Sinonasal Plasmablastic Lymphoma: Case Report and Literature Review

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Abstract

Introduction
Plasmablastic lymphoma (PBL) was first described in 1997 as a rare aggressive form of non-Hodgkin's B-cell lymphoma associated with human immunodeficiency virus (HIV) & Epstein-Barr virus, and most commonly located in the oral cavity. We report a rare case of sinonasal PBL from our institution.

Methods
A case report of sinonasal PBL in an HIV+ patient at a tertiary medical center is presented, including clinical data, imaging, histopathology, and treatment. In addition, a review of the literature was performed. The PUBMED database was searched for all cases of sinonasal PBL between 1978 and 2023. Age, sex, immunocompromise status, treatment, and outcomes were collected. Studies that did not separate individual cases of sinonasal PBL from aggregated data, and studies not written in English were excluded.

Results
A PUBMED search of "plasmablastic lymphoma" returned 928 articles, of which 55 studies included sinonasal PBL. Two studies could not be accessed beyond the abstract. Three studies with 29 cases were excluded because individual data was not separated from aggregate data. The final analysis included 74 cases of sinonasal PBL from 49 studies. Age ranged from 13 to 91 years. Five patients were missing sex information, two missing HIV status, and nine missing treatment status. Females comprised 20.3% (14/69). 30.6% (22/72) were HIV positive. The most common treatment regimen was chemotherapy alone at 41.5% (27/65), followed by chemoradiation at 33.8% (22/65). Three patients (4.6%) received surgery with chemotherapy, and another three received surgery with chemoradiation. Outcomes varied from death within 12 days (2 patients) to alive at 15 years. Mortality rate was 25.5% (12/47).

Conclusion
PBL is a rare malignancy in the sinonasal region. Most cases are treated with chemotherapy. Overall, it has an aggressive course with a poor prognosis.

Introduction

Sinonasal plasmablastic lymphoma (PBL) is a rare form of non-Hodgkin's B-cell lymphoma commonly seen with human immunodeficiency virus & Epstein-Barr virus that was first described by Delecluse in 1997.[1] It carries an aggressive clinical course, frequent relapse, and poor prognosis. PBL is characterized by plasmablasts or immunoblasts with expression of plasma cell markers and a dearth of B cell markers.[2] The most common head and neck location is the oral cavity.[3] Here we report a case report of sinonasal plasmablastic lymphoma and review the literature on this rare location of PBL.

Methods

We describe a case report of sinonasal PBL in an HIV+ patient at a tertiary medical center, including clinical data, imaging, histopathology, and treatment. We also perform a literature review of sinonasal PBL. The PUBMED database was searched for all cases of sinonasal PBL between 1978 and 2023. Age, sex, immune status, treatment, and outcomes were collected. Studies that did not separate individual cases of sinonasal PBL from aggregated data, and studies not conducted in English were excluded.

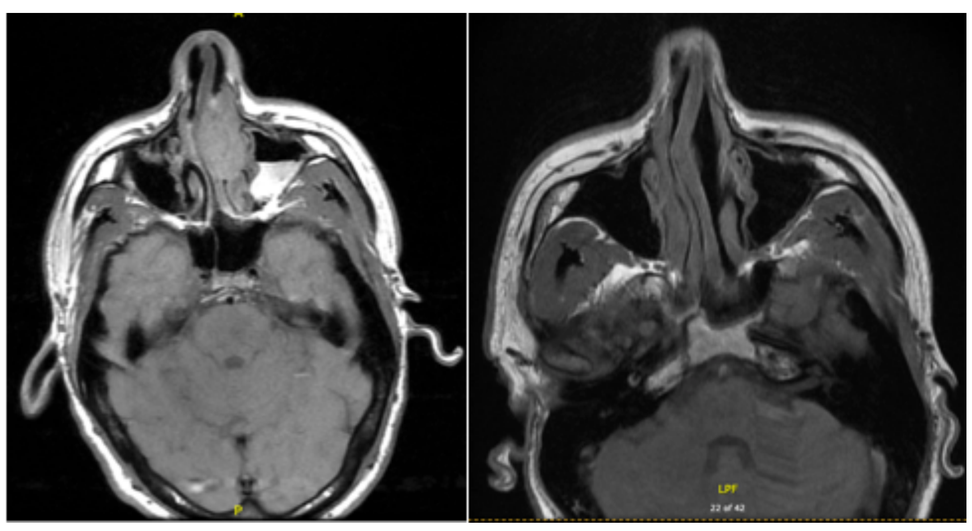

Figure 1. (left) MRI neck T1 pre-contrast with left nasal cavity mass and air-fluid level of left maxillary sinus. (right) T1 pre-contrast MRI face at 5 months' post-treatment with no evidence of residual or recurrent disease.

Results

Case Report
A 46-year-old male with HIV on highly active antiretroviral therapy with several months of bilateral epistaxis, anosmia, and nasal obstruction was found to have a large mass in the left nasal cavity. Imaging showed a 4.5cm mass of the left nasal cavity extending into the right side, without bony erosion or intracranial invasion. Biopsy resulted as plasmablastic lymphoma. Immunohistochemical studies showed plasmablasts in sheets with scant admixed plasma cells, positive for CD38, CD79a, CD138, MUM1, BCL2 (weak), and Ki67 proliferation index of 75%, and negative for CD19, CD20, PAX5, CD30, ALK, BCL1, BCL6, and MYC. He received chemotherapy with 6 cycles of etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin (EPOCH) and intrathecal methotrexate plus daratumumab-hyaluronidase-fihj, completed on March 11, 2023. Two months' post-treatment PET/CT had no abnormal enhancement. Five months' post-treatment MRI showed no residual or recurrent disease. He is currently pending evaluation with radiation oncology to evaluate whether adjuvant radiation would help prevent the risk of relapse.

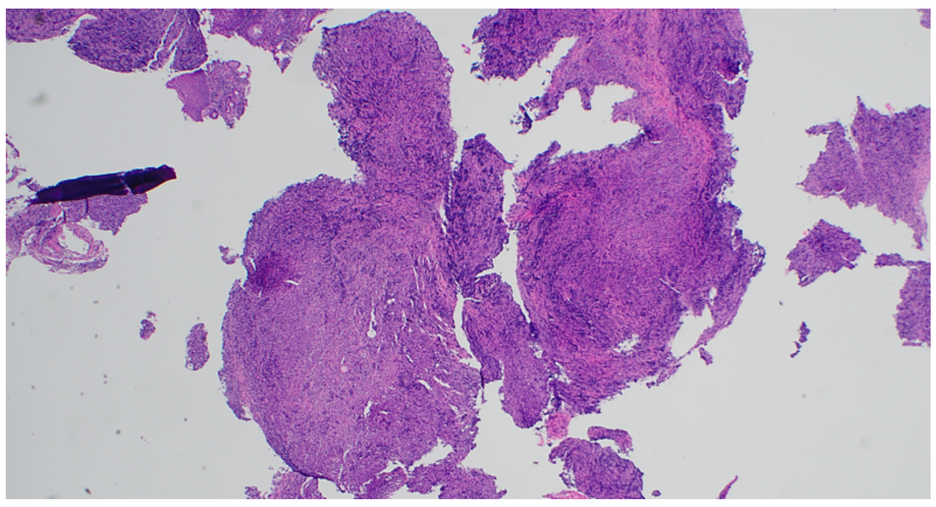

Figure 2. The patient's tissue pathology shows dense lymphoid infiltrate in diffuse sheets

Literature Review
Seventy-four cases of sinonasal PBL were included, and age ranged from 13 years to 91 years. Most were male (79.7%, 55/69), and 30.6% of all cases (22/72) had HIV. No treatment data was available for 9 patients. The majority (84.6% or 55/65) were treated with chemotherapy. The most common regimen was chemotherapy alone at 42.5% (27/65), with another 22 patients (33.8%) also receiving radiation and 3 (4.6%) receiving surgery and chemoradiation. Within chemotherapy, the most common regimen was cyclophosphamide, doxorubicin, oncovin/vincristine, and prednisone (CHOP) which 41.8% patients received (23/55). The next most common regimens were etoposide, prednisone, oncovin/vincristine, cyclophosphamide, and doxorubicin (EPOCH) and cyclophosphamide, vincristine/oncovin, doxorubicin/Adriamycin, and dexamethasone (CVAD), with 9 (16.4%) patients undergoing each.

Treatment Regimen	Number of patients (%)
Surgical resection alone	1 (1.5%)
Surgery + chemotherapy	3 (4.6%)
Surgery + radiation	2 (3.1%)
Surgery + chemoradiation	3 (4.6%)
Chemotherapy alone	27 (41.5%)
Chemoradiation	22 (33.8%)
Radiation alone	5 (7.7%)
Supportive care	2 (3.1%)
Total	65 (100%)

Prognosis and follow-up times varied but was overall poor, with mortality rate at 25.5% (12/47).

Conclusions

Sinonasal PBL is a rare and aggressive malignancy that is associated with HIV. Most patients are treated with chemotherapy, with an additional subset receiving chemoradiation, and a smaller subset receiving surgery. Length of survival varies, but sinonasal PBL is associated with a poor prognosis and high mortality rate.

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