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Case Report

Kimura's disease: rare inflammatory condition mimicing as soft tissue neoplasm

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ABSTRACT

Kimura's disease is rare benign chronic inflammatory condition of unknown etiology involving head and neck region. The characteristic features of this condition are elevated serum IGE levels and microscopic picture reveals lymphoid proliferation with eosinophilic infiltration. It is commonly misdiagnosed as soft tissue neoplasm. Surgical wide local excision is the main stay of treatment. Sometimes combined therapy (excision and postoperative radiotherapy) is needed. Here we are presenting one interesting rare case, 32-year-old male with post auricular swelling, diagnosed as soft tissue tumor preoperatively, after excision came out as kimura's disease.

Keywords: Eosinophilia, Kimura's disease, Lymphadenopathy, Lymphoid follicular hyperplasia

INTRODUCTION

Kimura's disease (KD) is rare benign chronic inflammatory condition of unknown etiology which is characterized by painless progressive subcutaneous nodules in head and neck region, elevated eosinophils and serum IgE levels.[1,2] It is generally seen in young asian males with mean age of 20-40 years with male to female ratio of 3:1.[3,4] The etiology of KD is unknown but may be due to impairment or interference with immune regulation, atopic reaction to a persistent antigenic stimulus by arthropod bites, virus, and neoplasm. Due to raised eosinophils and lymphoid follicular hyperplasia KD is hypothised to be a hypersensitivity reaction.[4] In current era there is no diagnostic criteria and the primary diagnostic tool is histopathological examination.[5] In this condition, simultaneous presentation of renal disease is seen in 10% to 60% and 10% to 12% of patients may suffer from nephrotic syndrome. Renal impairment is probably due to immunocomplex-mediated damage or to Th2-dominant immune response.[6] Here, we are reporting this rare inflammatory condition which can be mimics as soft tissue tumor with diagnostic difficulty.

CASE REPORT

A 35-year-old male patient presented to general surgery out patients department (OPD) with left retro auricular swelling in the last 1 year. The swelling was painless and had gradually increased in size since its appearance. The patient had observed that the swelling grew to its present size with cosmetic disfiguration. On clinical examination, an oval shaped firm swelling of size 5x4 cm was present behind left ear and lying over left mastoid region. The swelling was non tender and non-mobile. With the above-mentioned history and clinical examination, provisional diagnosis of soft tissue neoplasm was made. Routine blood examination revealed eosinophilia 2070 cells/dl (normal<500 cells/dl). USG was suggestive of vascular malformation and MRI revealed a well-defined T2 heterogenous hyperintense lesion arising from deep planes of left mastoid region with infiltration into left sternocleidomastoid muscle and enlarged left cervical

lymph nodes giving an impression of soft tissue neoplasm (Figure 1). With this diagnosis, wide local excision was planned after receiving valid consent. Under general anesthesia, wide local excision with 0.5 cm margin was done. Patient was discharged on postoperative day 3 and followed up in the OPD. Postoperative period was uneventful.

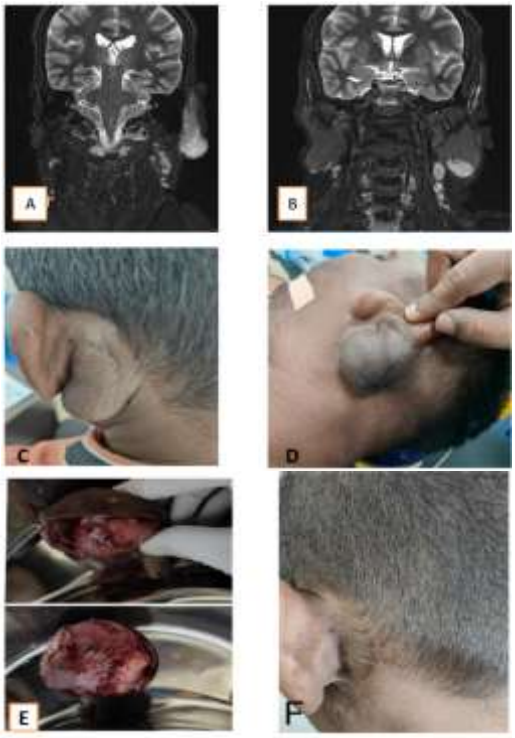

Figure 1: (A) Coronal T2 STIR sequence showing hyperintense soft tissue swelling in the subcutaneous plane with left sided cervical lymphadenopathy; (B) coronal T2 STIR sequence showing enlarged left parotid with intraparotid lymphnode; (C & D) per operative image showing lesion in left retroauricular region; (D) intra operative images showing 7×3 cm grey white to grey brown tumour with dense adhesion to skin (F) image after 6 months follow up.

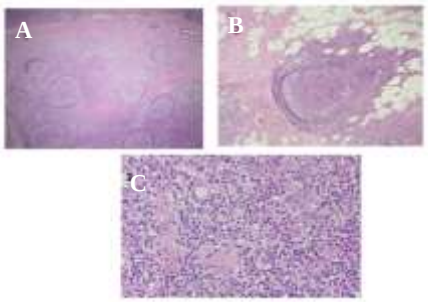

Figure 2 (A-C): Histopathological images showing in scanner image showing hyperplasia of germinal centre, low power microscopy showing lymphoid follicle with deposition of proteinaceous material and high power microscopy showing infiltration of eosinophils.

DISCUSSION

Kimura's disease was described by Kim and Szeto as "eosinophilic proliferative lymphogranuloma" in a Chinese journal in 1937. Later in 1948, Japanese scholars Kimura et al reported the related cases with the title "on the unusual granulation combined with hyperplastic changes of lymphatic tissue" and described its definitive histopathological features which later became widely known as Kimura disease.[7] It is generally seen in young asian males with mean age of 20-40 years with male to female ratio of 3:1 with mean age of affected individuals being 31 years.[8] The most frequently affected areas are the head and neck region, especially the parotid glands, salivary glands, and lymph nodes followed by other areas such as the groin, breast, arm and orbit have also been reported.[2] The underlying causes of Kimura's Disease (KD) remain a mystery. Several theories have been proposed, but none have been definitively proven. The most widely accepted hypothesis currently suggests that infections and parasitic factors, such as bites from arthropods, Epstein–Barr virus, HHV, and Candida albicans, may disrupt T cell regulation or trigger IgE-mediated type 1 hypersensitivity reactions. The presence of elevated eosinophil levels in the peripheral blood and bone marrow, along with increased serum IgE levels in KD patients, indicates that T helper cells (both 1 and 2 type) and T regulatory cells might play a role in the disease's development.[9] Renal complications occur in 10% to 60% of individuals with KD, and 10% to 12% may experience nephrotic syndrome. The renal issues are thought to arise from IgE deposits in the kidney glomeruli, eosinophil infiltration, and alterations in the permeability of the basement membrane due to cytokines.[10,11]Histopathology of KD shows tissue show lymphoid follicles in the subcutaneous areas which show hyperplasia of germinal Centers. Few follicles show deposition of proteinaceous material and variable amount of polykaryocytes. Few thickened vessels and marked infiltration of eosinophils are seen in perinodal and interfollicular area. No granulomas or malignant cells.[12]

The primary treatment for Kimura disease involves surgical removal of the affected tissue. This approach is often complemented by additional treatments such as radiotherapy, chemotherapy, or steroid therapy. The first-line management strategy is the surgical excision of nodules, which has a recurrence rate of up to 25% when performed as a standalone procedure. Oral corticosteroid treatments, such as prednisone at a dosage of 0.5-0.7 mg/kg per day and methylprednisolone at 0.5 mg/kg per day, are also used but have a higher recurrence rate of 60%. Other medications that may be employed in conjunction with steroids include cyclosporin, methotrexate, and leflunomide. For cases that recur, a combination of primary excision followed by postoperative radiotherapy may be considered.[13] Patients with a tumor size ≥3 cm, duration ≥5 years, eosinophil count ≥20%, or a serum IgE level ≥10,000 IU/ml has more chances to recur after surgical resection alone hence

combination therapy is preferred with 3.7 years (3-5 years) as mean duration of reccurance.[14]

CONCLUSION

Kimura's disease is rare benign disease which has more chances of recurrence if treated with single modality and there is neither fixed regimen nor duration of treatment. KD is difficult to diagnose through imaging alone and histopathology is the definitive diagnostic method. KD is usually misinterpreted as soft tissue neoplasm and thus needs to be treated accordingly

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