**Scholars Journal of Medical Case Reports**
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

**Otorhinolaryngology**

# Diffuse Large B-Cell Lymphoma with Plasmablastic Differentiation: Case Report

Yessenia M. Aguilar, MD[1*], Katherine E. Cordova, MD[2], Carla M. Dominguez, MD[3], Maria A. Montesdeoca, MD[4]

[1]Resident Otorhinolaryngology, University of Buenos Aires, Viamonte 430, C1053 Cdad. Autónoma de Buenos Aires, Argentina
[2]Resident San Jose Hospital Humanitarian
[3]General Medicine
[4]Occupational Medicine

**DOI:** https://doi.org/10.36347/sjmcr.2025.v13i01.040 | **Received:** 16.12.2024 | **Accepted:** 22.01.2025 | **Published:** 25.01.2025

***Corresponding author:** Yessenia M. Aguilar, MD
Resident Otorhinolaryngology, University of Buenos Aires, Viamonte 430, C1053 Cdad. Autónoma de Buenos Aires, Argentina

**Abstract** **Case Report**

***Introduction*:** Plasmablastic lymphoma is a subtype of diffuse large B-cell lymphoma. It is more frequent in immunosuppressed patients, with the oral cavity being the most common site of involvement, with less frequency in other extranodal regions such as paranasal sinuses, orbit, skin, bone and soft tissues. It is aggressive and has a poor prognosis, with an average survival of 1 year [1]. ***Clinical Case*:** We present he clinical case of a 69-year-old male patient, derived from neurosurgery due to a tumor in the frontal region of two weeks' evolution, nasal ventilatory insufficiency of the left nostril of 2 years' evolution, for 2 months he presented recurrent epistaxis. On physical examination a tumor in the frontal region was evident. CERS plus bicoronal craniectomy, resection of the rhinosinusal tumor was performed. ***Discussion*:** Plasmablastic lymphoma is a subtype of diffuse large B-cell lymphoma. It is more frequent in immunosuppressed patients, the oral cavity being the most common site of involvement, less frequently in other extranodal regions as in the case of the patient of rhinosinusal origin [5]. ***Conclusion*:** It is a diagnostic challenge, both its clinical and histological characteristics are ambiguous, which reduces the capacity for an accurate and rapid diagnosis, it requires intensive treatment with chemotherapy, however, it has a poor prognosis [7].

**Keywords:** Plasmablastic lymphoma, large B cells, R-CHOP.



## INTRODUCTION

Plasmablastic lymphoma (*plasmablastic lymphoma*; PBL) is a malignant clonal proliferation classified by the World Health Organization as a subtype of non-Hodgkin lymphoma big B [1].

Since its initial description in patients adults with virus of immunodeficiency human immunodeficiency syndrome (HIV)/immunodeficiency syndrome acquired (AIDS) as a tumor that affected preferably the cavity oral, he they have reported several cases in patients immunocompromised by other causes with location outside of the oral cavity both lymph node and extra lymph node [2].

Due to their characteristics morphological, immunohistochemical and to his course clinical aggressive and of fast growth, represents a true diagnostic and therapeutic challenge. generally, he diagnostic in stadiums clinical advanced and, to weigh of introduce a answer initial favorable to chemotherapy, the relapse rate is high and the poor prognosis [3].

## CLINICAL CASE

We present he clinical case of a 69-year-old male patient personal medical history not reported, habits: smoker IPA: 4 pack years, referred for neurosurgery due to a tumor in the frontal region of approximately two weeks' duration with an increase in size that intensified in the last week, accompanied by right eyelid edema, nasal ventilatory insufficiency of the left nostril of 2 years' duration, for 2 months he presented recurrent epistaxis that were self-limiting, for 1 month.

Physical examination revealed a tumor in the frontal region with a soft, fluctuating consistency, slightly painful on deep palpation, with undefined edges, edema of the upper right eyelid, without affecting eye movements, and preserved visual acuity.

**Figure 1: Tumor in the frontal region**

Rhinoendoscopy reveals a yellowish pink tumor in the left nasal cavity dependent on the medial wall of the maxilla and middle meatus, and purulent discharge is observed posteriorly in the throat.

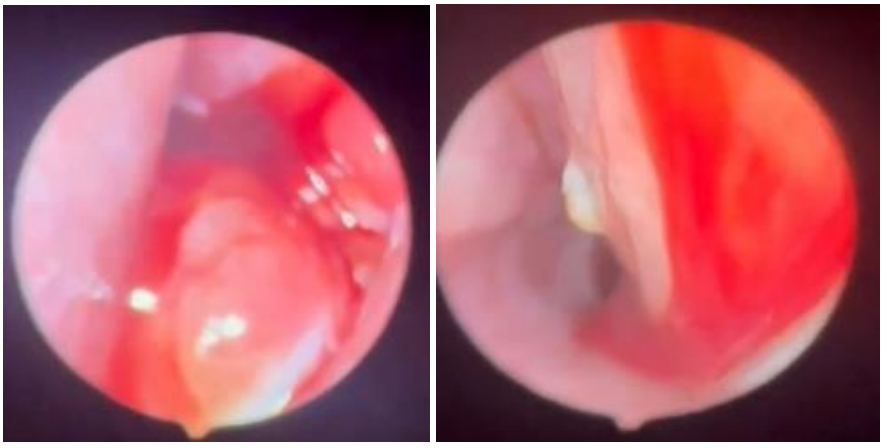

**Figure 2: Rhinoendoscopy reveals a yellowish pink tumor in the left nasal cavity dependent on the medial wall of the maxilla and meatus with slight bleeding that contacts the septum**

## RESULTS

In contrast-enhanced computed tomography of the facial craniomass, soft tissue density occupation of the frontal sinus, anterior and posterior ethmoid cells, right maxilla and sphenoid is observed. Erosion of the internal and external table of the frontal sinus, hypodense image in the frontal region with peripheral enhancement that gives the impression of a collection. Erosion of the lamina papyracea, roof of the right orbit and medial wall of the left maxillary sinus, soft tissue density occupation of the left nasal fossa with heterogeneous characteristics that gives the impression of capturing contrast.

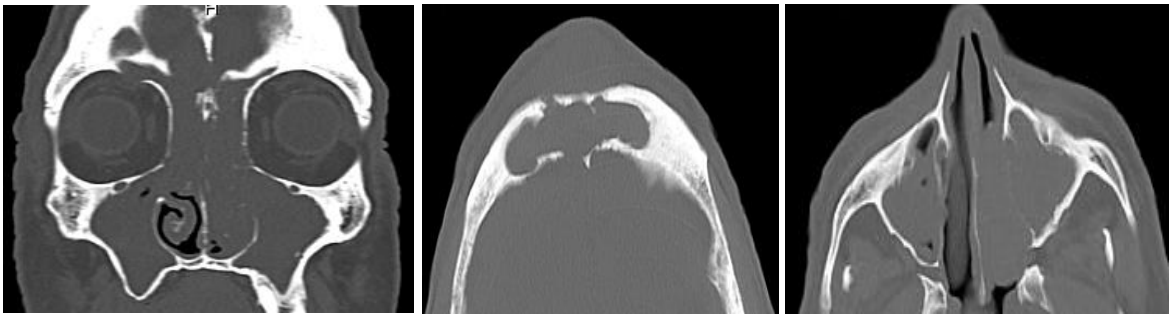

**Figure 3: Computed tomography of the facial craniomass with contrast, axial section. A). Erosion of the internal and external table of the frontal sinus is observed. B). Density of soft tissue occupation of the left nasal cavity with heterogeneous characteristics that gives the impression of capturing contrast**

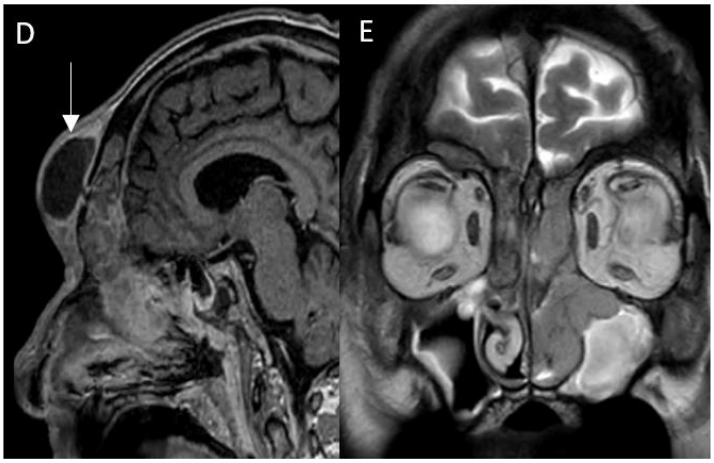


**Figure 4: D. Contrast-enhanced MRI of the brain: fluid-like collection in the frontal subcutaneous region (arrow). E: rhinosinusal formation with heterogeneous contrast enhancement**

A complicated endoscopic rhinosinusal surgery plus bicoronal craniectomy was performed resection of a rhinosinusal tumor with involvement of the external and internal table of the frontal sinus with communication to the anterior cranial fossa. During the intraoperative period, together with the neurosurgery team, a bicoronal incision was started, the flap was lifted anteriorly and the subdermal tumor was located at the frontal level, which extended by a pedicle through the external table of the frontal bone. Its excision was performed, which was infiltrative and friable, so it was removed in fragments. The external table of the frontal bone was exposed up to both orbital arcades and multiple perforations were found in it. A bone window was made by excision of the external table of the frontal sinus. An infiltrative and bleeding friable lesion is located, a sample is taken for pathological anatomy (atypical cell frozen biopsy), total excision of the intrasinus portion is completed and continuity with the cranial cavity is observed and the dura mater is detached, the mucosa of the frontal sinus is removed up to the level of the nasal bony septum, the frontal sinus is lined with temporal muscle and cranial fascia, and it is delimited with spongostan for CERS. It is performed with a 30-degree optic, maxillary antrostomy, right, posterior and left anterior ethmoidectomy are performed, the tumor is resected showing infiltration, the inferior turbinate body and tail and the left middle turbinate are resected due to infiltration, hemostasis is controlled with no evidence of tumor remains. The procedure is completed without complications.

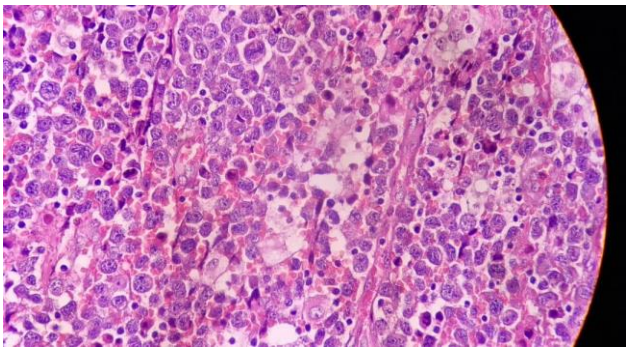

**Figure 5: Biopsy of the lateral maxillary wall; stained with hematoxylin and eosin (400x). A proliferation of large B cells with plasmacytoid features is observed**

He Follow-up was based on the biopsy results: *(diffuse large B-cell lymphoma with plasmablastic differentiation)*, the oncohematology service indicated 6 cycles of R-CHOP chemotherapy, with favorable clinical evolution.

## DISCUSSION

He lymphoma No Hodgkin represents he 65% of malignant diseases in children with HIV. PBL is a rare subtype of non-HIV lymphoma. Hodgkin, originally described as a variant of large B-cell lymphoma, with strong association to infection latent by he EBV. Yeah Although it is more common in adult males, the age range in which it occurs is wide [1].

In general, the most common sites of PBL are extranodal. In HIV-infected patients, the oral/nasal cavity is the primary site of involvement. In solid organ transplant recipients, the skin is a frequent site [2]. Other common extraoral sites of primary involvement include the gastrointestinal tract and lymph nodes. Nodal

localization is most frequently seen in HIV-negative patients [1, 2].

Although its clinical course is very aggressive and survival is generally less than one year, patients with long-term follow-up free of disease have been reported [3].

PBL represents a diagnostic challenge, as other entities may present with Histology and similar phenotype, implying the presence of large blast cells with little or absent expression of B-lymphocyte markers (CD20) and positivity for plasma cell-related antigens (CD138 and MUM1).5 Within the group of large B-cell lymphomas with plasmablastic differentiation, two main subtypes are described: PBL with plasmacytic differentiation and oral mucosa-type PBL. Both differ in clinical, epidemiological, morphological and immunophenotypic characteristics although with some overlap [4].

PBL with plasmacytic differentiation may present as nodal or extranodal disease It is seen primarily in patients who are immunosuppressed for reasons other than HIV [5].

Like oral mucosal PBL, this variant is associated with latent EBV infection and its prognosis is ominous. Histologically, it is composed of a heterogeneous population of plasmablasts and immunoblasts as well as small cells with plasmacytic differentiation with variable expression of immunoglobulin light chain restriction [6].

Oral mucosal PBL has a predilection for the oral cavity, nasal or paranasal area and is strongly associated with HIV/AIDS. It may also occur in patients with immunosuppression secondary to prolonged corticosteroid therapy or solid organ transplantation and in the elderly population. A small proportion of patients present with PBL secondary to the progression of another hematologic disease, mainly chronic lymphocytic leukemia and follicular lymphoma. EBER ISH is positive in 75% of HIV/AIDS-related PBL cases [1, 3, 4]. Histologically, a relatively homogeneous population of large lymphoid cells with immunoblastic features frequent mitotic figures and a high Ki-67 index is observed. Despite a favorable initial response, its clinical course is characterized by a high rate of relapses and death [7].

Differential diagnoses of PBL include various solid tumors (carcinoma, rhabdomyosarcoma, osteosarcoma) and lymphoproliferative processes with plasmablastic differentiation: plasmablastic myeloma, diffuse large B-cell lymphoma with plasmacytoid differentiation anaplastic kinase- positive (ALK+) lymphoma, primary cavitary lymphoma positive for human herpesvirus-8 (HHV-8), and HHV-8+ lymphoma associated with multicentric Castleman disease The presence of latent EBV infection, the absence of HHV-8 infection, association with HIV extranodal presentation the absence of monoclonal gammopathy, negativity for CD20 and other B-cell markers, as well as a characteristic plasma cell immunophenotype are the keys to differentiate PBL from other oncohematological diseases [8].

PBL is extremely rare in children. Of the patients published to date, 6 are case reports. Three case series including pediatric and adult patients and two case series of PBL in pediatrics are also described. Although the prognosis is generally poor, of the total of 25 patients published, 8 were alive at the time of the last follow-up [9, 10].

Recognition of the disease has been increasing in recent years. This is probably due to a greater suspicion of the entity and the retrospective search for patients with compatible symptoms in pathological anatomy databases [8].

In pediatrics, there is no standardized treatment due to the scarcity of data. In adults, cyclophosphamide, doxorubicin, vincristine, and prednisolone (CHOP) has been used for several years as first-line therapy. CHOP is currently considered inadequate and more aggressive therapeutic regimens, such as etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin (EPOCH), are proposed for both HIV-positive and -negative patients, although there is no evidence that they can offer better survival. In patients with HIV/AIDS, the use of intrathecal prophylaxis is advised. In the patient we present, a cytostatic regimen for high-grade non-Hodgkin lymphoma similar to CHOP was chosen with the addition of cytarabine and intrathecal treatment [9].

Antimyeloma agents such as bortezomib, as well as autologous bone marrow transplantation in first remission of patients with chemosensitive disease, has also been proposed [7].

Despite the potential for interactions with cytostatic therapy, it is suggested that HAART be started or continued. It is not known, however, to what extent antiretroviral therapy may offer a more favorable prognosis [10].

## CONCLUSION

It is a diagnostic challenge, both its clinical and histological characteristics are ambiguous, which reduces the capacity for an accurate and rapid diagnosis. It requires intensive treatment with chemotherapy however, it has a poor prognosis.

**Conflicts of Interest:** The authors declare that there is no conflict of interest regarding the publication of this paper.

**Financing:** Self-funded

## REFERENCES


1. Quintana, S., Castro, M., Hamdam, Z., Pérez, C. R., González, G., & Pallas, P. (2010). Manifestations of lymphoma in the ENT area. *SPORL J*, 48.
2. Delecluse, H., Anagnostopoulos, I., Dallenbach, F. E., Hummel, M., Marafioti, T., Schneider, U., ... & Stein, H. (1997). Plasmablastic lymphomas of the oral cavity: a new entity associated with the human immunodeficiency virus infection. *Blood, The Journal of the American Society of Hematology*, *89*(4), 1413-1420.
3. Avilés-Salas, A., Chanona-Vilchis, J. G., Candelaria-Hernández, M. G., Pen˜a-Torres, M. L., & Cuéllar-Mendoza, M. E. (2012). Initial presentation of a plasmablastic lymphoma as an ovarian tumor. *Gynecol Obstet Mex*, 80, 430-434.
4. Ghaleb Elyamany, G. E., Eman Al-Mussaed, E. A. M., & Alzahrani, A. M. (2015). Plasmablastic lymphoma: a review of current knowledge and future directions. *Adv Hematol*, 11. Article ID 315289.
5. Rafaniello Raviele, P., Pruneri, G., & Maiorano, E. (2009). Plasmablastic lymphoma: a review. *Oral diseases*, *15*(1), 38-45.
6. Astolfo, M. F., D'Antonio, F., Dartiguelongue, J. B., Arabolaza, M. N., Cheistwer, A., De Matteo, E., ... & Iriart, E. M. (2016). Oral plasmablastic lymphoma in a human immunodeficiency virus positive child: A case report. *Archivos argentinos de pediatria*, *114*(2), e95-e99.
7. Stein, H., Harris, N. L., & Campo, E. (2008). Plasmablastic lymphoma. In: Swerdlow, S. H., Campo, E., Harris, N. L., Jaffe, E. S., eds. WHO classification of tumors of haematopoietic and lymphoid tissues. 4th ed. Lyon: IARC, 256-257.
8. Delecluse, H., Anagnostopoulos, I., Dallenbach, F. E., Hummel, M., Marafioti, T., Schneider, U., ... & Stein, H. (1997). Plasmablastic lymphomas of the oral cavity: a new entity associated with the human immunodeficiency virus infection. *Blood, The Journal of the American Society of Hematology*, *89*(4), 1413-1420.
9. Castillo, J. J., Bibas, M., & Miranda, R. N. (2015). The biology and treatment of plasmablastic lymphoma. *Blood, The Journal of the American Society of Hematology*, *125*(15), 2323-2330.
10. Morscio, J., Dierickx, D., Nijs, J., Verhoef, G., Bittoun, E., Vanoeteren, X., ... & Tousseyn, T. (2014). Clinicopathologic comparison of plasmablastic lymphoma in HIV-positive, immunocompetent, and posttransplant patients: single-center series of 25 cases and meta-analysis of 277 reported cases. *The American Journal of Surgical Pathology*, *38*(7), 875-886.