Kimura’s Disease

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Abstract
A rare case of Kimura’s disease alongwith AV Malformation around left pinna in a 26 year old male is being described. AV Malformation had two feeders from superficial temporal artery and occipital artery. The lesion was excised along with ligation of feeders. Post operatively patient was put on systemic corticosteroid therapy with some benefits.

Key Words
Kimura’s disease, ALHE, AV Malformation (AVM)

Introduction
Kimura’s disease is an inflammatory disorder that presents as massive subcutaneous swelling in the periauricular and submandibular region in young asian men (1). Histologically prominent germinal centers with eosinophils are present in the subcutaneous tissues. Although blood vessels are in abundance, changes are less prominent than in angiolymphoid hyperplasia with eosinophilia (ALHE) which is an uncommon idiopathic condition that presents with isolated on grouped plaques or nodule in the periauricular region, forehead or scalp. ALHE is marked by a proliferation of blood vessels with distinctive large endothelial cells accompanied by a characteristic inflammatory infiltrate that includes eosinophils (2). Kimura is frequently accompanied by lymphadenopathy, eosinophilia and elevated IGE levels. The lesion is benign but may be persistent and difficult to eradicate. Various therapeutic modalities that have been tried for its treatment include intralesional and oral corticosteroids, cryotherapy, oral retinoids, vinblastine, surgical excision, laser therapy and Infalpaha2a (3,4).

Case Report
We report an unusual case of 26 year old male presenting with throbbing swelling around left pinna 6 months duration of symptoms with the swelling progressively increasing in size.(Fig-1).There was induration and redness of left pinna. The swelling was compressible with the positive bruit. The Vascular Doppler detected two feeders. MRI Angiography confirmed the presence of one feeder from Occipital artery and other from superficial temporal artery supplying the AVM around pinna.(Fig-2)

Patient underwent surgery in form of ligation of the feeders from the superficial temporal artery and occipital artery along with excision of the cutaneous lesions. However, pinna was spared. The histopathological features of the lesion corresponded to the soft tissue...
angiomatosis with accompanying lymphoid hyperplasia with reactive germinal center formation and Eosinophilia suggestive of Kimura’s disease. (Fig- 3) Post operatively patient was put on systemic corticosteroid therapy with some benefit.

**Fig-2: MRI Showing The AVM Around Pinna With Feeders From Occipital and ST Artery**

**Discussion**

Lu (5) in a comparative study of 12 cases compared Kimura’s disease (KD) with epitheloid hemangioma (EH). KD represented a lymphoid hyperplasia and EH a benign vascular tumor. Patient in our case had clinicopathological features of KD but associated with AV malformation around left pinna, Chan KM (6) has reported case of Kimura’s disease of the auricle Misago N (7) reported an unusual case of arteriovenous (AV) malformation (localized form of soft tissue angiomatosis): a reaction of angiolymphoid hyperplasia with eosinophilia (ALHE) developed over the lesion of this pre-existing AV malformation. However in our patient there were definite vascular feeders from occipital and superficial temporal artery along with histological features suggestive of Kimura’s disease a combination which is unparalleled in screened literature.

**Fig -3: Angiomatosis With Lymphoid Hyperplasia With Reactive Germinal Center Formation &Esinophilia ( HXE- 400)**

The treatment of Kimura’s disease is problematic. Surgical resection has been found to be curative although recurrence is common (4). Localized initial regrowth can be managed with surgical excision, other therapeutic options include radiation, systemic corticosteroids, Cytotoxic agents, cyclosporin and pentoxyfiline. Systemic steroids may be indicated in frequent relapses or cases complicated by nephrotic syndrome, Sato S et al (8) has suggested combined use of steroids with cyclosporine with good response in a patient. Radiation may be considered in cases refractory to surgical and medical therapy.

**References**