A case of Kimura's disease

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Abstract

Kimura's disease is not uncommon in Asia even though it is a rare entity in the West. It clinically manifest as solitary or multiple subcutaneous nodules in the head and neck region. It can be accompanied by regional lymphadenopathy and salivary gland hypertrophy.

Histology is characterized by vascular proliferation, hyperplastic lymphoid tissue and inflammatory infiltrate rich in *Eosinophills eosinophillia* can sometimes be seen. It can be associated with nephrotic syndrome. Prognosis is good and there is no malignant transformation.

We report a 24-year old male who presented with asymptomatic solitary subcutaneous nodule in L/S nasal bridge for 2 years duration. It has gradually enlarged in size. There were no regional adenopathy or salivary gland hypertrophy. Histology was suggestive of Kimura's disease and fungal studies were negative. There was no eosinophillia or evidence of nephrotic syndrome.

We treated him with intralesional steroid for which he responded. A man with Kimura's disease has been reported previously in Sri Lanka. To our knowledge this is the second case reported from Sri Lanka. We report this case to emphasize that Kimura's disease should be considered as a differential diagnosis for subcutaneous nodules in the head and neck region.

Introduction

Kimura's disease is a rare entity in the West. It is not uncommon in Asia. It was first described in Chinese literature in 1937. The definitive histological description was published by Kimura et al in 1948. There is male predominance. Peak age of onset is in the 3rd decade.

Usual clinical presentation consists of one or several asymptomatic subcutaneous nodules in the head and neck region which enlarge slowly. It can be accompanied by regional adenopathy and salivary gland hypertrophy. Overlying skin appears normal. Renal involvement is seen in 50% of patients.

Peripheral Eosinophillia and elevated serum IgE levels can be observed. Histology is diagnostic. The characteristic features are prominent lymphoid hyperplasia with proliferating germinal centres, inflammatory infiltrate rich in eosinophills and eosinophillic abscesses. Proliferation of post capillary venules is seen.

The main differential diagnosis is angiofollicular hyperplasia with eosinophillia. The etiology of this chronic inflammatory disease is unknown. An aberrant immune response to an unknown allergen is suspected. Prognosis is variable. Spontaneous remission can occur but recurrences are common. No malignant transformation has been reported.

Case history

We report a case of a 24-year old male who was reffered from the ENT unit with a history of progressive asymptomatic growth over L/S nasal bridge for 2 years duration. On examination there was a firm to soft subcutaneous nodule of 1 to 1.5 cm. overlying skin is normal. No regional lymphadenopathy or salivary gland hypertrophy.

Urine full report is normal. No peripheral eosinophillia. ESR is 8mm/hr. Lesional biopsy showed lymphoid hyperplasia with inflammatory infiltrate predominant of eosinophills. Post capillary venule hyperplasia was seen. Histology was negative for fungal studies. CT scan of the brain shows localized disease. It is a valuable test to determine the extent of the lesion. Kimura's disease was confirmed by histology.

Patient was observed for 6 months for spontaneous remission. Lesion increased in size. Considering the localization disease, he was given intralesional steroid. After 4 treatment sessions improvement of lesional size was noted.

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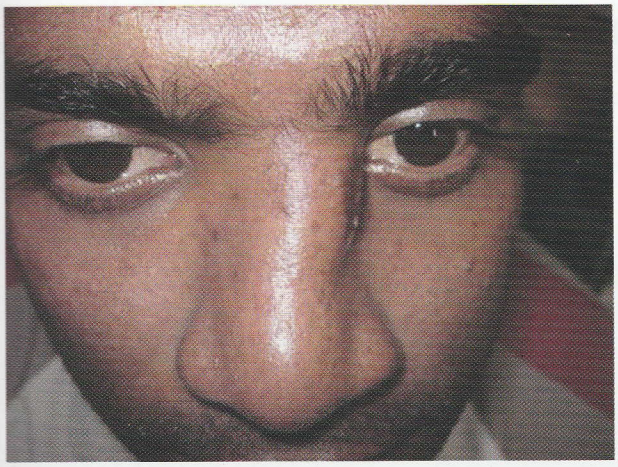
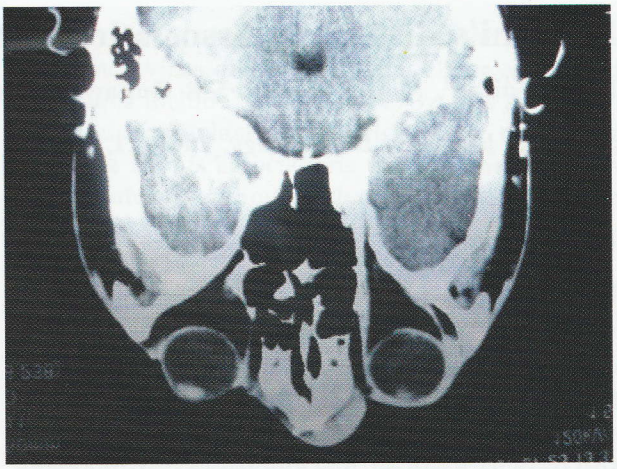
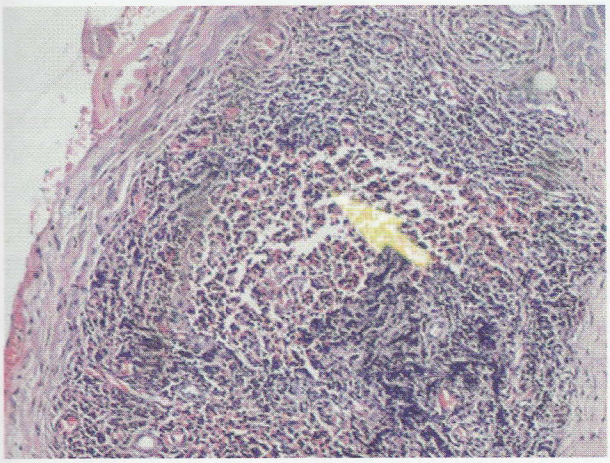


Subcutaneous swelling in the L/S nasal bridge.



CT scan showing localized disease.



Histology – hyperplasia of lymphoid tissue and cell infiltrate rich in eosinophills.

Discussion

Kimura's disease should be considered as a differential diagnosis of subcutaneous nodules in the head and neck region. Deep fungal infections and lymphoma should also be considered.

The aim of the treatment of Kimura's disease is to achieve cosmetically acceptable appearance and to prevent recurrences. The treatment of choice is surgical excision even though recurrences are common. Systemic and intralesional steroids are another option. Oral steroids are indicated if renal involvement occurs. Lesions reappear on withdrawal of steroids. Cytotoxic drugs eg. cyclosporine and pentoxipyline have been tried with variable results. Oral retinoids with or without steroids have shown success. Our patient had the localized disease, no renal involvement, and was treated with intralesional steroids. He showed improvement after 3 to 4 injections.

This case was reported to increase the awareness of Kimura's disease and to highlight its clinical features and the histology. With the predilection for head and neck region and the clinical presentation, it can be mistaken for malignancy.

References

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