An immunocompetent patient with EBV-related plasmablastic lymphoma in the small pelvis: a case report.

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Background

Plasmablastic lymphoma (PBL) is a rare form of aggressive non-Hodgkin's lymphoma. Although PBL is most frequently observed in the oral cavity of human immunodeficiency virus (HIV)-positive patients, rare cases of PBL have been reported in extra-oral sites, as well as in immunocompetent patients.

Case report

- 32-year old man
- No arguments for immunodeficiency
- Presenting with night sweats and raised inflammatory markers (C-reactive protein 20 mg/L, sedimentation rate 85 mm/h)
- Abdominal CT scan revealed an acetabular mass and osteolytic lesion in the small pelvis.
- Histological examination: lymphoproliferative process with plasmablastic differentiation, monotypic IgA lambda expression and a rather low proliferation index of 20%
- Immunophenotype: CD138 +, CD56+, CD20-, HHV8-
- CISH: EBV- encoded RNA +
- Therapy:
 - Stable disease after VCD, involved field radiotherapy and R-CHOEP
 - Partial metabolic remission after three courses KRd

Evolution of laboratory findings

References:

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Discussion

Plasmablastic lymphoma is a rare subtype of non-Hodgkin's lymphoma that displays some features of DLBCL and some features of MM (1). Despite its predominant occurrence in the oral cavity of HIV-positive patients, PBLs can also affect individuals without HIV infection, with different clinicopathological characteristics (2), such as short overall survival and lower rates of oral involvement and Epstein-Barr virus-encoded RNA expression (3). Therapy often consists of CHOP-like regimens, but prognosis is dismal with <40% of patients living beyond two years (1). However, age ≥60, advanced stage, bone marrow involvement, no chemotherapy, Ki-67 expression >80%, and HIV-negative status were associated with worse overall survival (4). Because of the distinct clinicopathological characteristics between HIV-positive and HIV-negative patients with PBL and no standard of care, further research is needed to evaluate different subtypes and develop treatment recommendations.

Conclusion

Despite the fact that PBL is associated with a short overall survival there seems to be an indolent course in this case, with no response to several chemotherapy regimens. It is known that immunocompetent patients with PBL have distinct clinicopathological characteristics with no standard of care, so further research is needed to evaluate different subtypes and develop treatment recommendations.