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Plasmoblastic lymphoma in the mediastinum in an HIV positive woman.

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Abstract

Plasmoblastic Lymphoma (PBL) is an hematologic neoplasia with a diffuse proliferation of large cells, most of which resemble B inmunoblasts or plasmablasts that have a CD20-negative plasmacytic phenotype. This disease affects the oral cavity of immunodeficient patients, but a few cases have been reported at extraoral sites.

A 44-year-old woman, positive for HIV, suffered from PBL detected in the mediastinum without involving the oral cavity. The primary diagnosis was made by cytology, but differentiation from other lymphomas was achieved by means of flow cytometry and immunostaining.

Flow cytometry: Lymphocyte population: 1% T lymphocytes; 0.04% B lymphocytes; 0.13% Natural Killer; 96% plasma cells monoclonal for kappa chain. CD38+, CD138++, CD19-, CD56 -/+ (21%), CD 81+, CD117-, CD27 (-/+), CD45-*Immunostaining of plasmacytoid cells*: CD138+; MUM1+; Ki67 90%; Kappa chains+ ; Lambda chains - ; CD20 - ; CD45 - ; CD30 - ; LMP1 - ; HHV8 -.

The strong association of this lymphoma with patients with HIV indicates that the presence of a PBL should be suspected in these cases.

Keywords

Cytology; VIH; plasmablastic lymphoma; pleural fluid; non-hodgkin lymphoma.

Introduction

Plasmablastic lymphoma (PBL) is an aggressive hematologic neoplasia with a diffuse proliferation of large cells, most of which resemble B inmunoblasts or plasmablasts that have a CD20-negative plasmacytic phenotype. The association between PBL and immunosuppression has been clearly proven [1].

Represents 2.6% of all non-Hodgkin lymphomas associated with HIV and was originally described as a disease that specifically affects the oral cavity of immunodeficient patients. Nowadays, the incidence of all the Non-Hodgkin lymphomas (NHL) in HIV patients is near 194 cases per 100,000 person. Most NHLs are PBL, but the exact data on this type of lymphoma is lacking in the literature [2]. The coexistence of PBL with HIV aggravates the outcome of this disease. Patients with HIV and PBL have a lower survival than those without PBL [3]. Several cases have been reported at extraoral sites, including the maxillary sinus, nasopharynx, stomach, small bowel, anus, lungs, skin, soft tissue, mediastinum, and heart. The oral cavity represents the primary site of origin in 51% of cases, while 20% of extraoral PBLs involve lymph nodes. Extraoral PBL occurs less frequently in non-deficient patients (70% of cases) and spreads more frequently (57% of patients are in stage IV at the time of diagnosis) [4-6].

Case Report

A 44-year-old woman, positive for HIV, was admitted to the Hospital for presenting asthenia, adynamia, and respiratory distress. She was diagnosed with a Plasmablastic Lymphoma (PBL) detected in the mediastinum without involving the oral cavity. The primary diagnosis was made by cytology, but differentiation from other lymphomas was achieved by means of flow cytometry and immunostaining.

The following tests carried out on pleural fluid

Cytology: Cell count: 15000 cells/mm^3, showing 100% immature cells of the lymphoid series with plasmoblastic differentiation (Figure 1). Intermediate to large size cells with moderate cytoplasm, round nuclei and prominent nucleoli were noted in pleural effusion.

Chemistry: pH: 6.8; Glucose <10 mg/dl; Protein: 3.7 g/dl; Albumine: 1.8 g/dl; LDH 2961 UI/l.

Flow cytometry: Lymphocyte population: 1% T lymphocytes; 0.04% B lymphocytes; 0.13% Natural Killer; 96% plasma cells monoclonal for kappa chain. CD38+, CD138++, CD19-, CD56 -/+ (21%), CD 81+, CD117-, CD27 (-/+), CD45-.

The following tests were carried out on blood

Cytology: haematocrit 34%; haemoglobin 11 g/dl; leucocytes 12340 /mm^3; platelets 497000/mm^3.

In Bone marrow aspiration puncture, plasma cells and B lymphocytes are not observed by flow cytometry. The following tests were carried out on *mediastinum biopsy*. H&E: Monomorphic proliferation of large, round to oval cells, with abundant cytoplasm, eccentrically placed nuclei with a single prominent central nucleolus and frequent mitotic figures.

Immunostaining of plasmacytoid cells: CD138+; MUM1+; Ki67 90% (Figure 2); Kappa chains+ ; Lambda chains - ; CD20 - ; CD45 - ; CD30 - ; LMP1 - ; HHV8 -.

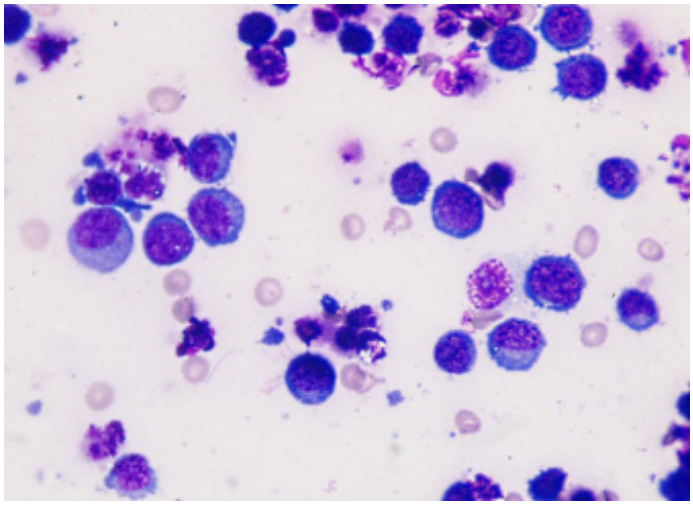

Figure 1: Efussion pleural: Immature cells of the lymphoid series with plasmoblastic differentiation (400x. Giemsa).

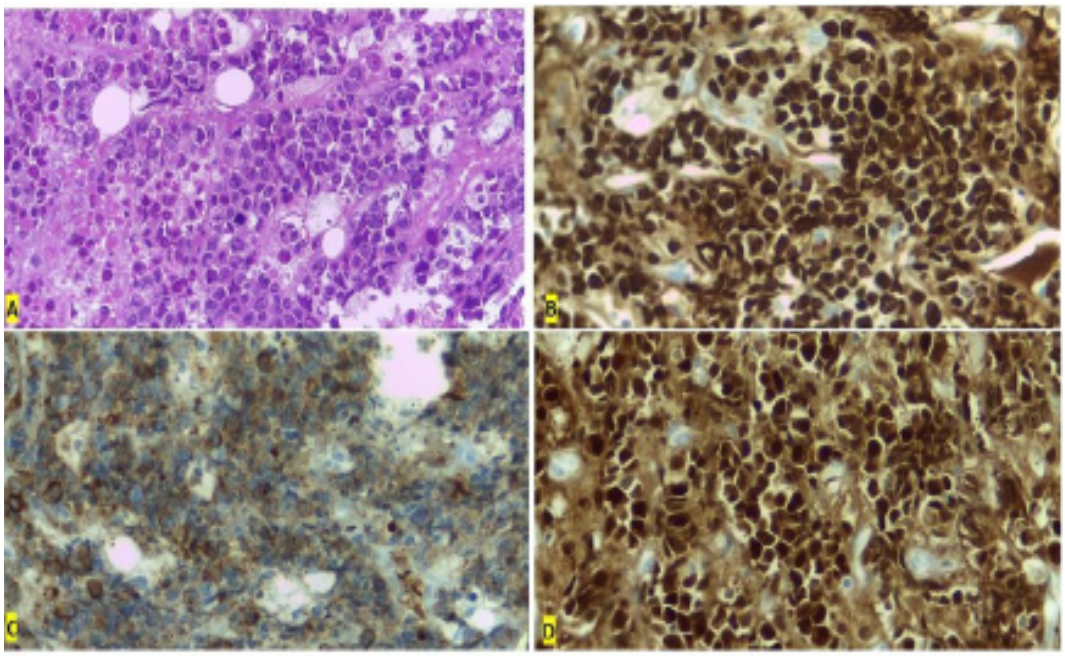

Figure 2: Mediastinal biopsy: a) H&E; b) Ki67+; c) CD138+; d) MUM1+ (400x).

Discussion

The diagnostic criteria for PBL are based on classical morphology (large lymphoid cells, with abundant cytoplasm, eccentric nuclei, prominent central nucleolus, and frequent perinuclear halo) with a terminally differentiated B-cell immunophenotype, characterized by minimal or absent expression of leukocyte common antigen (CD45) and B-cell antigens (CD20 and CD79a). In contrast, tumor cells in PBL are invariably immunoreactive for CD138. Markers associated with the post germinal center are generally expressed: MUM1 (multiple myeloma 1) and CD38.

The MUM-1 is a lymphocyte-specific transcriptional activator expressed in the final differentiation stage of intra-germinal center B cells. MUM-1 is expressed by post-germinal center B cells, plasma cells, and

subset of T cells VS38c (rough endoplasmic reticulum-associated antigen) is expressed by plasma cells and cells with plasmacytoid differentiation, therefore it could be used as a PBL marker.

However, both MUM1 and VS38c are not very specific, and the use of CD138 should be always required to confirm the diagnosis of PBL.

Rearrangements of the MYC oncogene and immunoglobulin gene in immunoblasts are likely responsible for the development of this type of lymphoma [7-9].

The case presented is a PBL of extraoral location, in this case in the mediastinum, in a patient positive for HIV. The presence of PBL in the mediastinum, without affecting the oral cavity, is a very rare event. I feel it would be better if you could change it in some other words. In HIV patients with associated lymphoma, the possibility of a PBL should always be considered, due to the strong association between both pathologies.

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