

## Angio-Lymphoid Hyperplasia With Eosinophilia- Kimura's Disease: A Manifestation Of HIV Disease? A Case Presentation

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**Kimura's Disease is a rare condition, its histological features are: Lesions characterized by hyperplastic lymphoid tissue an inflammatory infiltrate rich in Eosinophils and a proliferation of post capillary venules, the patients have high levels of Immunoglobulin E and Eosinophils in their serum. A nephrotic syndrome must always be sought in these patients. The ESR is also raised in most cases. We present an 18 year old male patient who presented with features of Kimura's disease with HIV infection and look at the possibility of there being a relationship between the two.**

### Introduction

Kimura's Disease is a rare condition first described by Kim and Szeto in 1937<sup>2,3</sup>. Later a definitive histological description was done by Kimura in 1948 hence the name<sup>3</sup>. It is endemic in parts of Asia and affects young Asian men. It has a high male preponderance with a Male to Female ratio of 2:1<sup>3</sup>.

The exact cause of this disease is unknown but it is believed to result from an immunological reaction that is allergic or autoimmune in nature. The lesion shows no evidence of malignant transformation. Recurrence may occur following surgical excision<sup>5</sup>. It has a tendency to present as a discrete, enlarging mass with associated lymphadenopathy, the lymphadenopathy may be generalized but it may be regional. It has a marked predilection for the head and neck. But other sites though rare have been reported including limbs and trunk, vulva, spermatic cord, inguinal and axillary lymph nodes and peripheral nerves<sup>5</sup>. A differential Diagnosis of Angio-lymphoid hyperplasia with eosinophilia must also be considered. The two conditions although similar are different entities<sup>4</sup>.

The association of HIV with Kimura's Disease has not been widely reported<sup>7</sup>. We present the case a young man with Kimura's Disease and HIV infection.

### Case report

#### History

**KB**, an 18 years old male came into our care in November 2004 having been admitted several

times to the Government hospital with a five months history of bouts of severe abdominal pain, diarrhoea, vomiting and sometimes fever. The symptoms were of duration during which time the patient had lost weight.

There was no significant past medical history and there was no family history of similar illness. The patient was a schoolboy. He lived in one of the low cost residential areas of Chingola with his grandmother, his mother and father having died of HIV related disease three years earlier. He denied any history of travelling outside Chingola in recent past neither did he admit to playing or bathing in dams or streams.

#### Physical Findings

There was obvious weight loss .On the day of admission he weighed 52Kg. He had no pallor, no lymphadenopathy and no finger clubbing but he had a papular rash on his face and chest. He was tender in the Epigastrium. The rest of the systems were normal.

#### Investigations

Baseline tests showed the following findings:

- WBC count of  $3.9 \times 10^9/L$ , Neutropenia of 30.3% and Eosinophilia of 34%. Haemoglobin was 14.19g/dL. The BUN and Electrolytes were normal.
- **Serology for Schistosoma Haematobium** was positive in serum and urine but there were no ova in stool or urine.
- **VDRL** was negative.
- Serial blood slides for Malaria parasites were negative

- *Ultra sound scan* suggested coeliac and Para-aortic nodal enlargement.
- *Upper GI Endoscopy* revealed a normal Oesophagus. The Cardia had signs of moderately severe Reflux oesophagitis. The Fundus and Body of the stomach were normal, however in the Antrum there was a mass which was perceived to be either pressing from outside of the stomach or a leiomyoma. The Duodenum was normal.

At this stage a laparotomy was advised and done in order to obtain tissue for histopathological examination and the findings were:

- Massive lymph node enlargement in the whole abdomen.
  - The celiac nodes were particularly large.
- The stomach was normal. A biopsy of the nodes was taken.

*The histology report* showed no malignancy, but the Lymph nodes had sinuses, which were enlarged and filled with Macrophages and Eosinophils. In some places, only focal collections of Eosinophils were seen. The Conclusion was that this was an Angio-lymphoid hyperplasia with Eosinophilia also called Kimura’s Disease.

**Patient Follow Up**

**Table 1.** Weight Progress for 16 months

Date	9/11/04	16/2/05	7/3/05	23/3/05	15/6/05	30/6/05	3/8/05	14/9/05	12/10/05	11/4/06
Wt (Kg)	52	45	47	45	59	57	59	57	57	57

**Table 2.** Blood count chart

Date	%Neut	%Eosin	WBC	Hb
9/11/4	30.3	34.9	3.9	14.2
30/12/4	34.0	28.6	3.2	14.5
16/2/5	59.0	11.0	10.0	16.2
7/6/5	63.0	2.5	4.3	14.5
4/10/5	46.0	3.0	2.7	13.5
11/4/6	58.0	0.0	2.2	12.8

Our patient was considered to be an extreme case of hypereosinophilia, so steroid therapy was administered for a month from mid November to mid December 2004, with a good initial response. However the Symptoms came back after the steroid was tapered down and withdrawn. A repeat Blood count showed an Eosinophilia of 28%. At this stage, because of the continuation of his symptoms, the HIV test was done after obtaining consent from the patient and his guardian. It was positive. The first CD4 cell count was 218cells/mLitre. A diagnosis of HIV disease WHO Stage 3 was made since the patient had had diarrhoea for more than one month and he had lost more than 10% of his body weight.

In February 2005 we decided to put him on HAART with stavudine, lamivudine and nevirapine. He continued coming into hospital with diarrhoea, vomiting and fever for a period of over one month. As the treatment continued the admissions became less with the last admission being on 8<sup>th</sup> of March 2005. He has since gained some weight and has been symptom free for a long time now. At one time he developed a raised Alanine and Aspartate transaminases therefore nevirapine was substituted with efavirnez.

He is now being seen every six months and he has continued on HAART.

**Table 3.** CD4 Cell progress

Date	22/2/5	19/7/5	4/10/5	4/4/6
CD4 Count (cell/mL)	218	121	164	359

**Table 4.** ESR Progress

Date	20/3/5	16/4/5	07/6/5	11/11/6
ESR(mm/1 <sup>st</sup> Hr)	20	06	04	02

**Table 5.** BUN and Electrolytes

Date	Urea	Na <sup>+</sup>	K <sup>+</sup>	Cl <sup>-</sup>	Creatinine
16/2/5	4.5	133	1.9	99	73
18/2/5	1.9	136	1.5	119	34
21/2/5	3.0	135	3.9	103	65

The ESR was not requested for at the time of first presentation but it is presumed to have been high. The rest of the record is shown in Table 4. The BUN and electrolytes were estimated on three occasions during one admission (Table 5).

We were not able to assay the Immunoglobulin E at our institution.

### Discussion

Kimura's Disease is a rare condition, its histological features are: Lesions characterized by hyperplastic lymphoid tissue an inflammatory infiltrate rich in Eosinophils and a proliferation post capillary venules. The patients have high levels of Immunoglobulin E and Eosinophils in their serum. A nephrotic syndrome must always be sought in these patients

The ESR is also raised in most cases<sup>3</sup>. In our patient there was no apparent renal involvement because the BUN and Electrolytes were normal all the time.

Kimura's disease is usually benign and it is of uncertain aetiology. It has a tendency to present as a discrete, enlarging mass with associated lymphadenopathy in 50% of the cases<sup>5</sup>. The lymphadenopathy may be generalized but it may be regional as was the case with our patient. It has a marked predilection for the head and neck. But may present else where like limbs and trunk,

vulva, spermatic cord, inguinal and axillary lymph nodes and peripheral nerve<sup>5</sup>. In our patient it was abdominal.

To make a Diagnosis of Kimura's disease there must be a histological examination of the lesion and the presence of hypereosinophilia with raised ESR<sup>5</sup>. Our patient did have a minimally raised ESR at one occasion; it is regrettable that the ESR estimation was not done at the first presentation stage. There are reports of it apparently originating from a mosquito bite or parasite infestation several years before presentation<sup>1</sup>.

Our patient had an Eosinophilia of 34%, which was very high. Eosinophilia may be Idiopathic but it can also arise in the following conditions: Connective tissue disorders, helminthic infestation, and neoplasia particularly T-Cell Lymphomas and as an Allergy<sup>6</sup>. There are some reports of eosinophilia arising as a result of Immune deficiency syndromes<sup>7</sup>. Our patient's serology test was positive for S Haematobium both in his urine and blood although we could not find the Ova of S Haematobium in both his stool and urine. He was also HIV positive. VDRL and serial malarial tests were negative. The presence of HIV and parasitic infestation could have predisposed him to have the resultant histopathological picture.

Our patient had lost more than 10% of his body weight and had a history of diarrhoea for five

months, besides his CD4 count was 218cells/mlitre. One could define him as a case of AIDS (WHO Stage 3).

Activated Eosinophils are known to produce a large number of cytokines such as Interleukins and Tumour necrosis factor alpha<sup>6</sup>. This is the basis for the various signs and symptoms of Eosinophilia for example; Eosinophilia due to parasitic infections may cause abdominal pain, diarrhoea, weight loss, cough and rashes. Our patient did have all these symptoms except cough. Eosinophilia due to Allergy leads to asthma presenting as wheezing and breathlessness. Eosinophilia may also cause and fever and congestive cardiac failure, our patient had fever from time to time.

Various forms of treatment may be given to these patients. Where a facial lesion does occur, Plastic surgery or local excision should be done and where there is recurrence, radiotherapy is recommended<sup>3</sup>. Anti parasitic therapy should be given. Our patient did receive a course of Praziquantel. In extreme cases steroids should be given<sup>3</sup>. Hydroxyurea is said to be useful.

Our patient responded only after HAART was administered, although to start with he continued coming in with his symptoms for over month, his last admission being on 8<sup>th</sup> of March 2005. This was about the time his Eosinophil count dropped to about 11% and he began picking up his weight because the diarrhoea and vomiting had reduced. The reduction in the Symptomatology could have been due the reduction in cytokine load. It must be noted also that the steroids did not reduce the Eosinophil count significantly because by the

time we stopped the steroid the Eosinophil count was still 28%.

However, we notice a steady drop in the Eosinophil count the moment we started him of HAART. The drop in the Eosinophil count could have come about by the reduction in the viral load as the patient started on HAART. It could therefore be considered that this was another manifestation of HIV infection. However it can also be said that the symptoms exhibited by our patient could have been caused by the usual causes of these symptoms in these patients. We suspect the former.

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