CASE REPORT

Extranodal NK/ T-Cell Lymphoma in an African

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Abstract

Background: Extranodal Nasal NK (Natural Killer) / T-Cell lymphoma is one of the rare tumours of the head and neck region. Its prevalence is unknown among Africans. It is characterised by progressive destruction of the structures in the nasal cavity, may erode the palate and in some cases, it may metastasize. Hallmark of diagnosis is immunohistochemistry. With delayed presentation, the prognosis is very poor.
Methods: We present a 30 year old lady with complaints of two years' history of thick nasal discharge, progressive nasal blockage, hyponasal speech and pains in the nose. She was diagnosed with retroviral disease few weeks before presentation. Anterior rhinoscopy revealed a mass in the right nasal ala, destroyed nasal septum, crusts filling both nasal cavities. Patient had biopsy of the nasal mass and was screened for syphilis and Tuberculosis. She also had full blood count, serum electrolytes urea and creatinine assessment.
Results: Mantoux test and VDRL (Veneral Disease Research Laboratory) test were negative, histology result revealed malignant lymphoma and immunohistochemistry was positive for CD45, CD3, CD79a and Epstein-Barr virus-encoded RNA (EBER) but negative for CD10, CD20, EMA and Vimentin. By the time of making the diagnosis, the patient was lost to follow-up.
Conclusion: Extranodal NK/T-Cell lymphoma, being rarely seen among Africans, required high index of suspicion for diagnosis. Physicians should be on the lookout for it, multicentre reports should be collated to find its prevalence among Africans.

Key words: African, NK / T-cell lymphoma, immunohistochemistry,

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Introduction

Extranodal NK-/T-cell lymphoma is one of the EBV (Epstein-Barr virus) associated aggressive lymphomas. Midline destructive lesions involving the face were initially described in the later years of 19th century, precisely 1897[1,2]. These lesions are characterised by progressive destruction of the midline facial structures associated with unrelenting ulceration and necrosis of the midline facial structures[3,4]. A unique feature of midline destructive lesions of the face is their association with Epstein-Barr virus[5,6]. Majority of the non-Hodgkin lymphomas in HIV infected individuals are strongly associated with herpes viruses, especially EBV[7]. There is an increasing incidence of peripheral T-Cell lymphomas in HIV infected patients compared to immunocompetent patients[8] . The lesions are very rare, with sporadic cases reported in the Asian continent, North and Central American regions. However, the prevalence among black Africans is unknown[6].The lesion is seen in both sexes, but with slight male preponderance[6]. High index of suspicion is needed for diagnosis, as most patients will present with features suggestive of rhinosinusitis. Immunohistochemistry is the gold standard in diagnosis[9,10], to be supplemented with imaging. The mainstay of treatment is early diagnosis and timely commencement of chemoradiation. However, with late presentation the prognosis is very poor.

Case Report

We present a 30 year old lady, a Nurse, with complaints of two years' history of thick nasal discharge and progressive nasal blockage, hyponasal speech and pains in the nose. She developed loose upper right teeth, regurgitation of feeds in to the nose, but no epistaxis, weight loss or fever. Our patient has had several medications from peripheral hospitals since onset of the symptoms without improvement in her symptoms. She was diagnosed with retroviral disease and commenced on Highly Active Antiretroviral Therapy (HAART) one week before presenting to our facility.

She was found to be having facial asymmetry, swollen right ala, loss of the right nasolabial fold and tenderness over the right side of the nose. She had a growth on the right side of the hard palate and a fistula on the right. Anterior rhinoscopy revealed crusts filling the two nasal cavities, destroyed cartilaginous part of the nasal septum, with a remnant of the bony septum visualized. Both the inferior and middle nasal turbinates

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were destroyed and a fleshy mass was seen in the region of the right ala, which was biopsied.

Initial assessment of mitotic right maxillary tumour to rule out chronic granulomatous lesion of the right maxilla was made. Patient was placed on nasal douching, steam inhalation, oral antibiotics and an empirical antifungal agent. Computed Tomographic (CT) Scan of the paranasal sinuses Figure 1 below showed irregular soft tissue in the right nasal cavity, absent inferior turbinates, minimal mucosal thickening of the walls of the right maxillary sinus, other paranasal sinuses were free, so also the osteo-meatal complex.

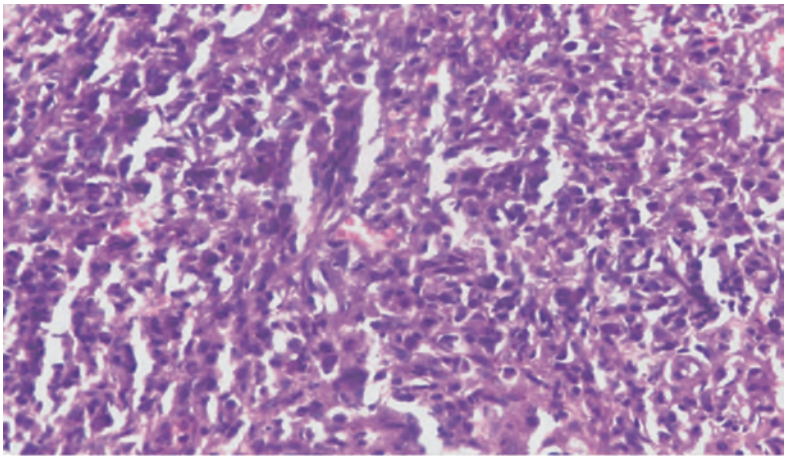

Figure 2: Photomicrograph of the initial Histology (Haematoxylin & Eosin x400)

VDRL was negative and Mantoux test was non-reactive. She had a packed cell volume (PCV) of 26%, WBC of $6.5X10^{9}$/L, Neutrophil count of 55% and Lymphocyte count of 44%. Electrolytes, Urea and Creatinine showed Sodium; 135mMol/L, Potassium; 4.2mMol/L, Chloride; 98mMol/L, Bicarbonate; 27mMol/L, Urea; 6mMol/L (slightly elevated) and Creatinine; 68μMol/L.

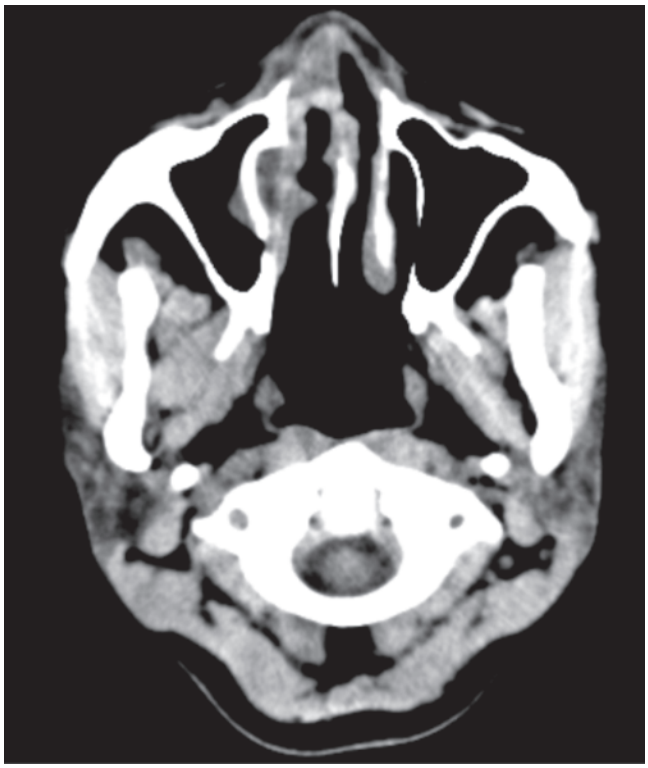

Figure 1: CT Scan of the Paranasal Sinuses showing destroyed nasal turbinates and septum with clear maxillary sinuses

Histopathological examination of the biopsy showed, polypoidal fragments of tissue containing a malignant neoplasm composed of sheets of small blue cells with hyperchromatic nuclei, indistinct nuclei and very scanty cytoplasm. The cells were arranged in sheets with few abnormal mitotic figures as shown in Figure 2 above.

Immunohistochemistry was positive for CD45, CD3, CD79a and Epstein-Barr virus-encoded RNA (EBER) and negative for CD10, CD20, EMA and Vimentin. Figures 3a, 3b and 3c below showed the photomicrographs of the patient's immunohisto-chemistry

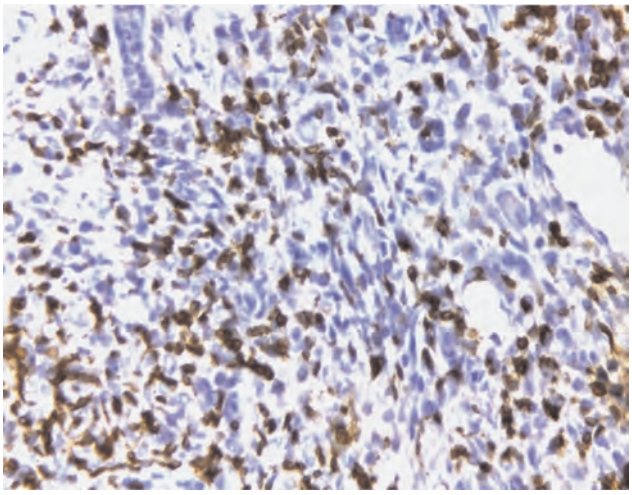

Figure 3a: Immunohistochemistry for CD3 (x400)

A final diagnosis of Extranodal NK-/T cell lymphoma was made. As at the time of writing this report, the patient has defaulted from follow-up.

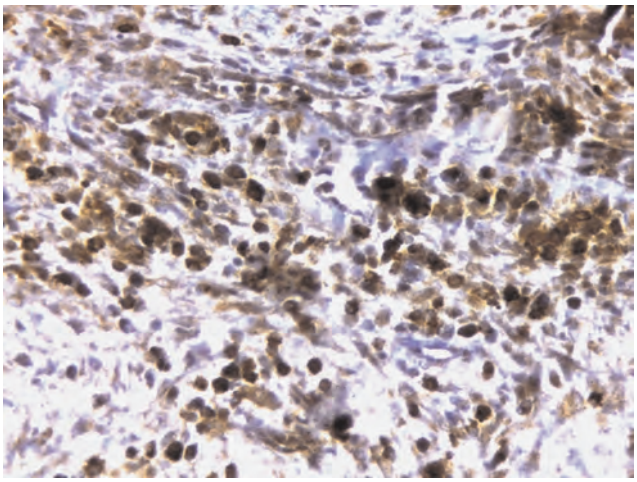

Figure 3b: Immunohistochemistry for CD79 (x400)

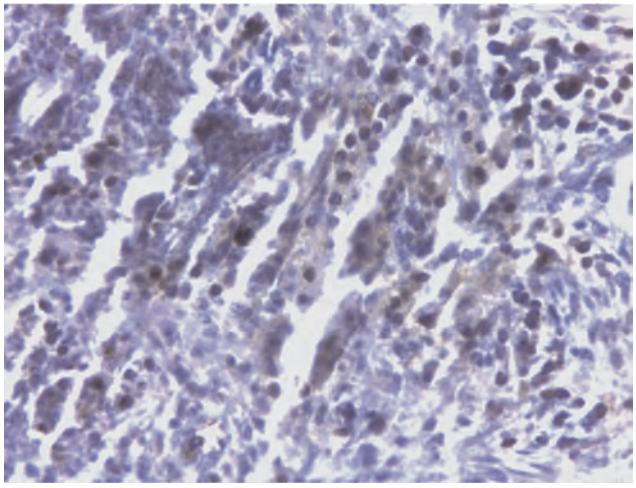

Figure 3c: Immunohistochemistry for EBER (x400)

Discussion

Extranodal NK / T-cell lymphoma being one of the aggressive tumours of the head and neck region, is characterised by progressive destruction of the structures in the nasal cavities, and occasionally the paranasal sinuses. Non specificity of symptoms to make a diagnosis of these lesions contributes significantly in delayed diagnosis and hence late commencement of treatment of such lesions[11]. Gregory et al[5] reported the mean age of presentation as 45, with a range of 19 to 80 years. Our patient was 30 years of age at the time of presentation. Our patient has had two years history of progressive nasal blockage and nasal discharge, mimicking rhinosinusitis. Acute invasive fungal rhinosinusitis, commonly seen in immuno-compromised patients[9] just like our patient could have been a possibility.

Our patient developed progressive destruction of the nasal cavity over the two-year period and had various medications from peripheral hospitals without improvement. Despite the duration of the illness, systemic features like weight loss and fever[11] were not present.

Diagnostic nasal endoscopy carried out on our patient revealed complete destruction of the cartilaginous and most of the bony nasal septum, destroyed inferior and middle turbinates, mostly of the right nasal cavity. This finding corroborated the reports of Bruno[1], Ashraf[2] and Martha[6]. Computed Tomographic (CT) Scan of the paranasal sinuses of our patient showed destroyed mid-nasal structures and the nasal turbinates. However, both the maxillary sinuses were not involved. There was a slight thickening of the mucosa of the right maxillary sinus. The hard palate on the right side was breached. Other paranasal sinuses were free of the disease. This finding is similar to the report of Meha[9] but contrasts the report of Martha [6] where the CT scan of their patient showed increased density in all the sinuses and also the nasopharynx. Generally, there has not been agreement on specific radiologic features of extranodal nasal NK/T-cell lymphomas. Imaging modalities help to delineate the lesion, assess the level of progression over a given period of time and most importantly, assess the effectiveness or otherwise of treatment.

The hallmark of diagnosis of extranodal T-cell lymphomas is the demonstration of T-cell markers such as CD2, CD3, CD4, CD5, CD7,CD45RO and other cell markers such as CD45[5]. The absence of B-cell markers such as CD20 and CD10, and Vimentin strengthens the diagnosis in favour of a T-cell lymphoma. The NK associated marker, CD56 is usually present, but other NK cell markers such as CD16 and CD57 might not be usually seen[5]. Our patient had immunohistochemistry with positivity of CD3, CD45 and CD79a. B-cell marker CD20 and Vimentin were negative. Epstein-Barr virus is strongly associated with the development of Non Hodgkin Lymphomas like extranodal NK/T-cell lymphoma. Our patient had EBER positivity, further strengthening our diagnosis of extranodal NK/T-cell lymphoma. Castillo et al[8] found out that in their series, 50% of the cases of NK/T-cell lymphomas in HIV-infected patients was associated with EBV infection. Upon establishing diagnosis, all efforts to trace the patient for commencement of chemoradiation failed. Without treatment, the condition is mostly fatal with about 50% of the deaths occurring due to systemic spread[11]. Arthur[9] documented the median survival time of 9 months in patients with disseminated disease. Castillo and Pantanowitz[12] found the median survival among treated patients to be 1.1years, in another study [8], the median survival in untreated cases was found to be one month after establishing diagnosis. However, with the introduction of chemoradiation in the treatment of

the disease, there seem to be improvement in survival rate[3].

Conclusion

We presented a 30 year old African lady with clinical and pathologic features of extranodal NK/T-cell lymphoma who defaulted from follow up after being diagnosed. High index of suspicion in the sub-saharan Africa should be the watchword for early diagnosis. Multi centre collaboration should be encouraged to determine the prevalence of the disease among Africans.

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