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KIMURA DISEASE – A CASE REPORT

Pathology

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ABSTRACT

Kimura disease is a rare chronic inflammatory disorder of unknown etiology and is seen in an endemic form in Asia and also in other parts of the world which includes United States and Europe. Kimura disease usually presents as a mass lesion located in the subcutaneous tissue of head and neck region or the major salivary glands and is often associated with regional lymphadenopathy. In few cases lymph node enlargement is the only presentation of Kimura disease. This disease is a rare entity and only few cases have been reported and published in the literature. Herein, we describe a rare case report of Kimura disease seen in 13 years old male child presented with post auricular swelling.

KEYWORDS

Kimura, Lymphadenopathy, Eosinophilia

INTRODUCTION

Kimura disease is a very rare chronic inflammatory condition with approximately 200 cases reported in the literature worldwide [1]. The clinical presentation includes multifocal subcutaneous mass lesions of head and neck which involves cervical lymph nodes, soft tissue and salivary glands. The pathogenesis of Kimura disease is largely unknown. Some believe that there may be a concordance with atopy due to the presence of eosinophilic infiltrates with in the mass lesions, peripheral eosinophilia and elevated IgE levels [2]. Histopathological analysis revealed marked hyperplasia of germinal centres, The germinal centres are most often well vascularized and contain polykaryocytes, interstitial fibrosis, and deposition of a proteinaceous material. In addition, there is an extensive infiltration by mature eosinophils and formation of eosinophilic abscesses. Hyalinized vessels are also seen in the paracortical region along with variable degree of sinus and paracortical sclerosis [3-5]. Kimura disease most commonly affects age group of 20 to 40 years and also can be seen in young children. Males are commonly affected than females. People with Southeast Asian descent are commonly affected. [6, 7]. There is no standard of care established due to the rare occurrence. However immune-modulating agents such as corticosteroids as well as cyclosporine are implemented as the first line treatment. Upon medical failure, other modalities like radiation as well as surgery are employed [8, 9].

Case Report

A 13-year-old male child came to the Surgical OPD with complaints of post auricular swelling insidious in onset and gradually progressive for 5 years duration. He had no other co-morbidities. Clinical examination revealed swelling of size 4x2 cm in the post auricular region. Excision biopsy was done for this patient. The specimen received includes 2 Lymph nodes. The largest node measures 1.8x1.5x0.5 cm and the smallest node measures 1.2x1x0.4 cm. Histopathological analysis revealed Kimura's Lymphadenitis with prominent eosinophilic infiltration and eosinophilic micro abscess. Also seen in this patient is Eosinophilia in peripheral blood smear. Haematological examination revealed Haemoglobin 12.8gm/dl, Total WBC count 7480 cells/cu.mm (Neutrophils 40%, Lymphocytes 41%, Eosinophils 12%, Monocytes 06%, Basophil 01%)

Microscopic Findings

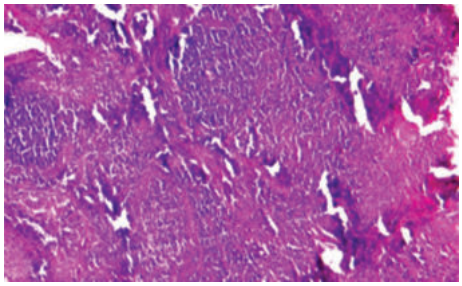

Figure 1: Hyperplastic Follicles With Germinal Centers

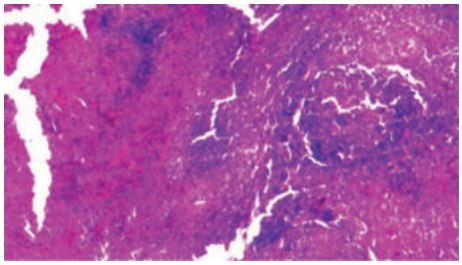

Figure 2: Eosinophilic Microabscess Formation

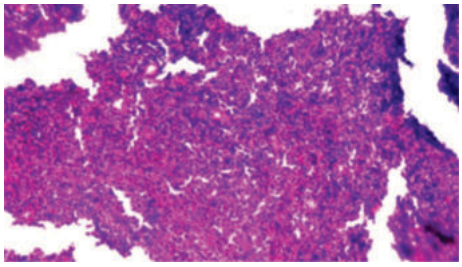

Figure 3: Eosinophilic Infiltrates (High Power View)

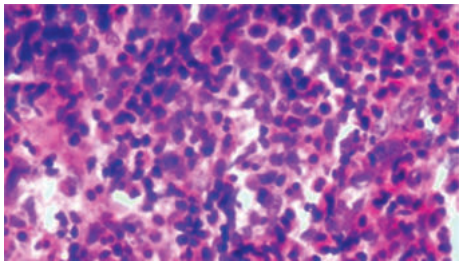

Figure 4: Polymorphous Population of Lymphoid Cells with Eosinophilic inFiltrates

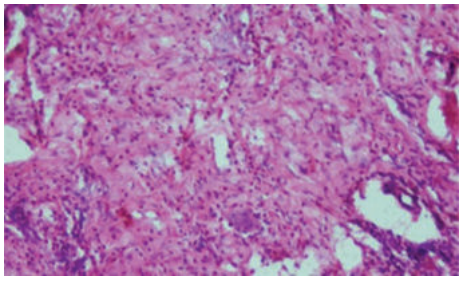

Figure 5: Vascular Proliferation in Interfollicular Areas with Sclerosis Admixed with Eosinophils

DISCUSSION

Kimura disease is a very rare chronic inflammatory condition and it was first described by Kim and Szeto in 1937 in Chinese literature as "Eosinophilic hyperplastic Lymphogranuloma" and has been known as Kimura's disease after its description in 1948 by Kimura et al. in the Japanese Literature. Kimura disease most commonly affects the age group of 20 to 40 years and also can be seen in young children. Males are commonly affected than females with a ratio of 3:1[10].

The clinical course of this disease is usually benign and it is self-limited. The other organs which can be involved includes Orbit, Lacrimal glands, Eyelid and Kidney. The renal involvement can be seen in the form of Nephrotic Syndrome and Extra membranous glomerulonephritis. The involvement of Kidney is a warning sign for patients with Kimura disease [10].

The histological characteristics of Kimura's disease had been described by Hui et al. The findings include preserved nodal architecture, eosinophilic infiltration, germinal centre hyperplasia and post capillary venule proliferation. Also seen are Subcapsular sinusoidal obliteration and capsular fibrosis. The nearest differential of Kimura's disease includes ALHE (Angio Lymphoid Hyperplasia with Eosinophilia). ALHE demonstrates prominent vascular proliferation and forms aggregates with epithelioid and histiocytoid changes [3]

CONCLUSION

Kimura disease is an extremely rare chronic inflammatory disorder of unknown etiology. The relevance of this case report is mainly due to rarity of the disease. The diagnosis of the Kimura disease has remained challenging due to the limited clinical presentation. Knowledge of this disease will make the physicians include Kimura disease in the differential diagnosis of patients with painless cervical lymphadenopathy and peripheral Eosinophilia.

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