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Case Report

Kimura's disease as an uncommon cause of post auricular swelling

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ABSTRACT

Kimura's disease (KD) is a rare chronic inflammatory disorder of unknown etiology, primarily seen in young Asian males. In India, only 200 cases have been reported worldwide since its histopathological diagnosis. The disease is characterized by painless subcutaneous swelling in head & neck region, blood and tissue eosinophilia and raised Immunoglobulin E (IgE) levels. A systematic multidisciplinary approach is mandatory to rule out the other common causes of post auricular lymphadenopathy. The diagnosis of KD can be difficult and misleading and patients with this disease are often evaluated using avoidable procedures by just not being aware of KD. Here, we present a case of a 20 year old male who presented with nodular swellings in the bilateral post auricular region. The diagnosis of KD was done based on characteristic histopathologic finding in conjunction with peripheral eosinophilia.

Keywords: Bilateral post auricular lymphadenopathy, Eosinophilia, Histopathology, Kimura's disease, Immunosuppressant's

INTRODUCTION

KD is a rare, benign disorder associated with chronic inflammation of unknown etiology.[1] It usually presents as subcutaneous mass in the head and neck region and is frequently associated with regional lymphadenopathy or salivary gland involvement.[2]

KD is rare in India, only 200 cases have been reported worldwide since its histopathological diagnosis.[3] It is generally seen in young adults, with most patients being in the age group of 20 and 40 years; men are affected more commonly than women, with a 3:1 ratio.[4] Peripheral blood eosinophilia and elevated serum IgE levels are constant features of Kimura's disease.[5] The disease is endemic in Asians, but occurs sporadically in other racial groups. Coexisting renal disease is common, with an incidence ranging from 10% to 60%.[6,7] The diagnosis of KD is often difficult, and the biopsy or excision of the involved mass for a histopathological study is necessary. Here we report a rare case of a 20-year old male with bilateral post auricular swelling.

CASE REPORT

A 20 years old Indian male presented with the complaints of swelling behind both ears, which was gradually increasing in size since 14 years, insidious, gradually progressive, painless and associated with neither pruritis nor dermatitis. No history of similar swellings in other parts of the body.

Clinical examination revealed on inspection distinct 4×2 cm ovoid, solitary swelling in the bilateral post auricular region with smooth surface with no visible pulsations. Skin over them was normal. On palpation there was no local raise of temperature, no tenderness, all inspectory findings confirmed, firm in consistency with normal overlying skin, mobile in both horizontal and vertical directions, non reducible, non fluctuant, non transparent (Figure 1). Auscultation, no bruit heard.

There were no other neck nodes palpable. There was neither axillary nor inguinal lymphadenopathy. The rest

of the general physical as well as systemic examinations were normal.

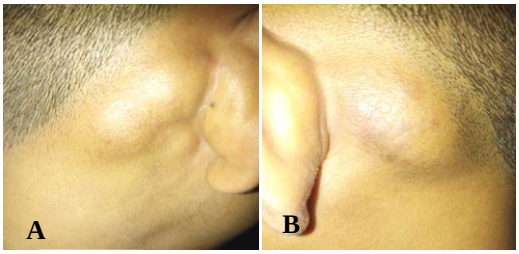

Figure 1: Bilateral post auricular regions showing ovoid, 4×2 cm swelling behind (A) the right pinna and (B) left pinna.

Investigations revealed marked peripheral eosinophilia and normal renal function (Table 1). Fine needle aspiration cytology (FNAC) of post auricular swellings was done which showed reactive lymphadenitis. Sputum samples for tubercular bacilli were negative. A chest radiograph and ultrasonography abdomen and pelvis were normal. Routine and microscopic examination of urine was within normal limits. X-ray of bilateral mastoids showed normal study. Audiological evaluation of both ears was within normal limits.

Table 1: Panel of investigations.

Investigation	Result
CBC	
Haemoglobin	14.5 gm/dl
TLC	9,500 cells/cumm
Neutrophil	32%
Lymphocyte	42%
Eosinophil	24%
Monocyte	2%
AEC	2,280 cells/cumm
Platelets	2.1 lakhs/cumm
RFT	
Blood urea	40.0
Serum creatinine	1.0

Patient underwent excision of bilateral swelling under local anesthesia and specimen was sent for histopathological examination (Figure 2). Microscopy showed thickened capsule with marked hyperplasia of follicles, and paracortical region with thickened blood vessels and infiltration of eosinophils with occasional formation of eosinophilic abscess which are on impinging on germinal centre (Figure 3). No Reed-Sternberg cells identified. Hence all the above features were suggestive of kimura's disease. Postoperative wound healed well (Figure 4) and subsequent follow ups were uneventful. Treatment with methyl prednisolone (24 mg/day) and leflunomide (20 mg/day) was given for 1 month. Laboratory test revealed no improvement in eosinophil count. Hence same treatment was continued for 4 months with observation of decrease in eosinophil count 10%. Methylprednisolone was then tapered by 4mg/day every 2 weeks to a maintenance dose of 8mg/day.

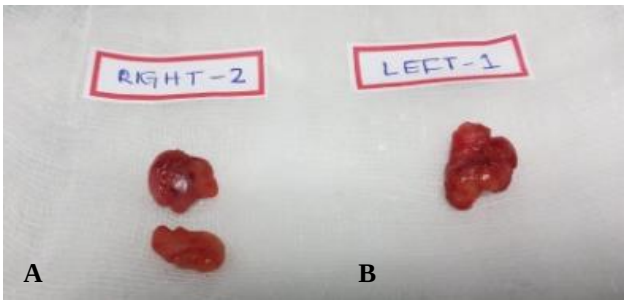

Figure 2: Gross specimen, (A) two grey white nodular lymph nodes, 2×1.8×0.8 cm and 2×1.2×0.5 cm respectively, (B) a grey white lymph node 3×1.5×0.5 cm.

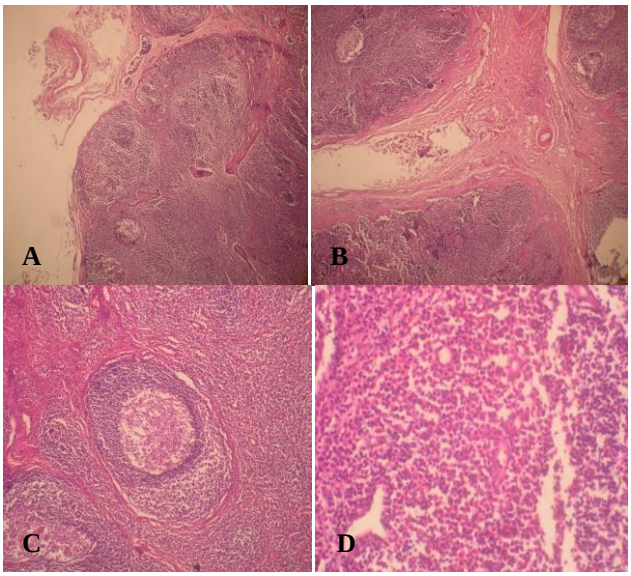

Figure 3: (A) Section of lymph node, scanner view: hyperplasia of follicles, (B) thickened capsule with marked hyperplasia of follicles, (C) paracortical region, infiltration of eosinophils & thickened blood vessels, (D) formation of eosinophilic abscess which are impinging on germinal centres.

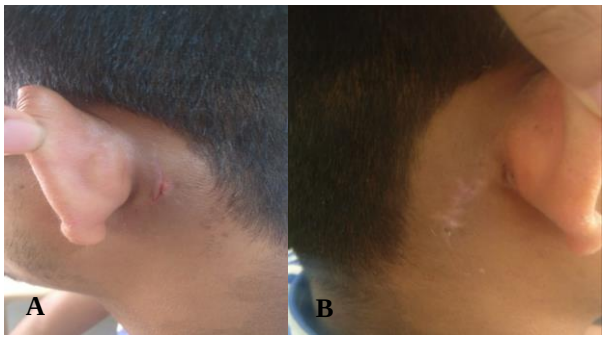

Figure 4 (A and B): Bilateral post auricular one-week post-operative status.

Complete blood count (CBC), serum IgE, and liver and renal function tests were normal at multiple follow-up visits during the following a year. The patient is currently maintained on methylprednisolone(4mg/day) plus leflunomide (10 mg/day) and has remained well without recurrence of symptoms.

DISCUSSION

KD was first described, in 1937, in the Chinese literature, by ST Kimm and C Szeto, as an eosinophilic hyperplastic lymphogranuloma.[7] This disease got its name after the histological description 'unusual granulation combined with hyperplastic changes of lymphatic tissue published by Kimura et al. in 1948.[9]

The common mode of clinical presentation is in the form of painless subcutaneous nodules usually found in the head and neck region. These nodules may be associated with pruritis and dermatitis. The condition is commonly associated with regional lymphadenopathy and occasional involvement of the salivary glands (parotid and submandibular glands). It is often accompanied by peripheral eosinophilia and markedly elevated serum IgE levels.[10]

Although eosinophilia and increased IgE, tumors necrosis factor-alpha, interleukin IL-4, IL 5, IL-13, and mast cell levels in peripheral blood, as well as in the lesion, were observed in these patients, no specific antigens have been identified.[11] It has been proposed that autoimmunity, allergy, neoplasm's, and parasite infestation are possible risk factors. FNAC is useful as an initial investigation of KD, with the main cytological features of high number of eosinophils in a background of lymphoid cells. Nevertheless, histopathology examination of excised lesion is required for a definitive diagnosis as cytology may sometimes be difficult to interpret. The histological picture with the formation of lymphoid follicles and intense aggregates of eosinophils (which sometimes form micro abscesses), vascular proliferation, and fibrosis are highly suggestive of KD.[12,13]

Differential diagnosis of KD would include angiolymphoid hyperplasia with eosinophilia (ALHE), Kikuchi disease, Mikulicz's disease, and most importantly Hodgkin and non-Hodgkin lymphoma.

Even though KD is a benign process, its treatment is not well established. Surgical excision, radiotherapy, steroids, anti-allergic drugs, and cytotoxic drugs have all shown varying results. The side effects of steroids and cytotoxic drugs made them not appropriate for long-time use, especially for young patients with fertility requirements, which motivated us to find other effective immunosuppressants with fewer side effects.

By now, only one report was found about the treatment of leflunomide in KD, which showed that leflunomide was effective in a KD patient with renal involvement.[14] Leflunomide is a novel immunosuppressant approved for rheumatoid arthritis and it is also reported to be effective in treating lupus nephritis.[15-17] upon entering into the human body, leflunomide is quickly metabolized to the active metabolite A77 1726, which in turn inhibits pyrimidine synthesis in activated lymphocytes by suppressing dihydroorotate dehydrogenase. A77 1726 inhibits the proliferation of activated T and B lymphocytes and down regulates immunoglobulin production.[17] Since KD is a chronic inflammatory condition with markedly elevated serum IgE levels, leflunomide may have been effective in this KD patient due to a combination of anti-inflammatory and immunosuppressive effects, as well as by inhibiting IgE production.[18]

Major side effects of leflunomide include leucopenia increased hepatic enzymes and gastrointestinal symptom (diarrhea, nausea, indigestion, etc).[18] No severe side effects were found during the follow-up periods in our patient.

Recurrence rate is as high as 40% despite early treatment of the patients; however there is no consensus for the treatment of recurrent disease with the overall outcome is good as there is no association with malignancy.[19,20]

CONCLUSION

KD is one of the rarest diseases. However, in all patients presenting with post aural swelling with eosinophilia, KD should be considered as a differential diagnosis to avoid unnecessary diagnostic workup. As a cervicofacial swelling that mimics malignancy, it is of high importance that otorhinolaryngologists be aware of this condition to provide early and appropriate treatment. Although prognosis is good, KD is notorious for recurrence and a complete cure may not always be attainable.

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