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A RARE CASE OF PLASMABLASTIC LYMPHOMA PRESENTING WITH A BULKY RENAL MASS IN AN IMMUNOCOMPETENT PATIENT

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INTRODUCTION

Plasmablastic lymphoma (PBL) is a rare and aggressive subtype of large B-cell lymphomas. PBL patients are predominantly immunocompromised, or HIV infected. Activated B cells undergoing somatic hypermutation and class-switching recombination, are believed to be the tumor cell origin. Histologically, the tumor exhibits a diffuse growth pattern leading to impairment of structure with large, atypical cells resembling immunoblasts. The specificity of PBL cells is lacking B-cell markers (CD19, CD20, and PAX-5) expression and having an immunophenotype similar to plasma cells such as showing CD38, and CD138 positivity. EBV-encoded RNA (EBER) has been seen in around 80% of cases and over 66% of cases present MYC rearrangements.

More than half of PBL arises in the oral cavity but may develop in various tissues or organs and it has been shown regardless of the location and presence of HIV and EBV, PBLs display comparable clinicopathologic features.

Intensive treatment protocols such as DA-EPOCH with the addition of new drugs like bortezomib are recommended treatment for fit patients.

CASE

A 59-year-old HIV-negative male presented with decreased urine output, high creatinine (5,85 mg/dl), high serum LDH level (588 IU/L) levels, and anemia (Hb: 6,8 g/dL, MCV:88fL) with bilateral grade 2 hydronephrosis on USG. PET-CT showed a bulky mass in the pelvis sized 18*15*25cm which covered nearly the right half of the pelvis and infiltrated the renal parenchyma (SUDmax:7,5), and a mass sized 30*24 mm in the liver (SUDmax:4,5). A biopsy of the kidney was decided. The histology revealed diffuse large-cell infiltration. The infiltrating cells were CD20, CD3, and PAX5 negative but CD138, and MUM1 positive with a high Ki67 labeling of 90%. EBER expression by in situ hybridization was strongly positive (70%) and clonality by kappa light chain restriction was documented.

He had a monoclonal gammopathy of IgG kappa type. Bone marrow did not show any evidence of lymphoproliferative disease, or plasma cell neoplasia. The karyotyping result was normal. DA-EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin) with bortezomib was chosen as treatment. The response was rapid. After two courses of chemotherapy serum creatinine decreased to normal levels (0,6 mg/dl) and hemoglobin increased to 8,4 g/dL.



PET CT image after 3 courses

CONCLUSIONS

In our case, PBL arises with a rare form as developing in immunocompetent biology, and in extraoral locations as huge masses.

Intensive chemotherapy combined with antimyeloma drugs such as proteasome inhibitors, immunomodulators, and monoclonal antibodies provides significant improvement in PBL but the management of PBL remains challenging.

The aggressive nature and propensity for relapse dictate there is still a need for clinical studies with large case series especially patients with relapsed and resistant PBL.

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