Kimura's disease: A case report of recurring multiple swellings in the periauricular region

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Abstract
Kimura's disease is a rare, benign chronic inflammatory disorder that manifests with abnormal proliferation of vascular endothelium and lymphoid tissues. Ten years back, then 15-year-old Nepali (Asian) male presented to the otorhinolaryngology department with swelling of approximately 5 cm x 6 cm on the left and 6 cm x 7 cm on the right post-auricular region. The patient consulted for cosmetic reasons. Fine-needle aspiration cytology revealed Kimura's disease. A high index of suspicion is needed to diagnose this disease. Early diagnosis can have proper treatment and treatment of co-existing conditions related to this disease.

Key words: Eosinophilic lymphofollicular granuloma; Inflammatory disorder; Kimura's disease.

INTRODUCTION
Kimura's disease is an eosinophilic hyperplastic lymphogranuloma: a distinct pathologic entity.1 It is a not so common, benign chronic inflammatory disorder, manifested by abnormal proliferation of vascular endothelium and lymphoid follicles. The basic aetiology is still unrevealed, reasons like autoimmune causes, allergic reactions, insect bites and subclinical infestation of Candida is postulated to have contributed to the condition.2 The disease is more prevalent in the Asian subcontinent and is seen more in males. The head and neck are the areas of prevalence. These are subdermal lesions and few patients may have renal involvement as its other manifestation.

CASE REPORT
Ten years back, then 15-year-old male presented to the department of Otorhinolaryngology, the Ear Nose Throat - Head and Neck Surgery (ENT-HNS) department with a history of multiple swellings in the bilateral post-auricular region for three months. It was painless and slowly increasing in size. These were approximately 6 cm x 5 cm on the left post-auricular and 6 cm x 7 cm on the right post-auricular region. The patient had sought the surgeon’s advice only for cosmesis. Fine-needle aspiration was done at two institutions and both revealed reactive lymphadenitis. Then excisional biopsy was done. This revealed Kimura’s disease with features showing plenty of eosinophilic infiltrates and angiolymphatic hyperplasia. His renal function test was within normal limits. The post-operative period was uneventful. He was treated with intralesional steroids and oral Cetirizine. The patient was closely followed for recurrence but was negative for 10 years. He developed another similar swelling in the right parotid and right post-auricular regions, a little behind the first one. They were approximately 5 cm x 4 cm and 3 cm x 2 cm...
respectively (Figure 1,2). Fine-needle aspiration showed Kimura’s disease again, signifying recurrence in another site. The swelling was again non-tender and soft in consistency like before. Chest examination and X-ray showed normal findings. Renal involvement was also ruled out by parameters like routine urine examination, blood urea, and creatinine which were normal. The intralesional and oral steroid was given for recurrence, which led to a significant decrease in the size of the swelling.

DISCUSSION

Kimura’s disease is seen more in young adults with male:female ratio is 3:1. Only 200 cases have been reported worldwide since its histopathological diagnosis. Common sites of involvement are the parotid glands, epitrochlear, axillary, and inguinal nodes. Rare sites of involvement include the kidneys, orbits, ears, spermatic cords, and nerves. Co-existing renal disease is common with incidence ranging from 10-60%, while 10-12% of patients may suffer from nephrotic syndrome.

Histopathological evaluation clinches the diagnosis, and surgical excision is the mainstay of treatment. The recurrence rate is approximately 25%. Many drug therapies like Cetirizine 10 mg orally for a few months to prevent recurrence have been used in some studies. Intralesional or oral steroids have also been used in recurrence. Cyclophosphamide is given in some cases. Radiation therapy is applied in a few but for such benign lesions, these modalities should be weighed carefully before implementation. A total dose of 20-30 Gy has proved effective and has been proven to prevent relapse.

A study done by Zhang et al. showed 11 out of 24 experienced recurrences of disease after treatment by surgical resection (46.2%), five out of seven experienced recurrence of disease after treatment by surgical resection followed by oral corticosteroids (71.4%) and zero out of four experienced recurrence of disease after treatment by surgical resection combined with radiotherapy (0%).

The limitation of current study is the case is reported after 10 years of first treatment and during recurrence of the disease. Present study aims to develop high level of suspicion for cases with lymphadenopathy.

CONCLUSION

Kimura’s disease apart from its rarity tends to have a low suspicion index. The diagnosis can be difficult and misleading and patients with this disease are often evaluated using avoidable procedures by just not being aware of the disease. This case of a Nepali male is presented to add another case to the literature and increase awareness about the disease.

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REFERENCES


