A Rare Case Of Kimura’s Disease Presenting With Cough, Hemoptysis And Axillary Lymph Node Enlargement.
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Citation

Abstract
Kimura’s Disease is a benign, slow growing chronic inflammatory swelling with a predilection for the head and neck regions and has hence been restricted exclusively to literature published in those specialties. Here, we report a unique and interesting case of a 50 year old male who presented with cough, massive hemoptysis and significantly enlarged firm, non tender, mobile axillary nodes. An excision biopsy showed eosinophilic micro abscesses and associated features that gave a diagnosis of Kimura’s. The serum IgE levels and eosinophil counts were high, which corroborated the histopathology. He was treated with oral steroid therapy with the subsidence of symptoms and regression of histopathological parameters within two weeks. This case is highlighted to bring to attention an extremely uncommon presentation of a disease which normally presents in a very different manner although the complex of symptoms is very common in the context of respiratory disease.

INTRODUCTION
Kimura’s Disease is a rare benign chronic inflammatory condition of unknown etiology seen in primarily in young males of South and Far East Asia. It has been reported as an indolent benign disease which starts as a slow growing swelling involving the regional lymph nodes that mimics a number of lymphoproliferative and neoplastic conditions in its presentation. It is also known to be associated with nephrotic syndrome and connective tissue disease. Histopathological examination of the lesion or the lymph nodes are diagnostic of the condition and show eosinophilic microabscesses with eosinophilic invasion of the paracortical and sinusoidal areas of the lymph node. The blood picture shows marked peripheral eosinophilia and elevated levels of IgE. Thus far, the ases have been restricted to the head and neck region. At least one case has been reported of an inguinal swelling and there are also reports of thoracic and limb swellings. However, this patient who presented with stock respiratory symptoms of cough, massive hemoptysis and axillary lymphadenopathy is probably the first such atypical presentation.

CASE REPORT
A 50 year old male presented with complaints of cough with mucoid expectoration and hemoptysis of six days duration. He had two episodes of massive hemoptysis of around 200ml prior to hospitalisation. There was no preceding history of chronic cough, breathlessness, chest pain or fever. There was no history of subcutaneous or skin lesions. There was however, a history of a swelling in the left side of the neck two years back which regressed spontaneously with scar formation. He did not have any significant loss of weight or appetite and there were no systemic features of tuberculosis. He was not a smoker although he had occasionally smoked in the past.

A general examination revealed palpable lymph nodes in the axillary region. There was a conglomerated mass of about 5 X 4cm size, firm, non tender, mobile with matting but not adhering to the surrounding tissue. The respiratory examination revealed occasional diffuse rhonchi and crepitations bilaterally. All other systems were with normal limits. A chest X ray and HRCT were normal.

Blood investigations including a routine bleeding and clotting parameters were normal which ruled out any platelet or coagulation disorders. A peripheral smear was performed which showed 30% eosinophils with an absolute eosinophil count of 3900/mm^3, it was also negative for microfilaria which are endemic in this region. Urine did not show any proteinuria. IgE levels were much above normal at 5225 IU. Both P ANCA and C ANCA were negative which ruled out connective tissue disease.
He underwent a Fine Needle Aspiration of the axillary node. The smears showed a polymorphous population of small and large lymphocytes with immunoblasts and a few tingible body macrophages. Histiocytic aggregates and singly dispersed cells were seen along with many eosinophils among the lymphoid cells. A few plasma cells were also noted. The impression was of reactive lymph node disease with marked eosinophilia. A biopsy was requested to rule out Hodgkin’s Disease.

A bronchoscopy was performed on the patient in view of the hemoptysis to rule out endobronchial pathology. Diffuse intense mucosal inflammation with bleed was noted. The lavage fluid was taken for cytology. It showed a mixed inflammatory infiltrate with histiocytes. There was no evidence of malignant cells. BAL analysis showed 40% eosinophils, 48% alveolar macrophages and 12% neutrophils. Microbiology ruled out tuberculosis.

The histopathology of the lymph nodes reported eosinophilic infiltrates in the paracortex, germinal follicles and peripheral sinuses. Large numbers of eosinophilic microabscesses were seen. There was an increase in post capillary venules in the mantle zone and germinal follicles. Scattered areas of sclerosis were seen and adjacent adipose tissue showed eosinophilic infiltrate. A final diagnosis of Kimura’s Lymphadenopathy was made.

He was started on oral steroids and discharged. He returned two weeks later and the lymphadenopathy had completely regressed. A repeat of the blood investigations showed a decrease in the eosinophil count to 16% and IgE levels were down to 2430 IU. There have been no episodes of cough or hemoptysis since.

DISCUSSION

Kimura’s Disease has a higher predilection among the younger males and is exclusively seen in the Far East and South Asian populations. It has even been suggested that Asian ancestry could be one of the factors that plays a role in its incidence. However it is still a rarely diagnosed disease in India and only a few sporadic case reports have been published.

Thus far Kimura’s disease has been reported as an insidious slow growing swelling involving the regional lymph nodes that has even been known to temporarily subside with antibiotic therapy but with a high tendency for recurrence. It is locally proliferative and commonly arises in the region of the head and neck except very few reported exceptions involving the trunk and the inguinal region.

The symptoms of cough and hemoptysis have not been associated with this disease in the past. This patient had massive hemoptysis for which no alternative explanation or cause could be found. Bronchoscopy revealed a diffuse intense mucosal inflammation with bleed and BAL analysis showed increased eosinophilia suggestive of pulmonary eosinophilia along with tissue eosinophilia further confirming the lung tissue eosinophilic infiltration pointing to Kimura’s disease.

In the diagnosis of this disease histology plays an unparalleled role. Fine needle aspiration has been found to be ineffective as it is often confused with reactive lymphoid hyperplasia. This was true in our case. However the marked eosinophilia that is detected may be useful in diagnosing recurrences. Excision biopsy of the lesion is diagnostic of the condition and shows lymphoid follicles with prominent germinal centres and formation of eosinophilic microabscesses. There is also eosinophilic invasion of the paracortical and sinusoidal areas of the lymph node. Fibrosis and increased post-capillary venules are also associated features.

The blood investigations are most characteristic of this condition and show marked peripheral eosinophilia and elevated levels of Serum IgE. This is a uniform and diagnostic feature of this disease.

Surgical excision is the treatment of choice for both diagnosis and treatment although recurrence rates are high. Oral steroid therapy as employed in this case is an effective treatment which can also be used for recurrences. For recurrent lesions not responding to either modality, local
irradiation (25-30cGy) has been found effective.\textsuperscript{10}

**CONCLUSION**

In conclusion we may say that a suspicion of this condition may be maintained when eosinophilia or high IgE levels are seen and any lymphadenopathy or slowly progressive swellings must be looked for and investigated. It is important to note that FNAC may not be conclusive in this condition and excision biopsy is the diagnostic standard. It may not be fair to say however, that it should be a differential diagnosis for a respiratory complex of symptoms and there are still a number of conditions which are more common and more aggressive and must first be ruled out. It is hoped that this case will create an awareness of the disease and shed light on the fact that it may also arise in the axillary nodes and present in this manner.

**References**


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