Case Report

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Kimura disease masquerading tuberculosis: a rare case presentation

Rabin Paneru, MBBS[a], Madalasa Pokhrel, MD[e], Saral Lamichhane, MBBS[b,*], Subash Basnet, MBBS[d], Saroj Poudel, MBBS[c], Sudarshan Gautam, MD[f], Nava Raj Sharma, MBBS[a]

Introduction and importance: Kimura disease (KD) is an inflammatory disorder characterized by the development of subcutaneous lymphoid masses and regional lymphadenopathy. Due to its rarity and similarity to another disease, the diagnosis is complex.

Case presentation: Here, the authors present a case of KD in 26-year-old male from Nepal who initially did not respond to antitubercular therapy. Later on, KD was diagnosed based on histopathology. He was followed up in medical outpatient with a good response to corticosteroid therapy.

Clinical discussion: The diagnosis of KD is quite difficult in low-resource settings. The diagnosis is histopathological. Associated lymphadenopathy may mimic tuberculosis. Many patients respond well to the high-dose of steroid therapy; some might also require surgical excision or chemotherapy.

Conclusion: Hence, the physician should include KD as a differential when a male in his 20s or 30s presents with a subcutaneous nodular mass in the head and neck.

Keywords: eosinophilia, kimura disease, lymphadenopathy, Nepal, tuberculosis

Introduction

Kimura disease (KD) is a chronic inflammatory disorder presenting as subcutaneous lymphoid masses; regional lymphadenopathy; peripheral eosinophilia; and elevated levels of serum IgE[1]. Its clinical presentation includes painless lymphadenopathy or subcutaneous masses in the head and neck region[2]. Other sites of involvement are the oral cavity, axilla, groin, limbs, and trunk[3]. There is frequent regional lymphadenopathy or salivary gland enlargement[4]. KD is a rare disease whose exact prevalence is not well-studied and primarily affects people of Southeast and East Asian descent[1,5,6]. It shows a male predilection of 3:1 and a peak incidence at age 20–40 years[2]. The etiology and pathogenesis of the disease are largely unknown. KD's most important pathological feature is hyperplastic follicles with germinal centers surrounded by abundant eosinophilic infiltrations. Standard treatment for KD is currently not available. The main therapeutic methods are surgical excision, glucocorticoid therapy, cytotoxic therapy, and radiotherapy. The prognosis of the disease is good. However, it has a high local recurrence rate.

Here, we present a 26-year-old male diagnosed with KD from Nepal, who initially did not respond to antitubercular therapy. This case has been reported in line with the Surgical CAse REport (SCARE) 2020 criteria[7].

HIGHLIGHTS

- Kimura disease (KD) is a rare inflammatory disorder affecting lymphoid tissue.
- The diagnosis of KD might be challenging in developing countries where the incidence of tuberculosis is high.
- Treatment of KD is surgical removal along with anti-inflammatory agents.

Case presentation

A 26-year-old male was admitted with a chief complaint of painful swelling on the right side of his neck for 4 months and episodic low-grade fever for more than three months. His recorded maximum temperature was 103 degrees F which last for 3 days. He reported sweating, malaise, anorexia, and a significant weight loss of about 10 kg in the last 4 months. There was no history of tuberculosis, and he does not recall tuberculosis in close contact. The skin overlying the mass was smooth and nonerythematous. There was no history of swelling elsewhere in his body. There was no travel history.

[a]Manipal College of Medical Sciences, [b]Gandaki Medical College, Pokhara, [c]Kathmandu Medical College, [d]Tribhuvan University Institute of Medicine, Kathmandu, Nepal, [e]Montefiore New Rochelle Hospital, New York and [f]Maimonides Medical Center, Brooklyn, USA

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*Corresponding author. Address: Gandaki Medical College, Pokhara 33700, Nepal. Tel.: +9779849484752. E-mail: sarlmc.sl@gmail.com (S. Lamichhane).

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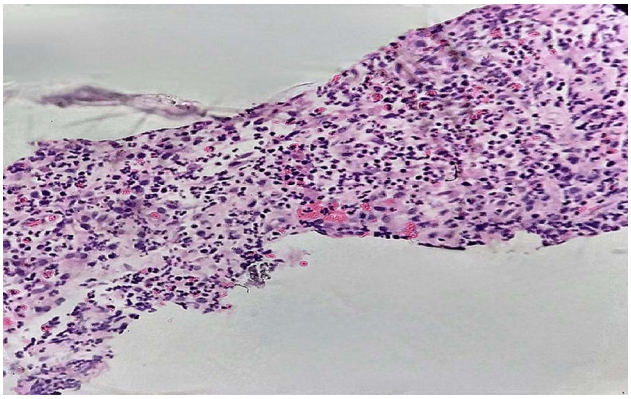

Figure 1. Specimen shows fibrous stromal tissue densely infiltrated by neutrophils, lymphocytes, histiocytes, plasma cells, and eosinophils.

The total leukocyte count was increased with predominant neutrophils. His C-reactive protein was positive. The eosinophil count was within the normal range. The serology was negative for HIV, HCV, and HBV. The Mantoux test was negative. GeneXpert of the biopsy sample was also negative for tubercular bacilli. C-ANCA and P-ANCA were negative. His renal function test and liver function test show no abnormalities. Blood and urine culture grows no organism after 48 h of inoculation.

Chest imaging showed necrotic cervical lymphadenopathy on the right side, a heterogeneously enhancing necrotic lesion in the anterior mediastinum, multiple mediastinal and right hilar lymphadenopathy, nodular opacity, and thickened interlobular septa with fibrotic changes in the right upper lobe with mild pericardial effusion. The left second rib's adjacent muscle, cartilage, and anterior aspect were invaded. Two similar lesions were noted in the right anterior chest wall superior to the lesion mentioned above and the in left lateral aspect of the lesion as discussed above.

Fine-needle aspiration cytology of the right cervical lymph node showed reactive lymphadenitis. In the meantime, the patient was started on intravenous broad-spectrum antibiotics for suspected infective lymphadenitis. Even though his pain and temperature reduced after a few days, the mass was still present with no improvement, even during follow-up. Although acid-fast bacilli were not seen in the microscopy of the fine-needle aspiration cytology sample, with the history and reactive lymphadenitis finding in the microscopy, a clinical diagnosis of lymph node tuberculosis was made during follow-up, and the patient was started on antitubercular therapy. The patient's condition was not improving even after starting antitubercular medicine, and thus the patient was admitted for further investigations and management.

Histopathology of the right cervical lymph node and right axillary tissue and a biopsy of the anterior chest wall was done. The specimens showed fibrous stromal tissue densely infiltrated by neutrophils, lymphocytes, histiocytes, plasma cells, and eosinophils, as shown in Figure 1. Histopathology of the sternal mass showed similar findings. There was the absence of Reed-Sternberg cells, granulomas, or any atypical cells.

Our patient showed clinical and symptomatic improvement with high-dose steroid therapy, so he was followed up in the clinic with drastic improvement during regular follow-ups.

Discussion

Kim and Szeto first described Kimura's disease in China in 1937[2]. The disease was initially known as 'eosinophilic hyperplastic lymphogranuloma' till 1948 and was named Kimura's disease later when its vascular component was observed. Although the exact prevalence of KD is unknown, only about 200 cases of Kimura's disease were reported worldwide till September 2020[1,2]. KD is a rare disease mainly affecting people of Southeast Asian descent. It shows a male predilection of 3:1 and peak incidence at age 20–40 years[3–5,8]. Our case is consistent with the expected age group, sex, and subcutaneous mass characteristics, whereas peripheral eosinophilia and an elevated serum IgE level were not reported. Though the mass is painless primarily in KD, the cervical mass present in our case was painful[4]. The pain and fever in our patient might be due to the infective etiology of cervical lymphadenitis, which was reduced after initial broad-spectrum antibiotics. Other sites of involvement include the oral cavity, axilla, groin, limbs, and trunk. The lymphadenopathy in our case was also observed in the axillary and mediastinal lymph nodes.

In contrast to our case, systemic symptoms like fever, weight loss, sweating, malaise, and anorexia are not common in KD[9]. The masses are primarily unilateral and grow over time. The number of masses found in the lesion is single or multiple. They have a firm consistency, and the overlying skin appears normal. Our patient had multiple lymphadenopathies with normal overlying skin. The renal disease, frequently Nephrotic Syndrome with clinically relevant proteinuria, is often seen in KD[9].

The etiology and pathogenesis of KD are primarily unknown. Due to the presence of peripheral eosinophilia, eosinophilic infiltrate within the masses, and elevated IL-5/IgE levels in the body, some clinicians and researchers believe there is a similarity between KD with atopy[10]. Impairment or interference with immune regulation and neoplasm may also cause this disease. The possibility of KD to be a type of hypersensitivity reaction is indicated by peripheral eosinophilia and the presence of eosinophils in the inflammatory infiltrate. T-helper 2 cells might also play a role in KD[1,4,10].

The diagnosis of KD is quite difficult. The differential diagnosis includes angio lymphoid hyperplasia with eosinophilia (ALHE), tuberculosis, Hodgkin lymphoma, Unicentric Castleman disease, eosinophilic granulomatosis with polyangiitis, Toxoplasma lymphadenitis, and angioimmunoblastic T cell lymphoma. KD and ALHE were often considered identical in the past literature. However, ALHE is a neoplasm of blood vessels, whereas KD is a chronic inflammatory disorder[8–10]. The definitive diagnosis of KD is made from a histological examination of the excised lesion. The typical histologic findings of KD include preserved nodal architecture; follicular hyperplasia with reactive germinal centers; eosinophilic infiltrate involving the interfollicular areas; vascularization of germinal centers; necrosis of germinal centers; and polymorphonuclear leukocytes, eosinophils in the germinal centers[2,3,11]. Imaging studies can provide details regarding the size and depth of nodules.

The primary treatment for KD includes surgical resection. In addition to surgical therapy, medical therapy with regional or systemic steroid therapy, cytotoxic therapy, and radiation have also been utilized[1,5]. KD has an excellent prognosis, although it may recur locally and wax and wane over time. It has a high local recurrence rate, with a 25% reoccurrence following surgery[9].

Conclusion

KD is a rare inflammatory disease with a very low prevalence worldwide. It is a rare disorder most seen in males in their 20s or 30s. It is often challenging to diagnose KD and involves some investigations that can be avoided due to its similarity in presentation with other rare diseases. Developing countries where tuberculosis is prevalent at a time can produce great confusion in diagnosing and managing KD. Thus, the treating physician should have a differential of KD in a patient presenting with lymphadenopathy and in whom antitubercular medications do not show improvement.

Ethical approval

None.

Consent

Written informed consent was obtained from the patient. A copy of written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

S.B., N.R.S. were involved in writing original draft. R.P., N.R.S., S.L., S.B., and S.P. were involved in conceptualization, design, and preparation of manuscript. S.L., N.R.S., and M.P. were involved in finalization of manuscript.

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