

中文題目：初始表現似晚期口腔癌的漿母細胞性淋巴瘤之個案報告

英文題目：Plasmablastic lymphoma initially presented as end stage oral cancer

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Introduction

Plasmablastic lymphoma(PBL) is an aggressive B-cell malignancy that is most commonly discovered in the rectum and the anal canal and has a strong relationship with HIV infection. However, it could also be found in HIV-negative persons. In this case report, we present a case diagnosed with plasmablastic lymphoma who had an initial presentation with an oral mass.

Case presentation

A 62-year-old male with medical history of hypertension, diabetes mellitus and cigarette smoking for 40 years suffered from an enlarging oral mass (figure 1) for about 2 months. Recent weight loss (5kg in 2 months), nasal obstruction and hemifacial numbness were also noted. Physical examination revealed affected territory of left fifth cranial nerve (V2 maxillary branch). There was no fever, night sweat, bone pain nor vision loss. Under suspicion of oral cancer, biopsy of oral mass was performed, and pathologic report revealed plasmablastic lymphoma. (Kappa stain negative, Lamda stain positive, LCA negative, CD-138 positive, EBER stain positive). Computer tomography of oral cavity showed maxillofacial tumor involving left maxilla, nasal cavity, palate, masseter and pterygoid muscles with severe bony destruction and erosion of left orbital floor. Positron emission tomography showed bone marrow, liver, lung, and the lymph nodes in neck and bilateral bronchopulmonary regions involvement (stage IV). Bone marrow biopsy showed confirmatory plasmablastic lymphoma, with CD138(+) and predominant lambda chain. Laboratory showed hypercalcemia (5.4 mg/dL), a normal albumin level (4.69 g/dL), beta2-Microglobulin (562 ug/dL), negative HIV, positive EBV and monoclonal lambda light chain in urine and blood. Therefore, the first course chemotherapy with EPOCH (Etoposide 45.1mg/m²+ Epirubicin 9mg/m²+ Vincristine 0.5mg/m²+ Endoxan 700.6mg/m² + prednisone) was initiated during 2021/04/01-04/05. After 5 cycles of EPOCH, follow up computer tomography showed markedly shrinkage of extensive maxillofacial plasmablastic lymphoma involving left maxilla, nasal cavity, palate, upper gingiva, masseter and pterygoid muscle.

Discussion

Plasmablastic lymphoma (PBL) is a rare subtype of large B-cell lymphoma that has an aggressive behavior and commonly associated with HIV-positive patients (but it can also be diagnosed in HIV-negative patients, as presented in our case). It is also found in patients infected by Epstein–Barr virus (EBV), and is more predominant in males [1,4]. In HIV-positive patients, the most common site of development of PBLs is the rectum and the anal canal. In HIV-negative PBL, the most common site of development of PBLs is the oral cavity[4]. Advanced clinical stage and elderly age are associated with poor survival and is identified as poor prognostic factors for HIV-negative PBL patients in Asian[3]. However, most patients present at an advanced stage (III or IV), with frequent bone marrow involvement when they are diagnosed [1]. There is no standard treatment for PBL (radiotherapy, chemotherapy or both) and several chemotherapy combinations (such as CHOP(cyclophosphamide, doxorubicin, vincristine, and prednisone), hyper-CVAD-MA (hyperfractionated cyclophosphamide, vincristine, doxorubicin, dexamethasone, and high-dose methotrexate and cytarabine), EPOCH etc.) have been used with hardly any differences on its survival outcome[1]. Bortezomib was used for treatment and showed promising with combination with EPOCH (complete response rate exceeding 90% as well as a 5-year overall survival rate of 65%)[5]. In conclusion, we should be aware of possible PBL when a patient is present with an oral mass, while a delayed diagnosis may negatively impact on the patient's treatment and survival.

References:

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