Case Report

Rare Encounter Behind the Ear: Unveiling Kimura Disease in the Postaural Region

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ABSTRACT

Introduction

Kimura disease, a rare chronic inflammatory disorder primarily impacting subcutaneous tissue, particularly affects young Asian males, with around 200 reported cases in India. Its etiology, linked to an abnormal allergic reaction, remains uncertain, featuring a characteristic indolent growth pattern. The condition manifests as swelling and lesions in the head and neck region, involving subcutaneous soft tissue, major salivary glands, and lymph nodes. Eosinophilia and elevated serum immunoglobulin E levels are common, and diagnosis relies on biopsy. Given its benign nature, optimal management lacks consensus.

Case Report

Presented is a case of Kimura disease in the postauricular region, treated with surgical excision under local anesthesia. Post-operative steroid therapy resulted in an absence of recurrence and reduced eosinophilia during follow-up.

Discussion

Managing Kimura disease is challenging due to its rarity and diverse clinical presentations. Treatment options encompass systemic steroids, antihistamines, immunosuppressants, chemotherapy, radiotherapy, and surgical excision. Despite interventions, the disease often locally recurs. Our case highlights the efficacy of surgical excision under local anesthesia, coupled with post-operative steroids, in preventing recurrence and reducing eosinophilia. Ongoing research and case reports are crucial for establishing standardized protocols in Kimura disease management.

Keywords

Kimura disease; Postauricular Region; Eosinophilia; Surgical Excision

Kimura disease is an uncommon benign disorder associated with chronic inflammation of unknown etiology.[1]

In 1937, ST Kimm and C Szeto documented Kimura disease in Chinese literature, referring to it as eosinophilic hyperplastic lymphogranuloma.[2] This disease got its name as Kimura disease after the histological description of "unusual granulation combined with hyperplastic changes of lymphatic tissue" by Kimura et al. in the year 1948.

It is generally seen in young adults, in the age range of 20 to 40 years. The condition predominantly affects men, with a ratio of 3:1 compared to women.[3] Kimura disease presents as a painless swelling of subcutaneous tissue in the head and neck region and may be associated with regional lymphadenopathy or salivary gland involvement. There are few cases reported with systemic manifestations like nephrotic syndrome in Kimura disease, within the range of 10% to 60%.[2] It has a good prognosis without the risk of malignant transformation. Due to its rarity, it is usually not included in differential diagnoses of head and neck pathologies.

A confirmatory histopathological result is essential, as the findings are pathognomonic and consistent with various literature reviews.[4] It is benign, and recurrence is most commonly seen; hence, regular monitoring of the case is very much needed in this disease. Here, we report a rare case of a 40-year-old male with swelling in the left ear.

Case report

A 40-year-old male patient came to our hospital with a 4-year history of swelling in the left ear that is insidious

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in onset, originally peanut in size and gradually progressed to present size, painless, and not associated with itching or any discharge from the swelling. No history of similar swellings in other parts of the body. Clinical examination revealed, on inspection, a 6 x 8 cm single, ovoid swelling seen in the left postauricular region, from the level of the superior part of the helix to the level of the ear lobule, with a smooth surface and no visible pulsations (Fig.1). The skin covering the swelling appears normal, without any visible scars or sinuses. On palpation, all inspection findings were confirmed, revealing a firm consistency, and non-tender swelling with no localized increase in temperature. The swelling is mobile both horizontally and vertically, non-reducible, and does not display any fluctuation. Additionally, the skin over the swelling appears normal characteristics. On examination, no bruit was heard. In both ears, the external auditory canal and tympanic membrane were normal. The remainder of the ear examination was normal. No other swellings or palpable lymph nodes were found in the body. Both side's facial nerves were intact. The general and neurological examinations were normal.

The audiological examination of both ears was within normal limits. Blood investigations showed mild eosinophilia (Absolute Eosinophil count of 1100/ microliter) with normal renal function and elevated serum immunoglobulin E levels. An ultrasound examination revealed no evidence of fluid in the swelling. Computed tomography shows no involvement of underlying structures or bones. Fine-needle aspiration cytology of the swelling showed numerous mature lymphocytes, scattered histiocytes and interdigitating cells with folded nuclei along with scattered eosinophils, suggestive of reactive lymphadenitis (Fig. 2).

Surgical Procedure - The patient underwent excision of the swelling through a post aural approach under local anaesthesia**,** with a corrugated drain kept for 48 hours. Excess skin tissue was surgically excised, and the resulting wound underwent primary closure using silk sutures and a pressure bandage dressing was applied.

Histopathological examination of the swelling - The specimen was submitted for histopathological analysis, revealing a stratified squamous epithelium accompanied by extensive regions of fibrocartilaginous connective tissue. Prominent collections of mature lymphocytes, forming lymphoid follicles, were observed (Fig.3). These lymphoid cells were intermingled with eosinophils, forming focal aggregates. Additionally, mild capillary proliferation was noted. The histopathological finding of infiltration of lymphoid follicles into the subcutaneous tissue supports the diagnosis of Kimura disease.

Postoperatively, the patient was on 60mg of methylprednisolone (1mg/kg/day) in three divided doses for the first 5 days, 40mg in two divided doses for the next 5 days and 20mg once daily for the last 5 days and showed decreased peripheral eosinophilia (Absolute eosinophil count of 450/microliter) in the follow-up.

Post-operative Challenges-The postoperative pictures of the surgical site of the patient are shown in (Fig.4a and Fig.4b). As can be seen in the postoperative pictures (Fig.4a and Fig.4b), there seemed to be an initial challenge in wound healing. Following the surgery, meticulous post-operative care was administered with antibiotics, including regular wound dressing on the third day to ensure cleanliness and reduce the risk of infection. Initially on day 7, interrupted sutures were removed later remaining sutures were removed on the next week. This comprehensive approach to wound management aims to promote optimal recovery and minimize complications by using steroids postoperatively. The patient was followed up for about a year, and no recurrence of the swelling was noted.

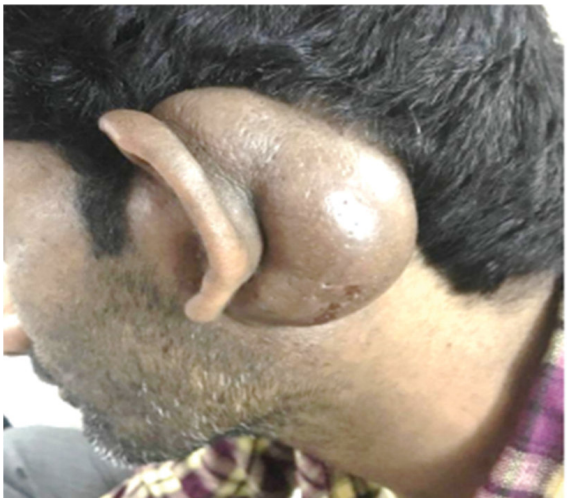

Fig. 1. Postaural swelling in the left ear

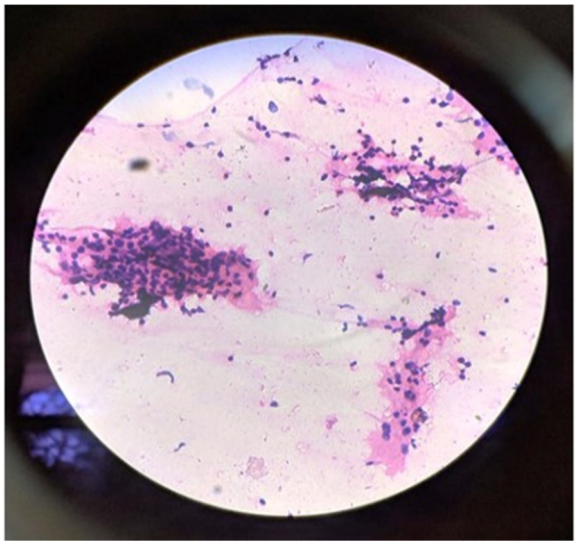

Fig. 2. Fine Needle Aspiration Cytology Examination of the swelling using eosin and hematoxylin stain at 10x magnification showed numerous mature lymphocytes, scattered histiocytes and interdigitating cells with folded nuclei along with scattered eosinophils.

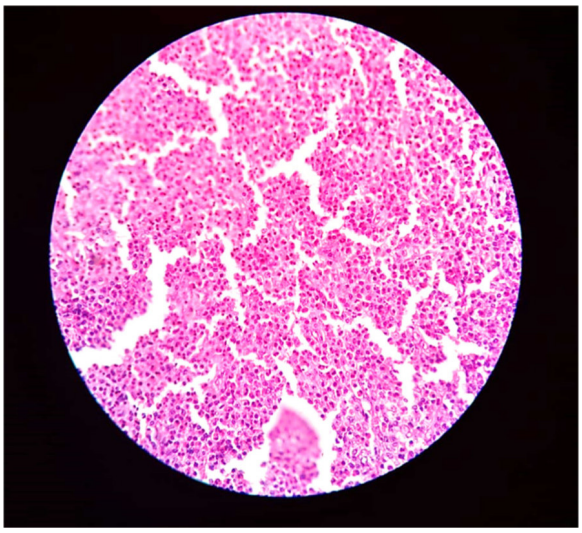

Fig. 3. Histopathological examination of the specimen using eosin and hematoxylin stain at 10x magnification showed prominent collections of mature lymphocytes, forming lymphoid follicles. Lymphoid cells were intermingled with eosinophils, forming focal aggregates.

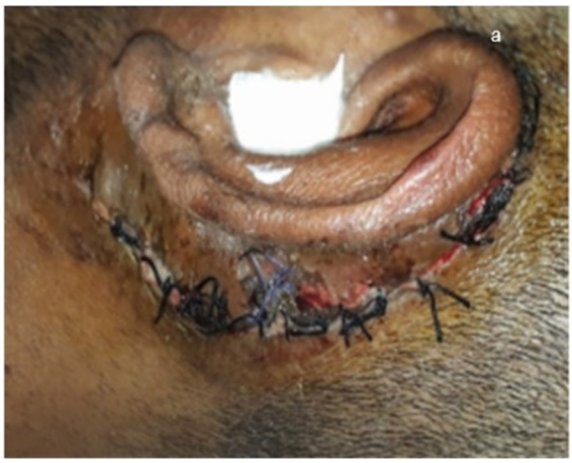

Fig. 4a. Shows the surgical site on the postoperative day 7 and

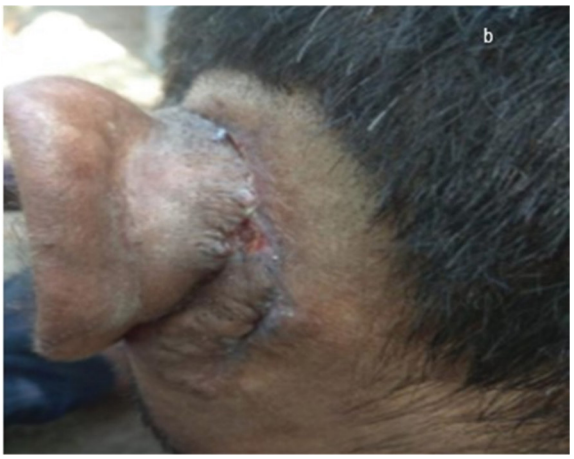

Fig. 4b. Shows the surgical site on the postoperative day 14

Discussion

Kimura disease is a rare, idiopathic, chronic inflammation of subcutaneous tissue with or without regional involvement. The classical sign of Kimura disease is peripheral blood eosinophilia, an increase in serum immunoglobulin E levels,[5] swelling in the subcutaneous tissue, involving most commonly the head and neck region, or it can be associated with regional lymphadenopathy and occasional involvement of salivary glands. Although eosinophilia and increased immunoglobulin E levels are seen, interleukin 4,5,13, mast cells, and tumour necrosis factor-alpha without specific antigens have been

identified.[6] Autoimmunity, allergies, neoplasms, and parasite infestation are possible risk factors for Kimura disease.

Fine needle aspiration cytology is the initial investigation of Kimura disease, showing eosinophils with a background of lymphoid cells. Histopathological examination is the definitive diagnosis of the disease, showing lymphoid follicles with eosinophil aggregates, vascular proliferation, and fibrosis.[7] Ultrasound and Computed tomography scans help to determine the extent of the disease. Computed tomography scans may be non-specific for Kimura disease.[8]

Surgical excision, radiotherapy, steroids, anti-allergic drugs, and cytotoxic drugs like cyclosporine, oral pentoxifylline, and all-trans retinoic acid in combination with oral steroids[9] are the treatment options available. Surgical excision is a widely used therapy, but relapses are frequent. Steroids and cytotoxic drugs for long-term usage have side effects, especially in young patients. Few reports show leflunomide's effectiveness in Kimura disease patients with renal involvement.[10] Tomizuka et al. highlighted the relationship between juvenile temporal arteritis and Kimura's disease.[11]

Differential diagnoses include angiolymphoid hyperplasia with eosinophilia, Kikuchi disease, Mikulicz's disease, and Hodgkin's and non-Hodgkin's lymphomas. The prognosis of Kimura disease is good, with no potential for malignant transformation. It is chronic and may persist or recur despite treatment. Failing to identify and treat the disease may result in the formation of significant and disfiguring lesions. The recurrence rate is high, about 40%, despite early treatment. Treatment of recurrence and overall outcome is good, as there is no association with malignancy. In our case, the timely detection, surgical removal, and post-operative follow-up with steroid therapy contributed to the absence of recurrence and a reduction in eosinophilia.

Conclusion

Kimura disease is an uncommon, chronic inflammatory condition of the subcutaneous tissue, characterized by unknown causes. Accurate clinical diagnosis, coupled with histopathological assessment, plays a crucial role in effectively managing this condition. Timely identification and treatment are essential in addressing the extensive and aesthetically challenging lesions associated with Kimura disease. Although recurrence rates are notable, the prognosis is generally favourable, as there is no established link between the disease and malignancy. In our case, the timely detection, surgical removal, and post-operative follow-up with steroid therapy contributed to the absence of recurrence and a reduction in eosinophilia.

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