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Published Case Study– A report of three cases of NK/T-Cell Lymphoma enrolled in the prospective Kamuzu Central Hospital Lymphoma study in Malawi

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Tamiwe Tomoka from the University of Malawi College of Medicine, Blantyre, Malawi, and colleagues published a case report of three patients of African descent with Extranodal NK/T-Cell Lymphoma (ENKTCL) in the journal BMC Cancer on 6th September 2017.

The three patients were seen between September 2013 and March 2014 and enrolled in the prospective Kamuzu Central Hospital Lymphoma Study cohort.

Key Highlights:

- Case 1: Male, 29 years of age; >6-month history of abdominal mass
- Case 2: Male, 51 years of age; 4-month history of a sinonasal tumor eroding the medial wall of the maxilla associated with right eye proptosis
- Case 3: Female, 60 years of age; 3-month history of progressive left maxillary sinus mass involving the medial left and right orbits
- All three patients were HIV-negative and had systemic B symptoms
- Two patients had ECOG PS of ≥2
- All three cases showed diffuse proliferations of intermediate to large atypical lymphocytes with high mitotic activity and areas of extensive background necrosis
- All were positive for CD3 and CD56, as well as negative to CD20
- *In situ* hybridization found all three were positive for EBV-encoded RNA; baseline plasma EBV DNA was markedly elevated in all three patients ranging from 3,412 to 2,098,940 copies/mL
- All three did not have bone marrow infiltration
- Other laboratory investigations were normal, except for mild anemia in one patient and elevated LDH in two patients
- Local standard of care for the majority of Non-Hodgkin Lymphoma subtypes in Malawi is CHOP
    - One patient was too ill to initiate cytotoxic treatment
    - Two received a single dose of CHOP but experienced rapid disease progression
- All three patients passed away due to progressive Lymphoma within 3 months of diagnosis

The authors highlight that reports of ENKTCL from Sub-Saharan Africa are rare despite EBV being highly prevalent, possibly indicating how under-diagnosed this malignancy is. The group hope that their report will increase familiarity of ENKTCL among healthcare professionals in the region and emphasizes the requirement for earlier and more accurate diagnosis. Lastly, radiotherapy is not available in Malawi, and intensive chemotherapy regimens with asparaginase or high-dose methotrexate are inhibited by limited supportive care infrastructure and drug unavailability, and so other therapeutic options are desperately required for these patients.

Abstract:

BACKGROUND: Extranodal NK/T-cell lymphoma (ENKTCL) reports from sub-Saharan Africa (SSA) are remarkably rare, despite early childhood acquisition and high prevalence of the causative infectious agent, Epstein-Barr virus (EBV), and frequent occurrence of other lymphoproliferative disorders causally associated with EBV.

CASE PRESENTATIONS: At a national teaching hospital in Malawi, three patients of African descent were seen with ENKTCL between 2013 and 2014. Patients were aged between 29 and 60 years, two with craniofacial involvement and one with a primary abdominal tumor, and all were HIV-negative. All had systemic B symptoms, and two severely impaired performance status. On histologic review, morphology and immunophenotyping demonstrated classical ENKTCL features in all cases, including diffuse proliferations of intermediate-to-large atypical lymphocytes with high mitotic activity and extensive background necrosis, positivity for CD3 and CD56, and negativity for CD20. By in situ hybridization, all three tumors were positive for EBV-encoded RNA (EBER). Baseline plasma EBV DNA was also markedly elevated for all three patients. Due to radiotherapy and chemotherapy limitations, patients were treated with CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) with rapid disease progression. All three patients died from progressive lymphoma within 3 months of initial diagnosis.

CONCLUSIONS: Our experience with these three patients in Malawi can highlight that ENKTCL does indeed occur in SSA, increase familiarity with ENKTCL among clinicians and pathologists throughout the region, and emphasize the need for better diagnosis and treatment for this neglected population.

References

1. Tomoka T. et al. Extranodal natural killer/T-cell lymphoma in Malawi: a report of three cases. BMC Cancer. 2017 Sep 6;17(1):633. DOI: 10.1186/s12885-017-3612-y.