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Case report

Kimura disease – A case report and review of the literature



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ABSTRACT

Introduction: Kimura disease (KD) is a benign chronic inflammatory disorder attributed to an immune mediated hypersensitivity. KD is commonly presented with unilateral subcutaneous tissue swelling in the head and neck area. The course is usually benign except for the potential cosmetic disfigurement. There is no consensus for the treatment of recurrent disease.

Aim: To illustrate a case of an uncommon cause of head and neck swelling.

Case study: A 41-year-old male presented with a recurrent painless swelling at his of right lower cheek swelling for 6-months. Head and neck examination revealed a 7 × 7 cm firm, non tender mass over the right mandibular region. Other systemic examination was unremarkable. Magnetic resonance imaging of the mass showed a well defined lesion 6 × 3 × 5 cm in size anterior to the right body of mandible. Surgical excision of the mass was done and histopathological examination of the specimen showed fibrocollagenous and fibrofatty tissue infiltrated by chronic inflammatory cells in the pattern of perivascular lymphoid aggregates. The infiltrate is composed predominantly of small lymphocytes and eosinophils.

Results and discussion: Here we described a rare cause of painless subcutaneous head and neck swelling that occurred in a middle-aged Asian man. KD usually presents as a painless subcutaneous soft-tissue swellings and associated with regional lymphadenopathy in the head and neck region. Diagnosis of KD is always a clinical dilemma with no specific diagnostic guideline. Though there is no consensus for the treatment of recurrent disease, the overall outcome is good as there is no association with malignancy.

Conclusions: KD should be considered in the differential diagnosis of a recurrent head and neck mass.

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1. Introduction

Kimura disease (KD), a benign chronic inflammatory disorder, was first reported by Kimm and Szeto in 1937 in China.¹ In 1948, a Japanese doctor named Kimura et al. published a systemic description of the disease and formally coined it as 'Kimura disease.'²

The most frequent clinical presentation is unilateral subcutaneous mass in the head and neck region with a predilection for post-auricular area.³ The exact cause of chronic inflammation in KD is uncertain. However, some attributed it to a hypersensitivity reaction or immune mediated response toward environmental agents.⁴ Although the lesion has no potential for malignant transformation,⁵ it can grow and cause functional and cosmetic disfigurement.

2. Aim

The aim of this case report was to enhance physicians' awareness on KD to ensure its optimal management for the patients.

3. Case study

The case refers to a 41-year-old male complaining of right lower cheek swelling for 6-months duration. The swelling was painless, slowly growing but associated with mild itchiness. There were no symptoms to suggest pulmonary tuberculosis such as chronic cough, night sweats, anorexia or loss of weight. On further history, he had previous recurrent swelling at the same site, which resolved on initiation of a course of oral prednisolone. On general physical examination, he had stable vital signs: his temperature was 37°C, heart rate of 60 bpm and respiratory rate of 18 breaths/min. Head and neck examination revealed a 7 × 7 cm swelling over the right mandibular region, which was firm, non-tender and non-adherent to the skin. But there was no associated cervical lymphadenopathy. Other systemic examination was unremarkable. Initial laboratory investigations revealed normal indices of full blood count with platelet count of $228 \times 10^9/L$, hemoglobin of 14.3 g/dL and total white cell count of $6.8 \times 10^9/L$, except a slightly higher eosinophils count of 0.5 g/dL. Peripheral blood film also showed eosinophilia. Erythrocyte sedimentation rate was 17 mm/h. Serum immunoglobulin E (IgE) level was not performed for this patient. The results of serum urea, creatinine and microscopic urinalysis and also chest radiography were normal. Magnetic resonance imaging (MRI) of the mass showed a well defined lesion 6 × 3 × 5 cm in size anterior to the right body of mandible. There was no evidence of bony and lymph nodes involvement. Subsequently, surgical excision of the mass was done under general anesthesia. Histopathological examination of the specimen showed fibro-collagenous and fibro-fatty tissue infiltrated by chronic inflammatory cells in the pattern of perivascular lymphoid aggregates (Fig. 1). The infiltrate was composed predominantly of small lymphocytes and eosinophils (Fig. 2). Some of the lymphoid aggregates had prominent germinal centers. These

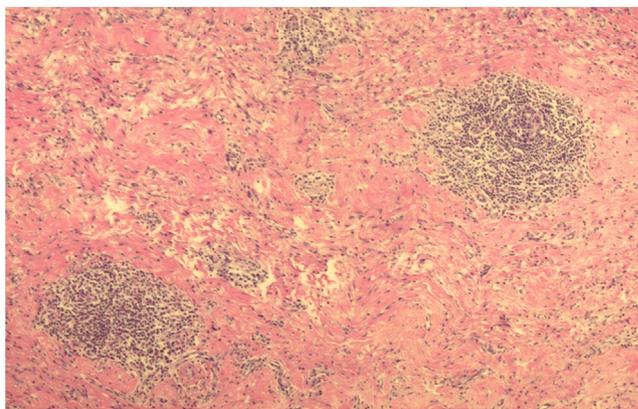


Fig. 1 – Presence of lymphoid aggregates arranged at perivascular area.

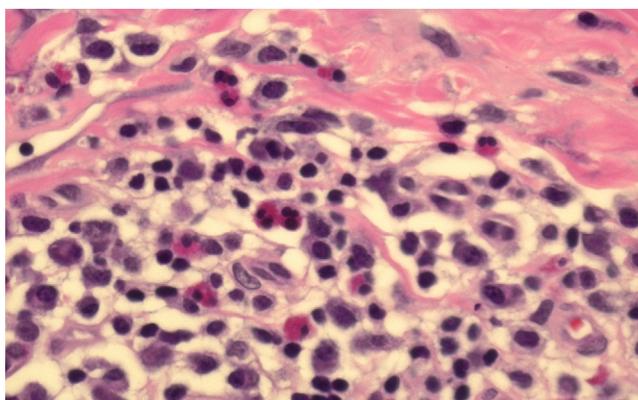


Fig. 2 – Aggregate of lymphocytes and scattered eosinophils within the follicle.

findings confirmed the diagnosis of KD. The patient's recovery was uneventful and he was discharged well.

4. Results and discussion

Here we described a rare cause of painless subcutaneous head and neck swelling that occurred in a middle-aged Asian man. KD, also known as eosinophilic lymphogranuloma, is a rare chronic inflammatory disorder with unknown etiology and pathogenesis.^{4,6} However, the presence of eosinophilia and increased serum levels of IgE, mast cell, interleukin (IL)-4, IL-5 and IL-13 in patients suggests an etiology of either an unusual autoimmune response or an immune reaction toward a parasitic infestation.⁷

The exact prevalence of KD is still undetermined,⁸ but most of the cases typically occurs in young and middle-age men originating from East Asia and South East Asia.⁹ For instance, two different studies done in Korea and China found similar male predominance of about 87% with mean age of late 1920s.^{6,10} In contrast, the incidence of KD amongst the Caucasian ethnicity is extremely rare, with the cases reported being sporadic with atypical presentation.^{11–13}

KD usually presents as a painless subcutaneous soft-tissue swellings and associated with regional lymphadenopathy in the head and neck region.^{12,14} It is also known to manifest in other sites including the limbs, groin and trunk.¹⁵ Occasionally, lymphadenopathy may be the only manifestation of the disease. Although the chronic inflammation is usually localized to the affected subcutaneous area without a systemic spread, there is a remarkable rise in serum IgE and eosinophilia in most patients. Intriguingly, nephropathy is associated with KD in about 12%–16% of patients, while the cause is unclear.⁷

Diagnosis of KD is always a clinical dilemma with no specific diagnostic guideline. Clinically, among other differential diagnosis of KD would include Kikuchi disease, Mikulicz's disease and most importantly Hodgkin and non-Hodgkin lymphoma. Fine needle aspiration cytology (FNAC) is useful as an initial investigation of KD, with the main cytologic 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.¹⁶ Similar to the histopathological findings in our patient, KD is commonly characterized by follicular hyperplasia with reactive germinal centers, micro-abscesses, fibrosis and proliferation of post-capillary venules.¹⁷

There is no consensus or standardized treatment protocol of KD. Therefore, its management involves various medical and surgical subspecialties including ear, nose and throat (ENT), dermatology, plastic surgery and oncology. Treatment options for KD include surgical resection, cytotoxic and radiation therapy,¹⁸ as well as regional or systemic steroids therapy.¹⁹ The role of surgery is mainly for diagnostic and cosmetic purposes. Systemic corticosteroids may be used with or without other treatment modalities. Treatment with intralesional corticosteroid (triamcinolone acetonide) had been reported to have good outcome.¹³

Relapses are common and recurrence rate is as high as 40% despite early treatment of the patients.²⁰ Though there is no consensus for the treatment of recurrent disease, the overall outcome is good as there is no association with malignancy.

5. Conclusions

Although unusual, KD should be considered in the differential diagnosis of a recurrent head and neck subcutaneous mass. FNAC is a helpful tool, but histopathology is necessary for diagnosis. The modality of treatment mainly depends on preserving function and cosmetics of the tissues affected. While prognosis can be good, a complete cure is sometimes impossible.

Conflict of interest

None declared.

REFERENCES

1. Kimm H, Szeto C. Eosinophilic hyperplastic lymphogranuloma, comparison with Mikulicz's disease. *Proc Chin Med Soc.* 1937;1:329.
2. Kimura T, Yoshimura S, Ishikawa E. On the unusual granulation combined with hyperplastic changes of lymphatic tissues. *Trans Soc Pathol Jpn.* 1948;37:179–180.
3. Viswanatha B. Kimura's disease in children: a 9 years prospective study. *Int J Pediatr Otorhinolaryngol.* 2007;71(10):1521–1525.
4. Calli C, Oncel S, Pinar E, Bener S, Calli A. Kimura's disease: two case reports with a review of the literature. *Kulak Burun Bogaz Ihtis Derg.* 2004;12(5–6):139–143.
5. Armstrong WB, Allison G, Pena F, Kim JK. Kimura's disease: two case reports and a literature review. *Ann Otol Rhinol Laryngol.* 1998;107(12):1066–1071.
6. Zhang JZ, Zhang CG, Chen ZZ. Thirty-five cases of Kimura's disease (eosinophilic lymphogranuloma). *Br J Dermatol.* 1998;139(3):542–543.
7. Sun QF, Xu DZ, Pan SH, et al. Kimura disease: review of the literature. *Intern Med J.* 2008;38(8):668–672.
8. Kar IB, Sethi AK. Kimura's disease: report of a case & review of literature. *J Maxillofac Oral Surg.* 2013;12(1):109–112.
9. Wang DY, Mao JH, Zhang Y, et al. Kimura disease: a case report and review of the Chinese literature. *Nephron Clin Pract.* 2009;111(1):c55–c61.
10. Kang M, Kim B, Kwon J, et al. Clinical characteristics of Kimura disease in Korea. *J Allergy Clin Immunol.* 2010;125(2):AB185.
11. 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;2014:415865.
12. Nyrop M. Kimura's disease: case report and brief review of the literature. *J Laryngol Otol.* 1994;108(11):1005–1007.
13. Lenk N, Artüz F, Kulaçoğlu S, Alli N. Kimura's disease. *Int J Dermatol.* 1997;36(6):437–439.
14. Li TJ, Chen XM, Wang SZ, Fan MW, Semba I, Kitano M. Kimura's disease: a clinicopathologic study of 54 Chinese patients. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 1996;82(5):549–555.
15. Chen H, Thompson LD, Aguilera NS, Abbondanzo SL. Kimura disease: a clinicopathologic study of 21 cases. *Am J Surg Pathol.* 2004;28(4):505–513.
16. Murthy SV, Geethamala K, Rao SM. Kimura's disease: a cytodifferential dilemma with brief review of literature. *Int Med J Sifa Univ.* 2015;2(3):62.
17. Kuo TT, Shih LY, Chan HL. Kimura's disease. Involvement of regional lymph nodes and distinction from angiolymphoid hyperplasia with eosinophilia. *Am J Surg Pathol.* 1988;12(11):843–854.
18. Fouda MA, Gheith O, Refaie A, et al. Kimura disease: a case report and review of the literature with a new management protocol. *Int J Nephrol.* 2011;2010:673908.
19. Madhavan M, Othman NH, Singh MS, Indudharan R, Sharma HS, Shamsuddin AR. Kimura's disease: a report of three cases with a brief review of literature. *Acta Otorhinolaryngol Ital.* 2000;20(4):284–289.
20. Göçmen H, Oguz H, Astarci M, Samim E. Kimura's disease: case report and brief review of the literature. *J Otolaryngol.* 2006;35(5):358–360.