



## Plasmablastic Lymphoma: Case and Review of the Literature

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### Abstract

Plasmablastic Lymphoma (PBL) is an uncommon and aggressive subtype of diffuse large B-cell lymphoma, diagnosed commonly in middle-aged male patients, and historically associated with HIV-positive patients, involving the oral cavity. It has recently been described in HIV-negative patients affecting other levels, such as the pleura, pericardium, etc. This entity is characterized by having positive CD38 and CD138 receptors, characteristic of plasma cell disease, negative CD20 receptors and high proliferative index (high Ki67). An early diagnosis and initiation of treatment is important, since it significantly improves the bad prognosis. Initial treatment recommended is conventional chemotherapy (CHOP, cyclophosphamide, Adriamycin, vincristine and prednisone) or more intensive chemotherapy (EPOCH, etoposide plus CHOP). New approaches to this lymphomas includes proteasome inhibitors (bortezomib) adding to conventional chemotherapy. We describe a clinical case of Plasmablastic lymphoma and review the literature.

### Case Presentation

A 63-year-old man was admitted in the emergency room in March 2019 with an episode of rectal bleeding and subsequent anemia. He was a heavy smoker of 30 cigarettes/day and a regular alcohol drinker, The personal medical history included arterial hypertension, ischemic heart disease with severe coronary artery disease revascularized (rug-eluting stent), atrial fibrillation, severe ventricular dysfunction, peripheral artery disease, chronic kidney disease, lacunar stroke, and previous rectal cancer in complete remission for 4 years before admitting in hospital. (The treatment was the resection and a complementary chemotherapy - , capecitabine-plus radiotherapy) During admission, a pelvic Magnetic Resonance Imaging (IRM) and colonoscopy were performed. A sessile colonic polyp (tubular adenoma) was removed and the IRM revealed a right pararectal presacral cavity with rectal fistulization, inside which an irregular mass, solid appearance suggesting granulation tissue, without being able to rule out rectal tumor recurrence. The complementary PET-CT scan showed intense pelvic hypermetabolism, apparently pararectal, and again without being able to rule out tumor recurrence or added septic process. No other findings suggestive of malignancy were identified. Given the high probability of tumor recurrence, an abdomino-peritoneal amputation and a definitive end colostomy were performed.

The result of the pathological anatomy of the presacral mass shows a diffuse proliferation, in a sheet, of a lymphoid population, with extensive areas of confluent necrosis, whose cellularity is of intermediate-large size, with vesicular chromatin nuclei, irregular and with 1 to 2 labeled nucleoli, reminiscent of immunoblasts. Immunohistochemical staining showed: CD45 LCA-o + focal, CD20 -, BSAP/PAX5 -, CD3 -, CD43 -, TdT -, CD5 -, CD10 -, cyclin D1 -, very focal BCL2 +, BCL6 -, CD21 -, CD68 + in macrophages, CD30 -, CD15 -, CD45 + focal, CD117 -, CD56 -, myeloperoxidase -, ALK -, HHV8 -, CD138 + intense diffuse with proliferative index (Ki-67) >90%. It shows no restriction of Kappa/Lambda light chains. The pathological anatomy and immunohistochemistry were compatible with the diagnosis of plasmablastic lymphoma.

A bone marrow aspiration and biopsy were performed, without observing infiltration by lymphoma and another neoplasm. With a diagnosis of primary plasmablastic lymphoma stage I (intestinal involvement) and IPI 1 (low risk), R-IPI 2 (good prognosis), NCCN-IPI 3 (low intermediate), we initiated treatment with V-CHOP, adding Bortezomib to conventional CHOP (receptors CD20 -, and CD38 and CD138 + receptors), for 6 cycles.

In the control PET-CT after 3 treatment cycles, an image compatible with an infectious complication in a presacral fibrous mass was observed, without being able to rule out neoplastic

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infiltration (SUVmax 3.9). No more hypermetabolic foci or lesions that suggest the existence of active neoplastic disease in other territories were observed, so we continued with treatment.

In the PET-CT scan after 6 cycles, the presacral mass that showed a higher peripheral metabolic degree was still appreciated, probably related to superinfection (SUVmax 7.7 - previous 3.9). Diffuse increase in bone marrow metabolic activity probably secondary to chemotherapy patient presented several and severe complications, requiring admission he presented a fistula to the skin of a presacral mass, with discharge of purulent content with negative cultures of the area, closing the fistula with local cures.

He required catheterization and cysto-urography to rule out fistulization of the presacral mass to the urethra because recurrent urinary infections, causing by *E. coli*. Lastly, he required two more admissions for decompensate heart failure and atrial fibrillation with rapid ventricular response. After adjustment of medication he improved and was discharged an echocardiogram was performed, observing a severely depressed ejection fraction, probably related to risk factors and previous cardiac pathology.

Faced with the doubt of progression or superinfection of tumor remains, it has been evaluated in the Hematology Consultation after a new PET-CT scan, the patient is currently in complete remission.

## Discussion

Plasmablastic lymphoma is a rare lymphoma, although the actual incidence is unknown. It is a very aggressive entity usually observed in the context of HIV disease, although cases are being described, as in our patient, in HIV-negative patients.

These tumors are composed of late B cells that express plasma cell markers (CD138) rather than the pan-B cell markers found in diffuse large cell lymphomas (e.g., CD20 and CD79) [1]. Some tumors that belong to this group have distinctive genetic or clinicopathologic features (Epstein-Barr virus positive, ALK rearranged). Due to the plasma cell differentiation of these tumors, the differential diagnosis with Anaplastic Myeloma is mandatory in case the bone marrow is affected.

Plasmablastic lymphoma is a predominantly male malignancy with a mean age of presentation of 55 years [1]. The majority (75%) of HIV-negative plasmablastic lymphoma patients are immunocompetent, although plasmablastic lymphoma has been associated with lymphoproliferative and autoimmune disorders, immunosuppressed patients, and after solid organ transplantation. Most of the HIV negative patients had extranodal involvement (89%, more frequent in the oral cavity, 21%, the gastrointestinal tract, 20%, the soft tissues, 17%, and bone marrow, 15% [2].

Overall survival is short, with medians of 11 to 19 months [3,4]. Poor prognostic factors include immunosuppression, as is our case, advanced stages, EBV negativity, treatment-resistant disease, and C-MYC mutations.

The therapeutic attitude given the low incidence of this lymphoma

is not clear. Conventional treatment with CHOP does not seem to be sufficiently effective and in general a more intensive or individualized treatment is recommended, including if possible transplantation after the first-line response (CODOX-M/IVAC; -EPOCH or V-CHOP) [5].

The use of CNS prophylaxis is not standardized. However, CNS prophylaxis or the use of regimens containing CNS penetrating agents is recommended because of the infiltrative capacity of these CNS lymphomas. Isolated radiation therapy has limited use.

In our case, and given the multiple pathologies of the patient despite age, we opted for using V-CHOP + TIT with the aforementioned results.

Immunochemotherapy is one of the novelties included in the treatment; however our patient did not have a CD30 receptor, so he was not susceptible to treatment with it.

CAR-T cells (chimeric antigen receptor T) are a form of genetically modified autologous immunotherapy that can target the precursor of B cells. This treatment with excellent results in patients with diffuse large cell lymphoma will be a future option for patients with plasmablastic lymphoma.

In conclusion, plasmablastic lymphoma is a rare type of lymphoma, in which there are not many cases referred to in the bibliography, from which it can be concluded, that it is more frequent in middle-aged male patients with HIV and EBV positive, although there are also cases described, like the one we present here, with negative serologies. They are characterized by sharing immunophenotypic characteristics with plasma cell neoplasms, such as CD138 +, instead of CD20 such as diffuse B-cell lymphomas. The cases studied tend to have a poor prognosis and treatment is performed with chemotherapies, such as EPOCH or V-CHOP, to perform a transplant of hematopoietic recipients in the second stage, and achieve a complete remission of the disease.

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