferated blood vessels lined by epithelioid-like endothelial cells surrounded by a heavy inflammatory infiltrate rich in eosinophils and containing lymphoid follicles with germinal centers.

**Laboratory:**
MRI of brain – negative except for soft tissue swelling over nasal septum
MRA of intra- and extracranial vessels: negative
No peripheral blood eosinophilia

**Treatment:** Intralosomal steroid

**Discussion:**
- ALHE is a benign uncommon skin disease occurring slightly more frequently in females in the third and fourth decade with no racial predominance. Clinically, the lesions affect the scalp, face, ears and periauricular area as well as the neck. Peripheral blood eosinophilia and lymphadenopathy are each associated in only 20% of cases. Rare extracutaneous involvement has been reported especially in the nasal mucosa, oral mucosa, muscle, bone and salivary glands.

- In some cases with subcutaneous extensions, an underlying A-V malformation could be demonstrated. ALHE has a benign course, with some cases showing spontaneous regression. However, a 33% recurrence rate has been reported even after successful treatment.

- The etiology of ALHE is still unknown, and proposed pathogenesis includes a neoplastic process, a hypersensitivity reaction, inflammatory vascular reaction or a tissue reaction to a previous trauma as seen in cases of acquired traumatic A-V fistulas.

- Therapy includes steroids (intralosomal, oral), oral retinoids, pentoxifyllin, intralosomal
chemotherapy (vinblastine, bleomycin, fluouracil), radiotherapy, cryotherapy. The most recommended management remains deep surgical excision. Vascular laser therapy has also been used.

- Angiolymphoid hyperplasia should be distinguished from Kimura’s disease. Kimura’s disease is associated with markedly elevated peripheral eosinophilia and elevated serum IgE levels as well as lymphadenopathy. Clinically, it is usually a deep solitary large lesion with normal overlying skin. Histologically, it is characterized by a marked lymphoid hyperplasia with an inflammatory infiltrate rich in eosinophils and fibrosis. Compared to ALHE, the vascular component is much less prominent and lacks the characteristic endothelial cells. Lymphadenopathy in Kimura’s disease involves eosinophilic microabscesses and heavy IgE deposition in the germinal centers.

References:

