KIMURA’S DISEASE – A CASE REPORT

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ABSTRACT
Kimura’s disease is a chronic inflammatory disorder of unknown cause primarily seen in Asian males, which involves the skin, subcutaneous tissues and lymph nodes. The Kimura’s disease is characterized by a triad of painless subcutaneous masses predominantly in head and neck region with blood and tissue eosinophilia and elevated serum IgE levels. We report a case of a 50 year old man for Union territory of Puducherry, Indian presenting with cheek swelling on both right and left side over the angle of mandible for the past 20 years. Based on the histopathological findings a diagnosis of the rare entity the Kimura’s disease was made.

Key Words: Kimura’s Disease, Eosinophilia, Serum IgE

INTRODUCTION
Kimura’s disease also known as epithelioid hemangioma is a rare chronic inflammatory disorder, which is endemic in Asian countries, which typically affects young and adult males of unknown cause. Kimura’s disease first described in 1937 in Chinese literature by Kimm and Szeto, termed it as “Eosinophilic Hyperplastic Lymphogranuloma” (Pitak-Arnnop et al., 2010). The disease became widely known as Kimura’s disease after the Kimura and colleagues reported two cases of unusual granulation combined with hyperplastic changes of the lymphoid tissue (Chung et al., 2010). Clinically Kimura’s disease is characterized by formation of subcutaneous nodular masses, localized mostly in the neck and the head. Peripheral blood eosinophilia and increased IgE serum levels are frequently encountered. In histopathology, tissue infiltration of eosinophils and lymphocytes as well as eosinophilic abscesses and lymphoid follicles with germinal centres and venular hyperplasia with flat or cuboidal endothelium were observed.

CASES
The patient, a 50 year old man, for the past 20 years had subcutaneous nodules developed in both right and left angle of the mandible which measure about 4 x 4 cm and 3 x 4 cm respectively. This nodule grew slowly and was associated with non-tender, no pain or local skin changes. He is a smoker for the past 20 years and had poor oral hygiene. Histopathological investigation by incisional biopsy using Fine Needle Aspiration Cytology (FNAC) was done. Incisional biopsy revealed that portions of lymph node with follicular hyperplasia, paracortical sclerosis and zones of eosinophilic abscesses. The eosinophilic abscess also shows collection of plasma cells (Figure 1). The germinal centres show proteinaceous material at places with increased vacularisation.

DISCUSSION
Kimura’s disease predominantly affecting the head and neck region, major salivary glands and lymph nodes. The etiopathogenesis of Kimura’s disease remains unclear, it is considered as an allergic disease of systemic immunological disorder. The cellular and humoral immunity is regulated by distinct subsets of
helper T-cells (Th), former is regulated by Th1 cells and later is regulated by Th2 cells (Marinko et al., 2001).

Figure 1: Incisional biopsy revealed that portions of lymph node with follicular hyperplasia, paracortical sclerosis and zones of eosinophilic abscesses. The eosinophilic abscess also shows collection of plasma cells

Two subsets of effector Th cells have been defined on the basis of their distinct cytokine secretion patterns and their immunomodulatory effects. Th1 cells produce predominantly IL-2 and IFN-gamma, which is required for cell mediated immune response, Th2 cells secret predominantly IL-4 which mediate B cell activation and differentiation of humoral immune response (Paul and Seder, 1994). In Kimura’s disease eosinophilia and elevated serum IgE levels may speculate that balance between Th1:Th2 ratio was augmented towards Th2 of humoral immune response. In Kimura’s disease, germinal centers are destroyed due to heavy infiltration of eosinophils, rarely even forming microabcesses (Ranka et al., 2004; Kuo et al., 1988). Kimura’s disease has a high recurrence rate so early and definitive diagnosis of the disease is vital for effective treatment usually involves the administration of parenteral steroids, surgically excising and radiotherapy.

REFERENCES