CASE REPORT

Kimura's disease: A case presentation of postauricular swelling

A Rajesh, T Prasanth, VC Naga Sirisha, MDS Azmi

Department of Oral and Maxillofacial Pathology, CKS Theja Institute of Dental Sciences and Research, Tirupathi, Andhra Pradesh, India

Abstract

Kimura's disease (KD) is a rare chronic inflammatory disease of subcutaneous tissues and occurs predominantly in head and neck region. It is seen primarily in young Asian males. Typical clinical presentations are painless subcutaneous masses, regional lymph node enlargement, blood and tissue hypereosinophilia, and increased serum IgE levels. Here, we present a case of a 27-year-old female who presented with unilateral single nodular swelling in the right postauricular region. The diagnosis of KD was done based on characteristic histopathologic finding in conjunction with peripheral eosinophilia and increase in serum IgE levels.

Key words: Angio lymphoid hyperplasia with eosinophilia, eosinophilia, Kimura's disease, serum IgE

Date of Acceptance: 28-May-2015

Introduction

Kimura's disease (KD) is a rare chronic inflammatory disease, which was first described in 1937 by Kim and Szetu in the Chinese literature as eosinophilic hyperplastic lymphogranuloma. The disease came to be known as KD since its description by Kimura *et al.* in Japanese literature in 1984.[1-4] It is endemic in China and Japan, although sporadic cases have been reported elsewhere.[5] KD is rare in India, with about 200 reported cases worldwide since its histopathological diagnosis.[6] The disease is characterized by the triad of painless subcutaneous masses in head and neck, blood and tissue eosinophilia, and increased serum IgE levels. Recent studies have shown that KD occasionally show the clonal proliferation of T-cells.[7] It affects young men between 20 and 40 years of age with a male: female ratio = 3:1.[11] The diagnosis of KD is often difficult, and biopsy/excision of involved mass are necessary.

Case Report

A 27-year-old female patient reported to us with a chief complaint of swelling behind the right ear since 6 months.

Address for correspondence:
Dr. MDS Azmi,
Department of Oral and Maxillofacial Pathology, CKS Theja Institute of Dental Sciences and Research, Chadalawada Nagar, Renigunta Road, Tirupathi, - 517 501, Andhra Pradesh, India.
E-mail: dr.saniashaik@gmail.com

On examination, a small nodular well-circumscribed swelling in the right postauricular region was observed. It is asymptomatic, and the surface of swelling was normal and no local rise in temperature was observed [Figure 1].

On palpation the swelling measured 2 cm × 2 cm in size, nodular with well-defined borders, rubbery to firm in consistency [Figure 2]. The patient was subjected to excisional biopsy with a provisional diagnosis of lipoma, and the specimen was sent for histopathological examination.

The macroscopic features showed the specimen measuring about 1.5 cm × 0.8 cm × 0.8 cm in size, creamish to brownish in color, soft to firm in consistency with regular borders.

The histopathology showed lymphoid tissue with reactive follicular hyperplasia, eosinophilic deposits within germinal centers, and mixed inflammatory infiltrate predominantly of eosinophils seen. Later blood investigations were done and peripheral blood smear revealed eosinophilia and

Access this article online

Quick Response Code:

Website: www.njcponline.com

DOI: 10.4103/1119-3077.164361

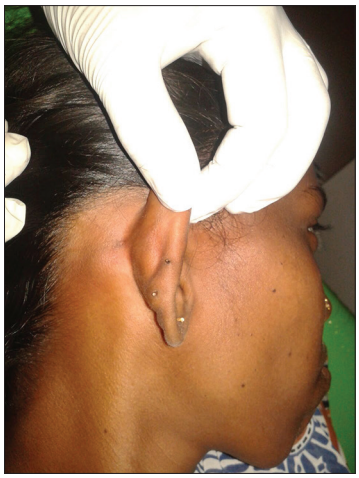

Figure 1: A small nodular well circumscribed swelling in right postauricular region

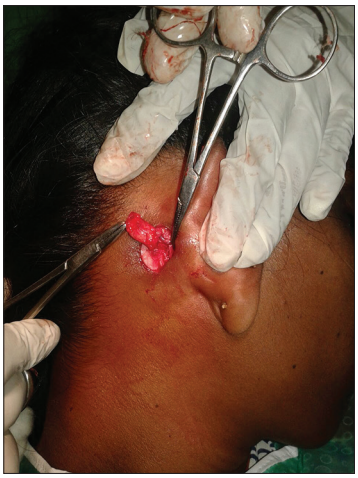

Figure 2: Intraoperative photograph showing well-circumscribed swelling, 2 cm × 2 cm in size with well-defined borders

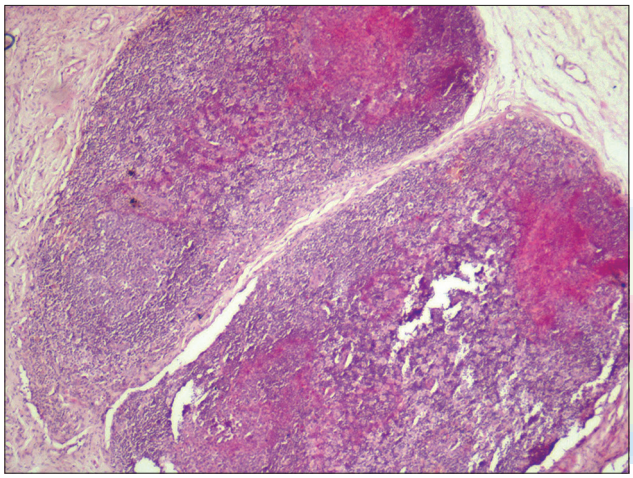

Figure 3: H and E, ×10 views, showed well-encapsulated lesion showing hyperplastic lymphoid follicles with prominent germinal centers

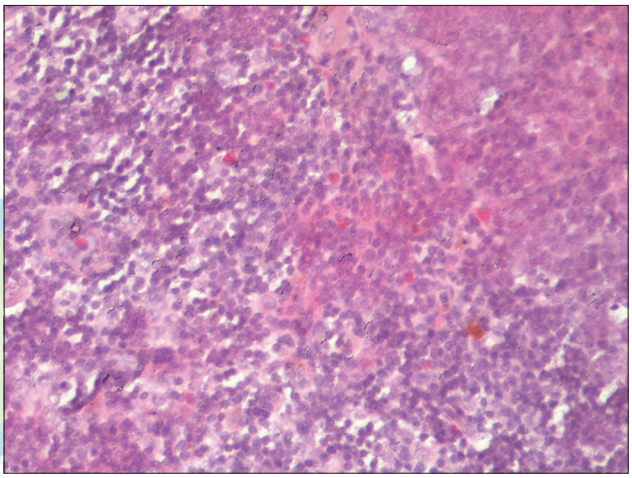

Figure 4: H and E, ×40 views, presence of eosinophils within the germinal centers and interfollicular area admixed with neutrophils, lymphocytes, and plasma cells. Multinucleated giant cells are seen interspersed in between

increase of serum IgE concentration. Based on the clinical, laboratory, and histopathologic findings, a final diagnosis of KD was made.

Discussion

Kimura's disease was first described in 1937 by Kim and Szetu in the China as eosinophilic hyperplastic lymphogranuloma but is more widely known as KD since its first description by Kimura *et al.* in Japanese literature in 1984. He reported similar cases under the title "On the unusual granulation combined with hyperplastic changes of lymphatic tissue." Over the years, there has been considerable confusion between KD and angiolymphoid hyperplasia with eosinophilia (ALHE). The confusion escalated by the introduction of an ever-expanding number of names applied to both diseases, including eosinophilic granuloma, eosinophilic granuloma of lymph node and soft-tissue, eosinophilic hyperplastic lymphogranuloma, eosinophilic lymphofollicular granuloma, eosinophilic lymphfolliculoid granuloma of the soft tissue, atypical pyogenic granuloma, inflammatory angiomatous nodules, histiocytoid hemangioma, epithelioid hemangioma, subcutaneous angioblastic lymphoid hyperplasia with eosinophilia, and subcutaneous ALHE.[1]

In 1979, the first good histological dissection of the lesions was reported, in which KD was separated from ALHE, the latter classified under the group of histiocytoid hemangioma.[1]

Kimura's disease is considered much more prevalent in young Asian lineage. There is a marked predominance of male patients; the male/female ratio is 2:1–5:1 and the onset of the disease occurs mostly in the third decade of life.[8,9] It occurs predominantly unilaterally as subcutaneous nodules in the head and neck, and it is frequently associated with

regional lymphadenopathy with or without the involvement of salivary glands.[3] Bilateral involvement is also seen sometimes.[8] Majority of the cases and those reported in the literature occurred in the head and neck, particularly infra auricular and retro auricular region. Orbit, eyelid, palate, and pharynx have also been reported, in addition to the axilla, groin, and arm.[1,8,9] KD may affect kidneys in up to 60% of patients. In those cases, it may present itself as glomerulonephritis and nephritic syndrome. Most common renal involvement occurs as membranous glomerulonephritis.[8,10] Cases with systemic clinical signs have been described in the form of pruritus, eczema, rashes, etc., but this is less common.[10] In our case, the patient was completely asymptomatic.

The delineating histologic features of the lymph nodes in KD outline constant, frequent, and rare histologic parameters. The constant features include preserved nodal architecture, florid germinal center hyperplasia, eosinophilic infiltration, and postcapillary venule proliferation. Our case showed lymphoid tissue with reactive follicular hyperplasia [Figure 3], eosinophilic deposits within the germinal centers, and interfollicular area. Giant cells (Warthin–Finkeldey) were also seen [Figure 4]. Other features are the proliferation of small blood vessels, many of which are lined by enlarged endothelial cells with uniform ovoid nuclei and intracytoplasmic vacuoles. These endothelial cells have been described as having a cobblestone appearance. Frequent features include sclerosis, vascularization of the germinal centers, proteinaceous deposits in the germinal centers, necrosis of the germinal centers, eosinophilic abscess, and a reticular IgE deposition within germinal centers. Although nodal architecture is largely preserved in most cases, capsular fibrosis with subcapsular sinusoid obliteration and perinodal soft-tissue involvement is frequently present. The presence of fibrosis, in general, is a valuable feature in distinguishing KD from ALHE. Eosinophilic folliculosis, a helpful feature in diagnosing KD, often coexists to some degree with eosinophilic deposits in the germinal center and germinal center necrosis. Small clusters of giant cells or small discrete eosinophilic necrosis with or without surrounding epithelioid histiocytes may also be seen sometimes. In a study done by Hong Chen *et al.*, one case showed a crystalline structure, resembling Charcot–Leyden crystal, within the cytoplasm of a number of histiocytes, a unique finding, when referring to phagocytosis of eosinophilic granules by tissue histiocytes. It was hypothesized that eosinophils infiltrate the germinal center, which undergoes folliculosis, resulting in an eosinophilic necrosis with or without an associated epithelioid histiocytic reaction. This contention is further strengthened by a recent report in which eosinophilic epithelioid granulomatous reactions were produced by phagocytosis of the apoptotic eosinophils by macrophages.[1]

The differential diagnosis of KD is broad and includes Hodgkin lymphoma, angio immunoblastic T-cell lymphoma, langerhans cell histiocytosis, florid follicular hyperplasia, Castleman's disease, dermatopathic lymphadenopathy, ALHE, lymphadenopathy of drug reactions, and parasitic lymphadenitis.[1] Absence of reed sternberg cells, langerhan cells, and prominent arborising vessels histopathologically excludes them from other differential diagnosis such as lymphomas, langerhans cell histiocytosis, ALHE. There was no drug history and other infections.

The etiopathogenesis of this rare disease remains unknown. It has been proposed that a common trigger factor such as viral, parasite infection or a toxin could alter T-cell immunoregulation or induce an IgE medicated hypersensitivity resulting in the release of eosinophilia trophic cytokines. The role of cytokines, such as interleukin-4 (IL-4), IL-5, and IL-13, was documented in patients with KD, which may precipitate high IgE levels and marked eosinophilia in this disease.[9]

Therapies for KD include surgical excision, steroid, and radiation, but the treatment of choice is still controversial. The effect of systemically administered steroids (prednisolone) show good effects on disease progression; however, withdrawal can often result in tumor relapse.[8] Radiation has been utilized for steroid-resistant lesions.[3] Besides, the treatment of KD with cyclosporine has also been reported.[9] Surgical excision may be considered first especially for the localized lesion, even if recurrence is possible, however, radical surgery should be avoided since no malignant transformation has been reported in the literature.[2,12]

Conclusion

Kimura's Disease is a rare chronic inflammatory disease of subcutaneous tissues commonly occurring in males. Although it is a rare disorder, it should be considered in the differential diagnosis of any lymph node demonstrating an eosinophilic infiltrate and prominent follicular hyperplasia. It is a distinctive clinicopathologic entity with characteristic histologic features and is important to separate from drug reactions, hypersensitivity, and infectious agents. We present this case in an Indian female so as to add another case to the literature.

References

1. Chen H, Thompson LD, Aguilera NS, Abbondanzo SL. Kimura disease: A clinicopathologic study of 21 cases. Am J Surg Pathol 2004;28:505-13.
2. Kimura T, Voshimura S, Ishikawa E. On the unusual granulation combined with Hyperplastic changes of Lymphatic tissues. Trans Soc Pathol Jpn 1948;37:170-80.
3. Patel CS, Dave RV. Kimuras disease: A case report. Int J Dent Clin 2011;3:115-6.

4. Osuch-Wójcikiewicz E, Bruzgielewicz A, Lachowska M, Wasilewska A, Niemczyk K. Kimura's disease in a Caucasian female: A very rare cause of lymphadenopathy. Case Rep Otolaryngol 2014:1-4.
5. Baidya DK, Chhabra A, Raj R. Anesthetic management of a patient with Kimura's disease for superficial parotidectomy. J Anaesthesiol Clin Pharmacol 2011;27:383-5.
6. Abhay H, Swapna S, Darshan T, Vishal J, Gautam P. Kimura's disease: A rare cause of local lymphadenopathy. Int J Sci Study 2014;2:122-5.
7. Rao K, Kumar S. Kimura's disease – A rare cause of head and neck swelling. Int J Otolaryngol Head Neck Surg 2014;3:200-4.
8. Priyokumar OS, Niteshore MS, Ratan K. Bilateral Kimuras disease with sinusitis and parotid gland involvement: A case report and a brief review of literature. IOSR J Dent Med Sci 2014;13:22-5.
9. Chen YM, Chen CH, Chen HH, Chen YH, Chen DY, Lan JL. Clinical manifestations, therapeutic response and disease outcome of Kimura's disease in Taiwan. Formos J Rheumatol 2009;23:33-9.
10. López-Arcas Calleja JM, Iturriaga TM, Romero MP, de María Martínez G, Martín-Moro JG, del Castillo JL. Kimura disease: Report of a case in a Caucasian male and revision of the literature. Rev Esp Cir Oral Maxilofac 2006;28:353-8.
11. Pastapur R, Ansari A, Bohra RB, Vare R, Mathew SM. Kimura disease – A rare case. J Evol Med Dent Sci 2013;2:8860-3.
12. Ragu R, Eng JY, Azlina AR. Kimura's disease of the parotid: A complete clinical-radiological-pathology report. Med J Malaysia 2014;69:199-201.

How to cite this article: Rajesh A, Prasanth T, Naga Sirisha VC, Azmi M. Kimura's disease: A case presentation of postauricular swelling. Niger J Clin Pract 2016;19:827-30.

Source of Support: Nil, **Conflict of Interest:** None declared.