PRIMARY MENINGEAL BURKITT-TYPE LYMPHOMA PRESENTING AS THE FIRST CLINICAL MANIFESTATION OF ACQUIRED IMMUNODEFICIENCY SYNDROME

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ABSTRACT - The purpose of this study is to report a rare case of primary meningeal high grade Burkitt-type lymphoma presenting as the first clinical manifestation of acquired immunodeficiency syndrome. A 38-year-old Caucasian man, with a negative past medical history, sought treatment after experiencing global headache for five days. CT-Scan revealed a right fronto-temporo-parietal hyperdense subdural expansive mass. A craniotomy was performed and a hard white subdural was microsurgically dissected. Some hours after the surgery, the patient developed hemispheric cerebral edema and intracranial hypertension syndrome. Decompressive craniotomy was performed and the patient had an excellent recovery. Screening blood tests diagnosed human immunodeficiency virus infection. Further investigation ruled out systemic diseases. Eleven days after the initial surgery, the patient developed an acute respiratory failure and sepsis, dying on that day. Pathological studies diagnosed Burkitt-type lymphoma.

KEY WORDS: Burkitt-type lymphoma, meningeal neoplasm, acquired immunodeficiency syndrome.

Linfoma de Burkitt primitivo da meninge como primeira manifestação clínica da síndrome da imunodeficiência adquirida

RESUMO - O objetivo desse estudo é relatar um caso de linfoma de Burkitt de alto grau primitivo da meninge, que se apresentou como primeira manifestação clínica da síndrome de imunodeficiência adquirida. Um homem branco, de 38 anos, previamente hígido, referia cefaléia holocraniana há cinco dias. A TC de crânio evidenciou coleção hiperdensa subdural na região fronto-temporoparietal direita. Após craniotomia fronto-temporal direita, um tumor branco e rígido de localização subdural foi microcirurgicamente ressecado. Algumas horas após, o paciente apresentou edema cerebral hemisférico e hipertensão intracraniana, tendo sido submetido à craniotomia descompressiva com excelente melhora clínica. Testes sorológicos evidenciaram infecção por vírus da imunodeficiência humana. Investigações complementares afastaram outras doenças sistêmicas. Onze dias após a primeira cirurgia, o paciente apresentou insuficiência respiratória aguda e sepse, evoluindo para o óbito. Análise histopatológica evidenciou linfoma de Burkitt.

PALAVRAS-CHAVE: linfoma de Burkitt, tumor meningeal, síndrome da imunodeficiência adquirida.
Primary CNS lymphoma has a much higher incidence in patients with established immune deficient status, and in acquired immunodeficiency syndrome (AIDS) patients it represents the most frequent brain tumor. A high prevalence is also observed in renal and cardiac transplants, patients with IgA deficiency, or Wiskott-Aldrich syndrome. A number of clinical reports suggest an increasing incidence of this tumor over the past decades, however there is no published data about it as the first clinical manifestation of AIDS.

The purpose of this study is to report and discuss a rare case of primary meningeal high grade Burkitt-type lymphoma, presenting as the first clinical manifestation of AIDS.

CASE
A 38-year-old Caucasian man, with a negative past medical history, sought treatment after experiencing global headache of moderate intensity for five days. There were no other complaints. The patient's neurological examination was intact. Computed tomography (CT-scan) revealed a right fronto-temporo-parietal hyperdense subdural expansive mass, with a 1cm midline brain shift, suggesting an acute subdural hematoma (Fig 1).

A right fronto-temporo-parietal craniotomy was performed. A hard white subdural mass with extension to frontal, temporal and parietal spaces was microsurgically dissected (gross-total removal) (Fig 2). A few hours after the surgery, the patient developed right hemispheric cerebral edema and intracranial hypertension syndrome. Decompressive craniotomy was performed and the patient had an excellent recovery. Screening blood tests (2 samples) diagnosed human immunodeficiency virus (HIV) infection some days after the surgeries. This fact was not known neither by the patient nor by his family. Further complementary radiological and hematological investigation ruled out systemic diseases. Eleven days after the initial surgery, the patient developed an acute respiratory failure and sepsis by a Pneumocystis carinii infection. The patient died on that day.

Pathological studies – At microscopy the biopsy was characterized by a diffuse monotonous infiltrate of medium sized cells, with a slight molding pattern. The distinct starry-sky pattern caused by interspersed tingible-body macrophages was present. The cells showed round nuclei, with coarse chromatin and basophilic nucleoli, with a notable rim of basophilic cytoplasm. Karyorrhectic debris and mitotic figures were frequent. Immunohistochemically, the tumor expressed positivity for B cell markers CD20 and CD79a. CD10 and Bcl-6 were also positive. The proliferative index measured by the Ki-67 was very high bearing 100%. The negativity to Bcl-2 also helped in the differential diagnosis with diffuse large B cell lymphoma. These features are compatible with high grade Burkitt-type lymphoma (Fig 3).

DISCUSSION
CNS lymphoma is a rare tumor, representing 1 - 2% of all primary brain tumors. The majority of CNS lymphomas consist of secondary involvement of the brain, spinal cord, or covering membranes by nodal or extranodal (rather than CNS) lymphomas. Primary CNS lymphoma is by definition an extranodal lymphoma arising in the central nervous system in the absence of systemic disease. These tumors are primarily the non-Hodgkin type, and are high grade and of B-cell lineage, with large cell morphological characteristics, although nearly all other types of malignant lymphoma of the CNS are on record.

Primary CNS lymphoma has a much higher incidence in patients with established immune deficient status, especially in AIDS patients. In its early stages, human immunodeficiency virus infection has no symptoms or causes only a flu-like illness with many of the following symptoms: fever, sore throat, rash, nausea and vomiting, diarrhea, fatigue, swollen lymph nodes, muscle aches, headaches and joint pain. When the number of CD4 cells drops significantly, the patient develops AIDS and opportunistic infections may overcome. Some of the germs that can cause these include Candida fungus, cryptococcosis, cytomegalovirus, herpes simplex virus, Mycobacterium...
Fig 2. Transoperative photography showing the meningeal Burkitt-type lymphoma with subdural extension.

Fig 3. A) Histological examination, H & E; B) CD10 (56C6): positive (neoplastic B-cells); C) CD 20 (L26): positive (neoplastic B-cells); d) Ki 67 (MIB 1) immunostaining proliferative index.
avium complex and Pneumocystis carinii. A tumor may also develop, including cervical cancer, Kaposi's sarcoma and certain types of non-Hodgkin's lymphoma, as brain lymphoma. Primary meningeal Burkitt-type lymphoma as the first manifestation of AIDS has not been reported previously.

The incidence of primary central nervous system lymphomas among immunocompetent and immunocompromised patients has been on the rise in the past three decades. This rise can be partially attributed to the prolonged survival of AIDS patients and the extensive use of immunosuppressive therapy in organ transplantation and autoimmune disease.

Most authors have found that primary CNS lymphoma is a disease of late middle age but some series describe a different age distribution, with the majority of patients being in the seventh, eighth or even in the ninth decades of life. In the present case, the patient was in the third decade of life, which is an atypical age for this disease according to the literature.

A preoperative diagnosis of primary CNS lymphoma can be made with a high degree of certainty if the lesion is hyperdense on CT-scan, is invasive to the surrounding parenchyma, shows homogeneous enhancement, and has broad contact with ependyma and/or leptomeninges. In a large number of cases, Lee et al. evaluated CT-scan findings in some brain lymphomas and have correlated these findings with the tumors' pathological features. These authors have clearly discussed that homogeneous enhancement of primary CNS lymphoma on CT scanning is a common phenomenon, but that peripheral enhancement may be rarely seen, especially in AIDS related lymphomas. They found central necrosis in the high-grade lymphomas with peripheral enhancement; none of those cases were the Burkitt type.

In conclusion, although a threefold increase in the incidence of primary CNS lymphomas has been reported recently, primary meningeal high grade Burkitt-type lymphoma is still a very rare tumor. The peculiarities of this case are the lesion's topography, its radiological characteristic and the fact that the primary meningeal high grade Burkitt-type lymphoma presented as the first clinical manifestation of AIDS.
REFERENCES


