

CD3 Positive Gastric Plasmablastic Lymphoma in a HIV Negative Patient: A Case Report

Betul Bolat Kucukzeybek* Aylin Orgen Calli* Mehmet Ali Uyaroglu* Fusun Ozdemirkiran**

Aysegul Akder Sari* Ayse Vatansever* Bahriye Payzin**

*Izmir Katip Celebi University Ataturk
Training and Research Hospital- Dept.
Pathology

** Izmir Katip Celebi University Ataturk
Training and Research Hospital-
Hematology

ABSTRACT

Plasmablastic lymphoma is a rare and aggressive lymphoma characterized by the diffuse proliferation of large neoplastic cells resembling immunoblasts with an immunophenotype of plasma cells. A 47-year-old male was referred to our hospital with gastrointestinal bleeding, and a mass, 10 cm in diameter, was detected. An endoscopic biopsy was performed subsequently. A histopathological examination of the biopsy material revealed ulcer, alterations associated with ulcer, and further presented a diffuse infiltration of atypical cells with abundant cytoplasm and pleomorphic nuclei, some with crush artifacts in lamina propria. Immunohistochemically, the tumor cells were negative for cytokeratin, CD2, CD20, and PAX5; but they were positive for CD3, MUM1, CD38, and CD138. Ki67 proliferation index was as high as 95%. The case was signed out as CD3-positive plasmablastic lymphoma with clinical, histopathological, and immunohistochemical findings. The plasmablastic lymphoma case with an aberrant CD3 expression will be presented here, which is rarely observed in stomach.

Key words: Plasmablastic lymphoma, CD3, gastric lymphoma

Corresponding Author: Betul Bolat
Kucukzeybek
Izmir Katip Celebi University Ataturk
Training and Research Hospital
Department of Pathology-Izmir/Turkey
E-mail: bbkzeybek@yahoo.com

INTRODUCTION

Plasmablastic lymphoma (PBL) is a rare and aggressive B-cell lymphoproliferative disorder [1,2,3]. It is characterized by the diffuse proliferation of large neoplastic cells mainly resembling immunoblasts with an immunophenotype of plasma cells [1,4,5]. The incidence is high in HIV-positive patients, mostly in males. It is originally described and mostly observed in the oral cavity but may be seen in other localizations, predominantly at extranodal sites [1]. HIV positivity is less frequent in non-oral type PBL and can be seen in maxillary sinus, nasopharynx, and more rarely in the gastrointestinal tract and soft tissue [6,7].

A 47-year-old male patient diagnosed with gastric plasmablastic lymphoma with an aberrant CD3 expression will be presented below.

CASE

A 47-year-old male was referred to our hospital with gastrointestinal bleeding and an ulcero-vegetating, polypoid mass measuring 10 cm in diameter in the antrum. No pyloric involvement was detected during the upper gastrointestinal endoscopy. A biopsy was taken and sent to our clinic. Neither upper nor lower abdominal computed tomography showed any anomaly, and no abdominal lymphadenopathy was noted. Thoracic computed tomography showed no abnormalities including the bone structures. The patient was HIV-negative serologically. Serum and urine electrophoresis test results were normal. Histopathological examination of the biopsy material revealed ulcer, alterations associated with ulcer; and further presented a diffuse infiltration of atypical cells with abundant cytoplasm and pleomorphic nuclei, some with crush artifacts in lamina

propria (Figure 1a,1b,1c). Immunohistochemically, the tumor cells were negative for cytokeratin, CD2, CD20 (figure 1d), and PAX5; but they were positive for CD3 (figure 2a), MUM1 (figure 2b), CD38, and CD138 (figure 2c). Ki67 proliferation index was 95% (figure 2d). The case was signed out as CD3 positive plasmablastic lymphoma with clinical, histopathological, and immunohistochemical findings. Bone marrow biopsy proved to be negative for infiltration. The patient received 6 cycles of R-CHOP chemotherapy regimen. The PET-CT scan was negative after chemotherapy. During the 33-month follow-up, the patient presented no recurrence.

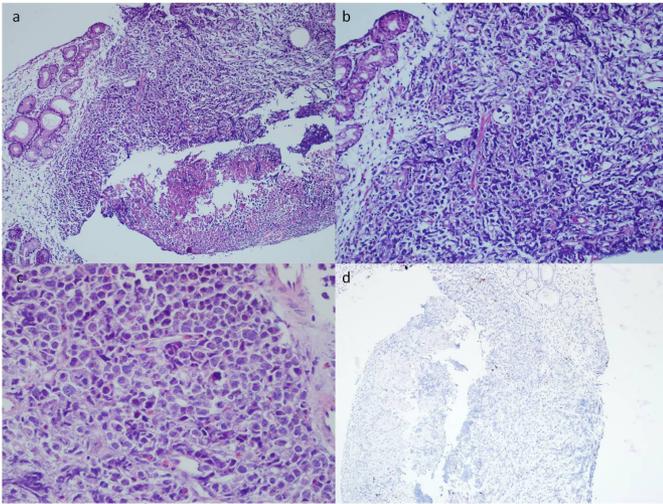


Figure 1a, 1b, 1c. Histopathological examination of the biopsy material revealed ulcer, alterations associated with ulcer, and further presented a diffuse infiltration of atypical cells with abundant cytoplasm and pleomorphic nuclei, some with crush artifacts in lamina propria.

Figure 1d. Immunohistochemically, the tumor cells were negative for CD20.

DISCUSSION

Plasmablastic lymphoma is a rare and aggressive B-cell lymphoma firstly described in 1997 by Delecluse et al. in the oral cavity of the immunocompromised patients [8]. In recent years, cases have been reported in HIV-negative patients, observed in the nasopharynx, maxillary sinus, orbita, skin, lung, gastrointestinal tract, soft tissue, anus, vulva, and mediastinum [1-6, 9-11]. HIV positivity is seen in 90% of the oral PBL cases, whereas the HIV positivity in non-oral PBL cases is less prevalent. EBV-positivity percentage is almost 100% in HIV-positive oral mucosal type PBL [6]. PBL is often associated with immunodeficiency such as HIV infection and EBV plays an important role in the tumorigenesis of the HIV-associated PBL [12]. However, detection of EBV positivity in 17% of HIV-negative PBL cases suggests that EBV may not be a unique causal agent in the pathogenesis

of PBL [13]. The case presented herein was not immunocompromised or HIV-positive.

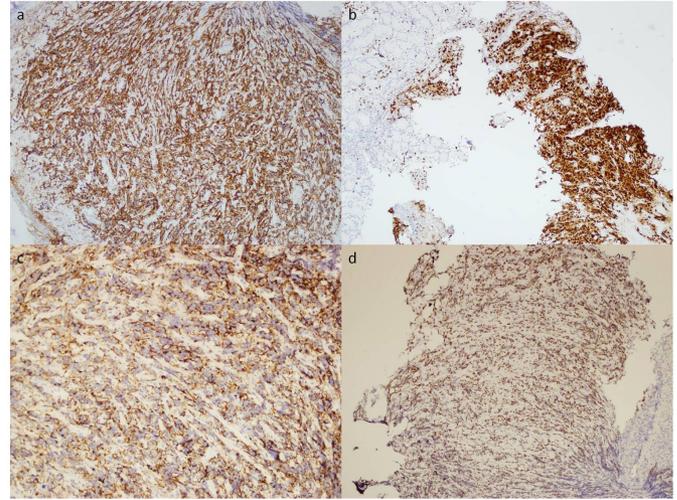


Figure 2. Immunohistochemically, the tumor cells were positive for CD3 (Figure 2a), MUM1 (Figure 2b) and CD138 (Figure 2c).

Figure 2d. Ki67 proliferation index was 95%.

Histopathologically, PBL is characterized by apoptotic cells and the proliferation of large neoplastic cells having numerous mitoses with a diffuse growth pattern. Immunohistochemically, pan B markers (CD20, CD22, PAX5, CD79a) are often negative and plasma cell associated markers (IRF4/MUM1, CD38, CD138) are strongly positive. Ki67 proliferation index is quite high and the aberrant expression of T-cell markers (CD3, CD4, CD43, CD7) is reported in rare cases [1,6,9]. Our case had a strong positivity of plasma-cell-associated markers and also of a T-cell marker, CD3. Immunohistochemically, CD3 positivity with CD20 and CD79a negativity may suggest a T-cell non-Hodgkin lymphoma in a differential diagnosis but that was excluded by the negativity of other T-cell markers (CD2, CD7) and a strong positivity of plasma-cell-associated markers.

Extramedullary plasmacytoma and plasmablastic plasma cell myeloma are other matters which should be considered for differential diagnosis as they have similar morphological and immunophenotypical features [2,6,9,14]. Clinicopathological correlation is essential in the differential diagnosis. Serum monoclonal protein negativity and high Ki67 proliferation index are also useful in this context. Serum and urine electrophoresis tests, high Ki67 proliferation index and computed tomography findings with no abnormalities of bone structures were helpful for the differential diagnosis of the case presented.

Plasmablastic lymphoma is very aggressive and most of the patients are lost within the first year of diagnosis [1]. Antiretroviral therapy and chemotherapy are reported to be quite helpful in improving the prognosis [15]. Our patient received 6 cycles of R-CHOP chemotherapy regimen. The PET-CT scan was negative

after chemotherapy. During the 33-month follow-up, the patient presented no recurrence.

Extranodal PBL is a rare and aggressive lymphoma that should be taken into account for differential diagnosis especially if the gastrointestinal tract shows a negativity with lymphoid and B-cell markers confused with poorly differentiated carcinomas; nevertheless, it poses diagnostic difficulties with T-cell lymphomas presenting aberrant expressions of T-cell markers.

REFERENCES

1. WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. Swerdlow SH, Campo E, Harris NL, et al, Eds. WHO Press, Geneva, 2008
2. Wang HW, Yang W, Sun JZ, Lu JY, Li M, Sun L. Plasmablastic lymphoma of the small intestine: Case report and literature review. *World J Gastroenterol.* 2012;18:6677-81.
3. Hashimoto M, Inaguma S, Kasai K, Kuwabara K, Noda N, Hayakawa M, et al. Plasmablastic lymphoma of the stomach in an HIV-negative patient. *Pathol Int.* 2012;62:763-70.
4. Rafaniello Raviele P, Pruneri G, Maiorano E. Plasmablastic lymphoma: a review. *Oral Dis.* 2009;15: 38-45
5. Castillo JJ, Reagan JL. Plasmablastic Lymphoma: A Systematic Review. *ScientificWorldjournal.* 2011 Mar 22;11: 687-696
6. *Diagnostic Pathology: Lymph Nodes and Spleen with Extranodal Lymphomas:* Published by Amirsys. Medeiros, L Jeffrey, editor. First edition. Salt Lake City, Utah, United States.: Amirsys, 2011
7. Angeleri A, Rocher A.E, Myburg C, Avagnina A , Aparo V, Palaoro L.A. Plasmablastic lymphoma involving the stomach in an HIV positive man. *Cytopathology.* 2016;27:293-5
8. Delecluse HJ, Anagnostopoulos I, Dallenbach F, Hummel M, Marafioti T, Schneider U, et al. Plasmablastic lymphomas of the oral cavity: a new entity associated with the human immunodeficiency virus infection. *Blood.* 1997;89:1413-20
9. Suzuki Y, Yoshida T, Nakamura N, et al. CD3- and CD4-Positive Plasmablastic Lymphoma: A Literature Review of Japanese Plasmablastic Lymphoma Cases. *Inter Med.* 2010;49:1801-1805
10. Bagul N, Mamatha GS, Mahalle A. Plasmablastic lymphoma of gingiva mimicking a reactive lesion: a case report. *Case Rep Dent.* 2012;2012:259307.
11. Mansoor M, Alani FS, Aslam MB, Kumar SN, Sahasrabudhe N, Khan D. A case report of cecal plasmablastic lymphoma in a HIV-negative patient. *Eur J Gastroenterol Hepatol.* 2012;24:332-5.
12. Castillo J, Pantanowitz L, Dezube BJ. HIV-associated plasmablastic lymphoma: lessons learned from 112 published cases. *Am J Hematol.* 2008;83:804-809
13. Lorbach RB, Hsi ED, Dogan A, Fend F. Plasma cell myeloma and related neoplasms. *Am J Clin Pathol.* 2011;136:168-182
14. Vega F, Chang CC, Medeiros LJ, Udden MM, Cho-Vega JH, Lau CC, et al. Plasmablastic lymphomas and plasmablastic plasma cell myelomas have nearly identical immunophenotypic profiles. *Mod Pathol.* 2005;18:806-15.
15. Sarode SC, Zarkar GA, Desai RS, Sabane VS, Kulkarni MA. Plasmablastic lymphoma of the oral cavity in an HIV-positive patient: a case report and review of literature. *Int J Oral Maxillofac Surg.* 2009;38:993-999.