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Vulvar and gastric involvement in plasmablastic lymphoma

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Summary

Plasmablastic lymphoma is highly malignant and invasive. It is most commonly found in gastrointestinal tract and is strongly associated with human immunodeficiency virus (HIV). The authors report a case of a 37-year-old HIV-positive African American woman with a vulvar mass diagnosed as plasmablastic lymphoma. Involvement of the upper gastrointestinal tract had been detected by endoscopic biopsy, suggesting that any patient diagnosed with plasmablastic lymphoma in unusual locations should be investigated for primary lesions in the gastrointestinal tract.

Key words: Plasmablastic lymphoma; Human immunodeficiency virus; Gastrointestinal tract.

Introduction

Plasmablastic lymphoma lesions are most commonly found in the mouths and gastrointestinal tracts of people infected with human immunodeficiency virus (HIV). Involvement of the vulva with any type of lymphoma appears to be very rare [1]. The authors present the case of a 37-year-old female with a vulvar tumor diagnosed as plasmablastic lymphoma.

Case Report

A 37-year-old African American woman with a ten-year history of HIV infection presented complaint of a painful mass with purulent discharge in the left side of the vulva for four months with rapid growth and the left lower limb hemiparesis for six months. She had received highly active antiretroviral therapy; however, at the time of admission, she was not using it. Physical examination revealed a subcutaneous, tender mass of seven cm in diameter with ipsilateral lymphadenopathy (Figure 1). The first hypothesis was an abscess or carcinoma of the Bartholin gland. A computed tomography scan was performed and showed lesions compatible with cerebral toxoplasmosis, whereas the abdomen showed right renal vein and inferior vena cava thrombosis. Fine-needle aspiration cytology was performed although the result was indeterminate for the vulvar aspiration, whereas the undifferentiated metastatic malignancy in the lymph node suggested a tumor of neuroendocrine origin. After this procedure, the patient developed both melena and hematemesis, and an endoscopy and colonoscopy with biopsy were performed. The gastric biopsy showed plasmablastic lymphoma infiltrating the gastric mucosa, and immunohistochemical analysis was positive for LCA and CD138 (Figure 2). In addition, a vulval biopsy was performed. A sufficient quantity of friable tissue was removed and histopathological examination revealed plasmablastic lymphoma with immunohistochemistry analysis showing positivity for LCA and CD138 (Figure 3).

Discussion

Cancer is an important source of morbidity and mortality among people infected with HIV. Advanced HIV infection characterized by marked immunosuppression is itself a risk factor for malignancy. Non-Hodgkin's lymphoma, Kaposi sarcoma, and cervical cancer are acquired immune deficiency syndrome (AIDS)-defining cancers, caused by a loss in immune control of oncogenic viruses [2].

Non-Hodgkin lymphoma is the second most common AIDS-defining cancers. Plasmablastic lymphoma is a clin-

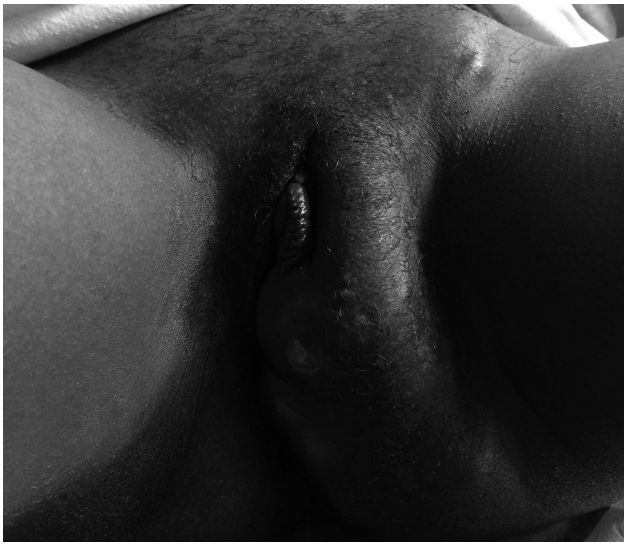

Figure 1. — Patient with a seven-cm diameter mass with ipsilateral lymphadenopathy.

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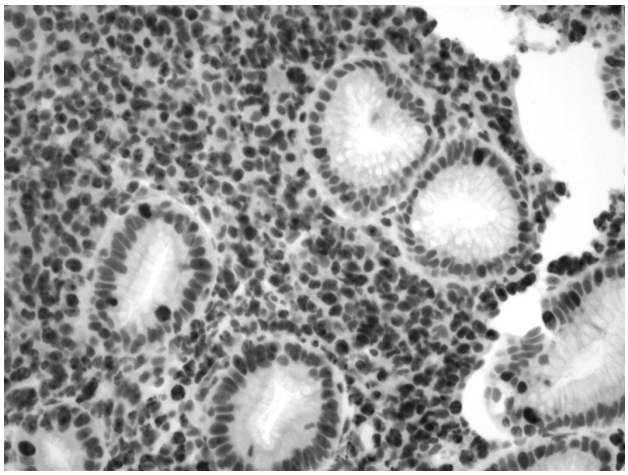

Figure 2. — Microscopic appearance of plasmablastic differentiation (hematoxylin–eosin, original magnification ×400).

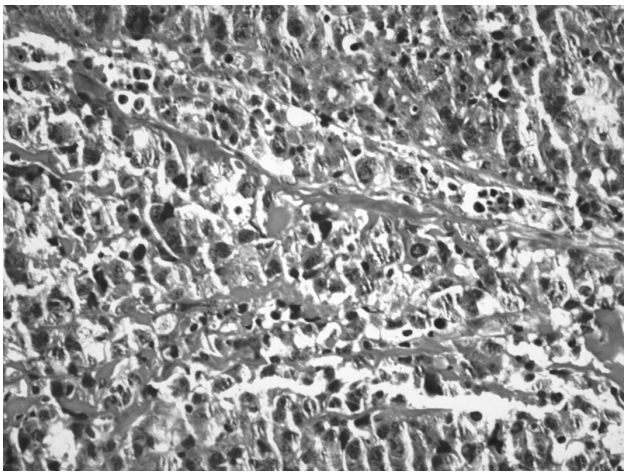

Figure 3. — Gastric immunohistochemical staining by CD138 staining showing positivity (original magnification x40).

ical entity defined by the World Health Organization as a form of non-Hodgkin's lymphoma characterized by plasma cell differentiation and an immunoblastic morphology. It is further characterized by its aggressive nature, plasmacytic differentiation, and for its predilection of involving the oral cavity and gastrointestinal tract of HIV-positive individuals [3, 4].

Plasmablastic lymphoma lesions are most commonly found in the mouths and gastrointestinal tracts of HIV-positive patients. The extranodal lymphoid organs, central nervous system, peripheral lymph nodes, paranasal sinuses, mediastinum, lung, liver, and bone marrow may also be involved. There have been recent reports of plasmablastic lymphoma in HIV-negative patients where lesions mostly occurred in extranodal tissues, including the skin, soft tissue, maxillary sinus, and gastrointestinal tract [4].

Non-Hodgkin's lymphoma constitutes 40% of malignant lymphomas. The involvement of the vulva, either primarily or secondarily, appears to be very rare [1].

Plasmablastic lymphoma remains a diagnostic challenge considering its peculiar morphology and an immunohistochemical profile similar to plasma cell myeloma. Furthermore, it is a therapeutic challenge with a clinical course characterized by a high rate of relapse and death [5].

The diagnosis of plasmablastic lymphoma can be complicated by its morphologic resemblance to other lymphoid and myeloid malignancies. Morphological features can be used to distinguish plasmablastic lymphoma from well-differentiated plasma cell neoplasms. However, highly aggressive plasma cell myeloma may contain a predominance of plasmablasts, which can closely resemble the malignant cells of plasmablastic lymphoma. Plasma cell myeloma and plasmacytoma may occasionally occur in the setting of AIDS; however, their association with AIDS is not as strong as that which has been reported for plasmablastic lymphoma [6].

The genetic and molecular mechanisms that may be involved in the pathogenesis are not well known. The contribution of HIV to the pathogenesis of plasmablastic lymphoma might develop because of the duration and degree of immunodeficiency or immunosuppression; the induction of chronic antigenic stimulation leading to a chronic B-cell proliferation; the loss of immune control of oncogenic herpesvirus such as Epstein–Barr virus or an incomplete immune reconstitution or factors unrelated to immune dysfunction [7].

A biopsy with accurate pathological and immunohistological testing and a high level of clinical suspicion are essential for correct diagnosis. The morphological differential diagnosis includes poorly differentiated and undifferentiated carcinoma, lymphoblastic lymphoma, plasmablastic variant of Burkitt's lymphoma, and anaplastic plasmacytoma [7].

Immunohistochemical studies have observed the lack of expression of the cell cycle inhibitors p27 and p16 and strong expression of p53 in a number of cases, suggesting that inactivation of these genes may contribute to the high rate of proliferation commonly seen in plasmablastic lymphoma [8].

Plasmablastic lymphoma is highly malignant and invasive, associated with poor prognosis, and has low sensitivity to chemotherapy. It is unknown if the outcome of patients with plasmablastic lymphoma has improved in the era of highly active antiretroviral therapy [4, 5].

There is currently no standard chemotherapy protocol, and most regimens are based on the CHOP regimen: cyclophosphamide, hydroxydoxorubicin, vincristine, and prednisone. A recent international multicentre analysis of HIV-positive cases with plasmablastic lymphoma showed that intense chemotherapy did not extend survival time; the rate of complete remission was 66% and the median overall survival was 11 months [4].

In the present case, the first diagnosis was vulvar plasmablastic lymphoma, and if the patient had not presented with specific gastric symptoms, such as hematemesis and melena, no investigation of the gastrointestinal sites would have been performed. This case suggests that any patient diagnosed with plasmablastic lymphoma in unusual locations should be investigated for primary lesions in the gastrointestinal tract.

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