



## Plasmablastic Lymphoma: A Challenging Diagnosis

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### Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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## ABSTRACT

**Introduction:** Plasmablastic lymphoma (PBL) is an aggressive lymphoma characterized by early relapses and subsequent chemotherapy resistance representing therefore diagnostic and therapeutic challenge with a very poor outcome.

**Case Presentation:** A 53-year-old- female patient presented with dyspnea and general status alteration due to anemia; diagnosed as hemolytic anemia and treated by corticotherapy without amelioration. The most recent hospitalization revealed worsening of her symptoms with weight loss of 5 kilos in one week, anorexia and lumbar pain. Physical examination showed isolated splenomegaly with no signs of bleeding, adenopathy nor a palpable breast mass. Extended laboratory tests were normal, except normocytic anemia with hemoglobin =8.76 g/dl; hct=26.6%; MCV=89.2 fl and a low haptoglobin concentration level <0.04 g/l with a high value of LDH 1165 U/litre. Hypercalcemia of 12 mg/dl was also noted with low PTH. To complete the workup: imaging showed a total body CT scanner with absence of adenopathy, a magnetic resonance imaging (MRI) with no metastatic bone lesions confirming the diagnosis and a full body PET CT scanner revealing diffuse hyperactivity of the bone marrow with hypermetabolic splenomegaly,

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hepatomegaly and bone lytic lesion of some cervical vertebrae. Bone marrow aspirate, biopsy and flow cytometry were done and in favor of lymphoma. An immunohistochemical profile shows positivity and correlates with morphology. It was compatible with Plasmablastic lymphoma (PBL). Patient received three cycles of chemotherapy (lenalidomide and CHOP) and immunomodulatory agents. But she passed away three months later.

**Conclusion:** Despite the recent advances in therapy of aggressive lymphomas, patients with PBL have the poorest outcome. Moreover, due to its challenging diagnosis and related complications, management of PBL remains to discuss on a case by case basis.

*Keywords: Plasmablastic lymphoma; diagnosis; cytometry; corticotherapy.*

## 1. INTRODUCTION

Plasmablastic Lymphoma (PBL) is an aggressive lymphoma, commonly associated with HIV infection. However, PBL can also be seen in immunocompetent patients and in patients with other immunodeficiencies [1-3].

Because of its distinct clinical and pathological features such as lack of CD20, plasmablastic morphology, and clinical course characterized by early relapses and subsequent chemotherapy resistance, PBL represent diagnostic and therapeutic challenge for pathologists and clinicians [4,5].

Despite the recent advances in therapy of aggressive lymphomas, patients with PBL, have the poorest outcome [6-8].

We describe a challenging case of a very aggressive form of plasmablastic lymphoma that had a dismal prognosis.

## 2. CLINICAL CASE

A 53-year-old- overweight women with history of gastric bypass and several hospitalizations presented for dyspnea and general status alteration due to anemia; diagnosed as hemolytic anemia and treated by corticotherapy without amelioration.

The most recent hospitalization revealed worsening of her symptoms, with weight loss of 5 kilos in one week, anorexia and lumbar pain.

Physical examination showed isolated splenomegaly with no signs of bleeding, adenopathy nor a palpable breast mass.

Laboratory studies revealed normocytic anemia with Hemoglobin =8.76 g/dl; Hct=26.6%;

MCV=89.2 fl; Platelets= 32 000/microliter; WBC = 7600/microliter with a slightly elevated reticulocyte count: 2.05; dosage of G6PD done was in low limit. ANA profile and coombs test were negative. Normal liver, renal function and coagulation tests were noted. Haptoglobin was very low < 0.04; LDH was high 1165 UI/litre.

Other laboratory studies showed: Corrected calcemia at 12 mg/dl and PTH level low.

Serum protein electrophoresis didn't reveal any peak, no hyperproteinemia was shown.

All these findings: Hemolytic anemia, hypercalcemia with low PTH and general status alteration were evocative of neoplastic process justifying the need of a total body CT scanner that didn't reveal any adenopathy.

Thus, bone marrow aspirate, biopsy and flow cytometry done were in favor of lymphoma.

On immunohistochemistry, a diffuse positivity of tumor cells with anti CD38 and anti-MUM 1 was noted. Anti CD3 CD20 CD25 CD68 MCO and CD117 were negative.

This immunohistochemical profile correlates with morphology and was compatible with PBL.

A magnetic resonance imaging (MRI) was performed and did not show any metastatic bone lesions, confirming the diagnosis.

Workup was completed by a full body PET CT scanner revealing diffuse hyperactivity of the bone marrow with hypermetabolic splenomegaly, hepatomegaly and bone lytic lesion of left cervical vertebra C1. Plus, several hypermetabolic left cervical lymphadenopathies were noted.

Our patient was diagnosed with stage 4 PBL IPI score 3.

HIV serology and Hepatitis B profile were negative.

The patient received three cycles of chemotherapy and immunomodulatory agents (lenalidomide and CHOP), that were not well tolerated with multiple opportunistic infections, deterioration of her mental status and major pain which all lead to death Three months after the diagnosis.

### 3. DISCUSSION

This case illustrates the difficulty of diagnosis and the aggressivity of PBL: It is a heterogenous disease.

Clinical features seen, are according to immunological status: HIV positive PBL, are more frequently seen in men at a median age of 42 years (in 78% of cases); but HIV negative PBL affect a higher proportion of female patients (34% of cases) at a median age of 55 years like our patient. Moreover, 76% of posttransplant PBL are man with a median age at presentation of 62 years old.

PBL in immunocompetent individuals seems to be more heterogenous in terms of sites of involvement.

Regarding pathological features, the diagnosis can be challenging because the tumor cells may be indistinguishable from plasmablastic myeloma or lymphoma with plasmablastic morphology.

The immunophenotype is similar to that in plasma cell neoplasm, positive for CD79a, CD38, CD138; and negative for B cell markers CD19, CD 20 and PAX-5.

Some cases express T cell markers CD2 and CD4.

Furthermore, the main differential diagnosis is plasmablastic or anaplastic multiple myeloma (MM).

The presence of monoclonal paraproteinemia and HIV infection can help to differentiate between the two as the first is seen in MM and the second is associated with PBL.

The PBL prognosis is poor; in HIV negative patients' median overall survival is 11 and 10 months in HIV positive; in post-transplant lymphoma the prognosis is very poor.

Nowadays, there is no standard treatment with current guidelines not recommending more aggressive treatments (such as EPOCH or hyperCVAD).

However, no proven survival benefit.

Given the association of plasmoblastic lymphoma with EBV, future therapy could include anti-viral agents.

In HIV positive patients, combination of Bortezomib and dose adjuvant EPOCH has shown efficacy.

Studies have shown efficacy of Brentuximab in cases expressing CD3.

Finally, Intrathecal (IT) prophylaxis should be considered with each cycle of EPOCH with or without Brentuximab; and considered of consolidative high dose chemotherapy (HDC) followed by autologous stem-cell transplantation (SCT) in first remission for candidates.

In HIV positive candidates, chimeric antigen receptor therapy (CART) should be started and optimized under supervision of an infectious disease specialist with experience in interaction between anticancer agents and Antiretroviral therapy (ART).

### 4. CONCLUSION

In conclusion, several factors have contributed to the bad prognosis in this clinical case; especially, the challenging diagnosis, aggressivity of the disease and complications related to immunomodulators agents. Palliative radiation therapy (RTH) should be considered on a case by case basis along with a concomitant chemotherapy.

### CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

### ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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