Case Report

Kimura disease of the supraauricular region: a rare presentation

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Received: 16 April 2019  
Revised: 06 July 2019  
Accepted: 11 July 2019

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ABSTRACT

Kimura disease is an idiopathic chronic inflammatory disorder involving the subcutaneous tissue. It favors the head and neck region, although presentation over atypical sites has been reported. This disease occurs spontaneously, has a characteristic indolent growth pattern but has a high tendency to recur locally after treatment. Due to its benign nature, there is still no clear consensus on the optimal management of this rare disorder. Various treatment options are available which include systemic steroids, antihistamines, immunosuppressants, chemotherapy, radiotherapy, and surgical excision. Presently, the disease remains a therapeutic enigma as the pathogenesis is still a mystery and its tendency to recur following treatment proves to be discouraging. Herein we report a rare case of Kimura disease involving the supra-auricular region who underwent surgical excision.

Keywords: Kimura’s disease, Head and neck, Supra-auricular

INTRODUCTION

Kimura disease (KD) was first discovered and was known as eosinophilic hyperplastic lymphogranuloma in 1937.1 It attained its current name and become widely known when Kimura et al described in more detailed of the similar pathology in 1948.2 Various conflicting hypotheses have been established regarding its etiology however there is almost complete unanimity that KD is a form of a chronic inflammatory disorder of the subcutaneous tissue with a predilection towards the head and neck region. It mainly manifested as a painless subcutaneous swelling and/or lymphadenopathy. However, there are rare reports of systemic manifestation particularly nephrotic syndrome in association with KD.3 This disease is diagnosed in average in the second and third decade of life but in actuality can become apparent at any age. It has a prolonged and indolent clinical course some lasts even decades. Yet it is noteworthy that it has a good prognosis without risks of malignant transformation. Due to its rarity, it is usually not included in the differentials of the head and neck pathology. A working diagnosis of KD is usually made when there is subcutaneous swelling and unusual presentation of hypereosinophilia. A confirmatory histopathological result (HPE) is essential as findings are pathognomonic and consistent with various literature reviews.4 Takeshi et al suggest a combination of magnetic resonance imaging (MRI) and ultrasonography is some of the superior diagnostic tools to diagnosed KD. They also favor surgical excision of the subcutaneous mass as not only tissue sample can be obtained to aid diagnosis, it provides prompt treatment of the disorder.5 In spite of numerous modality of treatments for this disease, recurrence is the most bothersome obstacle in its management. However, due to the benign nature of KD recurrence disease is often treated conservatively with regular monitoring.

CASE REPORT

In 2018, a 24-year-old Bisaya gentleman attended the Ear, Nose and Throat Clinic in Queen Elizabeth Hospital in Kota Kinabalu, Sabah for a swelling near his left ear. The swelling developed spontaneously 10 years ago,
gradually progressive in size but become more apparent recently. Hence, the reason for him to seek medical advice. He denies any pain over the swelling other significant associated symptoms. He is an occasional cigarette smoker and is allergic to seashells. Besides the noticeable swelling, the patient is fit without any remarkable medical history. On physical examination, the patient appeared healthy with normal vital signs. The neck examination revealed a diffused swelling involving the periauricular region which extends anteriorly to the zygoma.

**Figure 1:** Diffuse swelling over the left peri-auricular region.

**Figure 2:** Post excision wound over the supra-auricular region.

It measured 5×6 cms and is firm, non-tender with healthy overlying skin. A subcentimeter ipsilateral lymph node is palpable at the posterior triangle of the neck and the parotid region. The ear, nose, and throat examinations are unremarkable. The patient recently had a fine needle aspiration cytology (FNAC) performed which revealed normal lymphocytes with occasional macrophages. The cytological interpretation make the diagnosis of this obvious problem challenging. The patient was counseled for surgical removal of the lesion and he agreed. A computed tomography (CT) was scheduled and showed a homogenous subcutaneous lesion of the left temporal measuring 6.8×3.3×5.3 cms without bony extension. Further investigation revealed eosinophilia (41%) in the peripheral blood despite patient being well. This abnormal analysis aroused our clinical suspicion of KD. The surgical specimen obtained is mainly fibrotic tissue with a solitary lymph node which adhered firmly to its surrounding tissue. Microscopic examination of the tissue sample displayed follicular hyperplasia with lymphocytes, plasma cells, eosinophils and mast cells with fibrosis and eosinophilic micro abscesses within the germinal center. This substantiates the diagnosis of KD. Postoperatively, an active drain was inserted and removed after 48 hours. The patient is currently well and is under follow up for surveillance of disease.

**Figure 3:** Computed tomographic scan showing a subcutaneous homogenous mass in the temporal region (A) and ear lobe (B).

**Figure 4:** Excised fibrotic mass over the left supra-auricular region.

**DISCUSSION**

It is widely accepted that KD is a rare form of chronic inflammation of the subcutaneous tissue with or without regional lymph node involvement. The extraordinary findings of peripheral blood eosinophilia and an increase in serum IgE accompanying the subcutaneous swelling is almost classical of KD. To date, the pathophysiology of this remarkable disorder remains a debate among various scholars. One of the hypothesis includes an allergic
reaction due to the fact that there is evidence of an increased level of serum IgE however none can explain the cause that Figure 2: Post excision wound over the supraauricular region remains a challenge as there is no affirmation of the mechanism for apoptosis as it may be due to an eosinophil deficiency or apoptosis activated by foreign body reaction or both (Figure 2). The disease may also be attributed to genetic variations due to its predilection towards oriental patients but there are no literature reports regarding the study of genetics involving KD as of now. KD often involved the head and neck region to be more precise the periauricular region and the salivary glands. As for our case, the subcutaneous swelling involvement demonstrates an apparent deformity of the facial contour because it extends from the periauricular region encroaching the scalp. Our differentials prior to the histopathological reports include lipoma, lymphoma, tuberculosis, and malignancy. Histopathological studies are commonly diagnostic with findings of inflammatory eosinophils infiltrate and follicular hyperplasia, fibrosis and vascular proliferation such as discovered in our patient. Regional lymph node if involved will commonly demonstrate similar findings of the original tumor. Masayuki were able to demonstrate the relation of eosinophil count (EC) relative to the size of the granuloma which is directly proportional. Hence, they concluded that any treatment that was able to reduce the EC such as oral corticosteroids can be used to treat KD. A more radical approach in the treatment of KD is by utilizing either chemotherapy, immunosuppressant or radiotherapy. Chang et al evaluated 14 patients with KD who underwent radiotherapy with 64% have complete remission rate and 28% of the patients developed recurrence. None of their patients experienced any significant complications. Yuichi and friends successfully treated KD associated with lichen amyloidosis with low dose cyclosporine. We are in agreement with the majority of authors which inferred surgical excision as the first line of treatment of KD as it has a lower recurrence rate of 25%, requires less time as it allows immediate eradication of tumor with access to histopathologic confirmation for precise diagnosis and provide less systemic side effects compared to other forms of treatment. KD is a disease with a good prognostic value. Hence recurrence is better to be treated conservatively after diagnosis is confirmed.

**CONCLUSION**

Diagnosis of KD remains a challenging task due to its sporadic occurrence in the head and neck region. Clinical diagnosis with confirmatory histopathological evaluation is important in the management of KD to avoid unnecessary anxiety and misuse of investigative tools.

**Funding:** No funding sources

**Conflict of interest:** None declared

**Ethical approval:** Not required

**REFERENCES**


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