East African Medical Journal Vol. 94 No. 1 January 2017
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A. O. Adisa, BDS, FMCDS, Lecturer and Consultant, Department of Oral Pathology, University of Ibadan and University College Hospital Ibadan, Nigeria, S. E. Udeabor, BDS, FWACS, Lecturer and Consultant, Department of Oral and Maxillofacial Surgery, University of Port-Harcourt and University of Port-Harcourt Teaching Hospital, Port-Harcourt, Nigeria, B. F. Adeyemi, BDS, FWACS, MSc, A. O. Akinyamoju, BDS, FWACS, Lecturer and Consultant, Department of Oral Pathology, University of Ibadan and University College Hospital Ibadan, Nigeria and G. O. Ogun, MBBS, FMCPath, Lecturer and Consultant, Department of Pathology, University of Ibadan and University College Hospital Ibadan, Nigeria

KIMURA'S DISEASE IN A NIGERIAN: CASE REPORT

A. O. ADISA, S. E. UDEABOR, B. F. ADEYEMI,
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SUMMARY

Kimura's disease (KD) is a chronic inflammatory disease presenting as multiple subcutaneous swellings within the head and neck region. Aetiology has been suggested to be due to allergy or an immune response and it's predominantly seen in young and middle-aged males. Histopathologically, hyperplasia of lymphoid tissue, with well-developed lymphoid follicles and marked infiltration of eosinophils are seen. KD is a relatively rare disease previously reported to be endemic in Asians of Chinese decent. However, they can constitute a diagnostic challenge where there is low index of suspicion. Therefore, we report a case of KD in an adult male Nigerian.

INTRODUCTION

Kimura's disease (KD) is a chronic inflammatory disease of undetermined etiology. It presents as multiple painless solitary subcutaneous nodules mostly in the head and neck region with coexisting lymphadenopathy and peripheral eosinophilia. An allergic disease or immune response has been proposed (1). Although rare in other races, a study reported the disease as endemic in Asians of Chinese decent (2). That report characterised the disease as having the following features: A young and middle-aged male predominance, predilection for the head and neck regions, a long duration, single or multiple lesions, mainly involving subcutaneous tissues, major salivary glands and lymph nodes. Histopathologically, the lesion is characterised by hyperplasia of lymphoid tissue, with well-developed lymphoid follicles, marked infiltration of eosinophils, proliferation of thin-walled capillary venules, and varying degrees of fibrosis. Because this lesion is relatively rare, they can constitute a diagnostic challenge, especially in a population where there is low index of suspicion. Hence we report a case of KD in an adult male Nigerian.

CASE REPORT

A 36-year old male Nigerian from the eastern part of the country presented to our facility. He gave a history of a painless, progressively expansive recurrent right cheek mass following a surgical excision dating two years back (Figure 1). The initial mass had been present for approximately three years before he sought medical intervention. He also described the development of a right supra-orbital mass (Figure 2) and left parotid mass (Figure 3), two months and six months respectively, following the surgery. Histopathological report from the referring facility was a granulomatous inflammatory disease.

Family and medical history obtained were non-contributory. Clinical examination revealed all three facial masses as soft and painless with slightly hyper-pigmented skin covering. A clinical assessment of neurofibromatosis type I was made with a possible differential of multiple lipomatosis syndrome. An incisional biopsy of the right cheek mass showed a section with multiple lymphoid follicles having follicular hyperplasia aggregated in moderately dense and vascularised fibrous connective tissue that was markedly infiltrated by eosinophils (Figures 4, 5, 6). Initial microscopic review hinted reactive lymphoid hyperplasia likely due to a parasitic organism. However, with further discussion around the histology description and coupled with the facial features, a final diagnosis of KD was made. Subsequent laboratory tests requested showed marginally high peripheral blood eosinophilia and very high serum IgE levels. Analysis of the laboratory test results confirmed the diagnosis of KD (Table 1).

TREATMENT

The patient was commenced on oral prednisolone tablets following the regimen: 20mg 8-hourly for 5 days, 10mg 8-hourly for 5 days and 5mg 8-hourly for 5 days. Three days after the commencement of this therapy a marked reduction of all the facial swellings was noted and the swelling progressively reduced in size till there were no obvious masses to the untrained eye. However, four days following the completion of this trial, the facial swellings reappeared after one week without steroid therapy but were not as large as the initial masses. After a clinical review and laboratory testing, the patient was put on a similar steroid therapy cycle followed by a 5mg daily maintenance dose for 2 weeks.

Table 1
Requested laboratory investigation

Test	Test Value	Reference Value	Interpretation
Eosinophils	8.9%	0.1-6.0%	Marginally high
Serum IgE	2709.6 KU/L	1-70 KU/L	Extremely high
Urea	2.8 mmol/L	2.5-6.4 mmol/L	Normal
Creatinine	62 µmol/L	57-113 µmol/L	Normal

Table 2
Comparison of Kimura's disease in individual African American and Black African cases

Cases	Duration of Swelling (Years)	Age of Patient	Gender	Site	Recurrence
I	3	40	Male	Preauricular/ Zygoma region	Present
II	4	15	Male	Zygoma	Present
III	4	23	Male	Cheek	Present
IV	3	36	Male	Preauricular/ Cheek/Supra -orbital region	Present

*Case 1- African American, cases 2, 3 & 4- Black Africans living in Senegal, Brazil and Nigeria respectively.

Figure 1
Right cheek and left parotid masses

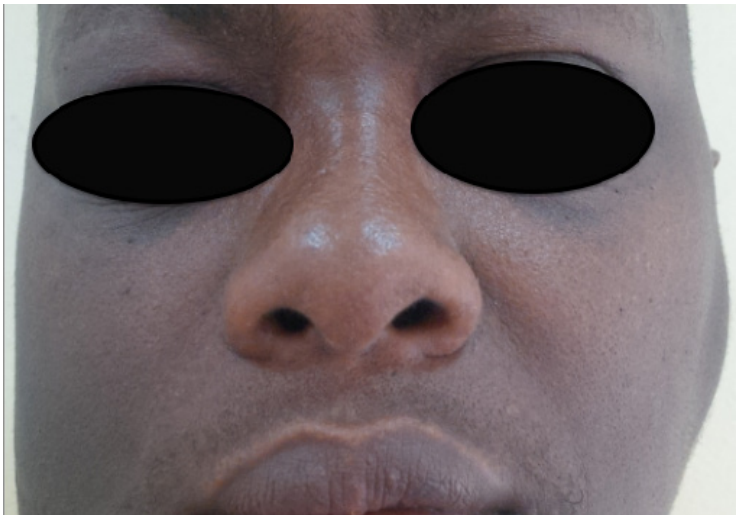


Figure 2
Right supra-orbital mass

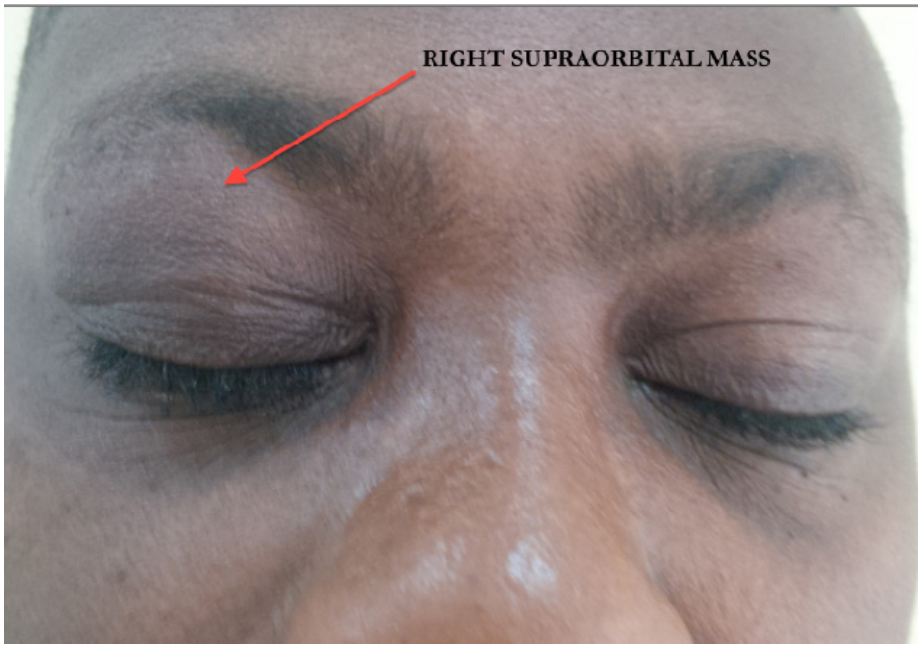


Figure 3
Left parotid mass

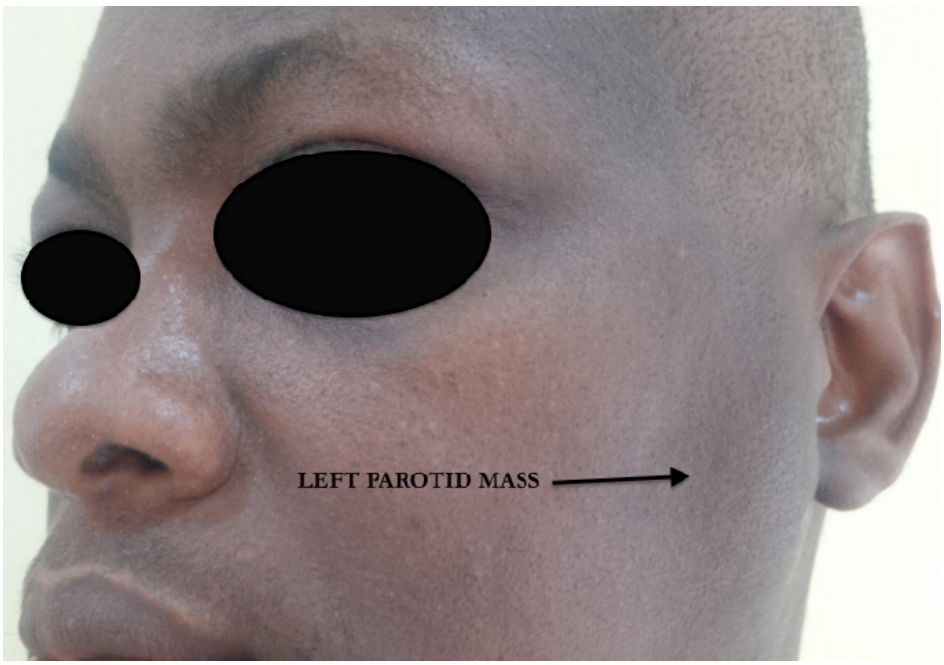


Figure 4
Multiple lymphoid follicles having follicular hyperplasia aggregated in moderately dense and vascularized fibrous connective markedly infiltrated by eosinophils (X40)

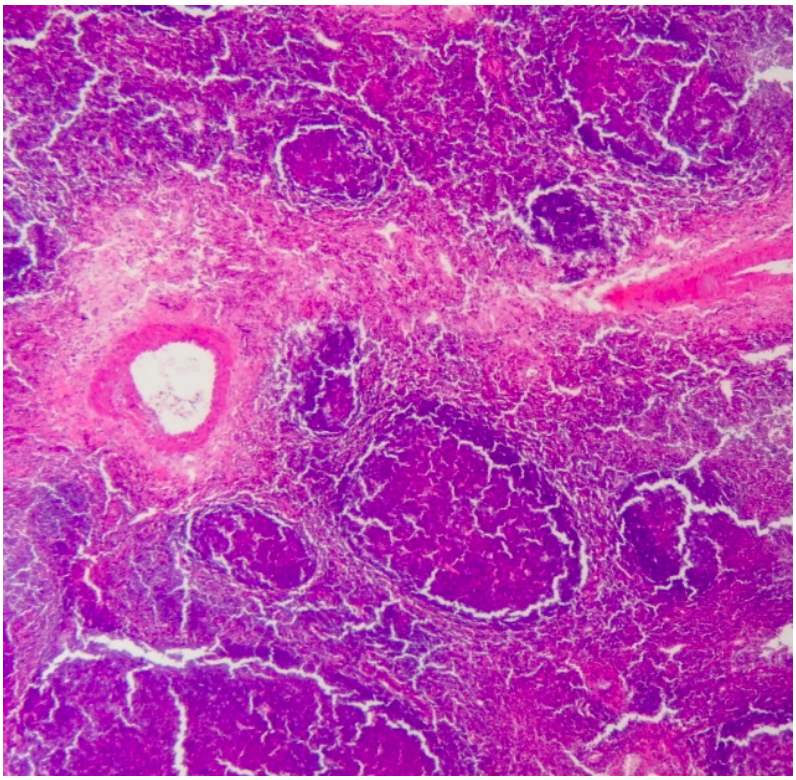


Figure 5
Multiple lymphoid follicles having follicular hyperplasia aggregated in moderately dense and vascularized fibrous connective markedly infiltrated by eosinophils (X100)

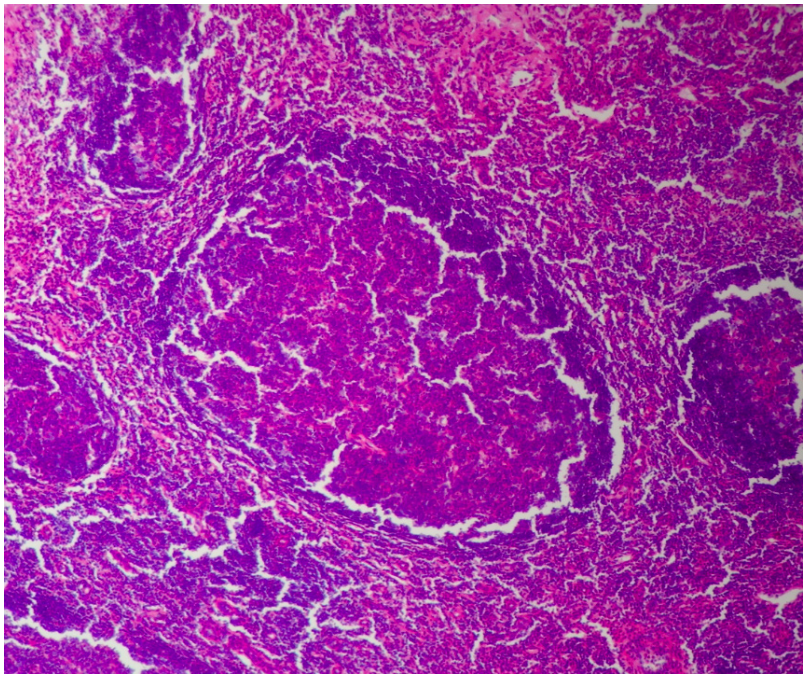
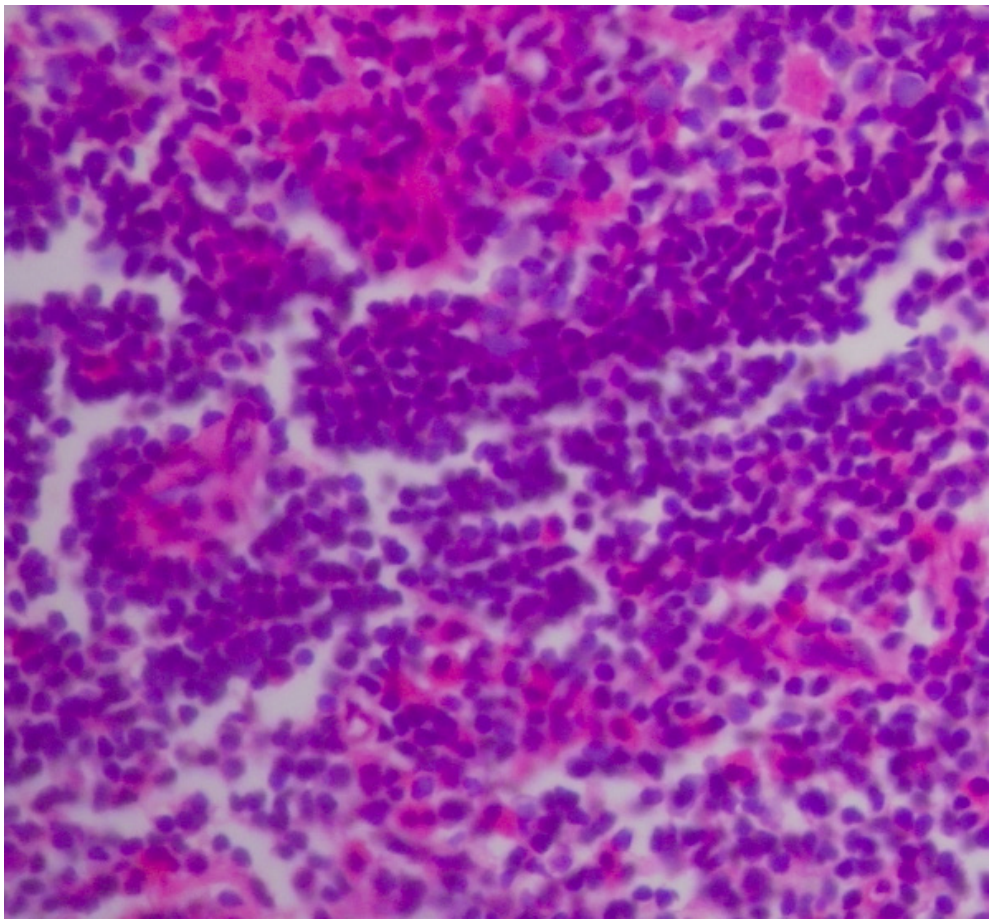


Figure 6
Lymphoid follicle having follicular hyperplasia adjacent to area markedly infiltrated by eosinophils (X400)



DISCUSSION

Most of the previous cases of KD reported have been in Asians and the documentation of this condition in black Africans is quite rare, though a few cases have been reported in patients of African descent in other climes (3). (Table 2). Thus, a low index of suspicion during clinical assessment and differential diagnosis is expected in an African population. The patients' history suggests a disease that had bewildered clinicians and pathologist for about five years prior to his visit to our facility. We also initially considered several other diseases such as neurofibromatosis type I, multiple lipomatosis syndrome clinically and a reactive lymphadenitis histologically. The only flag that raised further suspicion was the massive eosinophil population within the related connective tissue. Even then, we still considered that this might be due to a parasitic infection. Further review however led us to query an angiolymphoid hyperplasia with eosinophilia (ALHE). Differentiating between ALHE and Kimura's disease is quite challenging, however blood vessel walls containing hypertrophied and sometimes vacuolated endothelial cells with eosinophilic cytoplasm and rarely a typical nuclei never appear in KD (4).

English literature search identified KD in six blacks in the United States, a Nigerian patient living in Brazil, an African-American and a black African in Dakar (3, 4, 5, 6). The features in these cases are comparable to the case in our report.

In conclusion, the diagnosis of KD is a combination of clinical, histological and laboratory analysis and although it is rare in the African population, unexplainable subcutaneous multiple facial masses should highlight KD among other possible differential diagnosis.

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