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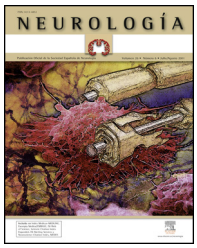

ORIGINAL ARTICLE

Primary central nervous system lymphoma☆

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KEYWORDS
Stereotactic biopsy;
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Methotrexate;
Brain tumour

Abstract
Introduction: Primary central nervous system lymphoma is a rare subtype of extranodal non-Hodgkin lymphoma that accounts for 4% of central nervous system tumours.
Patients and methods: Retrospective review of 24 patients diagnosed with primary central nervous system lymphoma between 1990 and 2010. All patients were diagnosed using magnetic resonance imaging and the diagnosis was confirmed surgically.
Results: Of the 24 patients analysed, all except 4 were immunocompetent. Median age at diagnosis was 59.3 years (range 13—79) and the sex ratio (male to female) was 1:1.1. Cognitive decline (in 33.4%) and headache (in 25%) were the most common complaints. Diagnosis was performed in 13 cases (54%) following craniotomy and in the other 11 cases (46%) after stereotactic biopsy. Breakdown by pathology was as follows: 22 cases of B-cell lymphoma (91.6%), 1 case of anaplastic large-cell lymphoma, and 1 case of T-cell lymphoma. Mean survival time was 12.8 months with an overall 1-year survival rate of 37.5%.
Conclusions: Primary central nervous system lymphoma often presents in the sixth decade with cognitive decline, headache, and focal neurological deficits. A single intracranial lesion was present in 75% of the patients (18 cases), and the remaining 25% (6 cases) had between 2 and 4 lesions. Preoperative clinical status was the most important factor determining prognosis.

PALABRAS CLAVE
Biopsia estereotáctica;
Linfoma no hodgkiniano;

Linfomas primarios del sistema nervioso central

Resumen
Introducción: Los linfomas primarios del sistema nervioso central son una variedad poco frecuente de linfomas no hodgkinianos que constituyen alrededor del 4% de los tumores del sistema nervioso central.

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Linfoma primario del sistema nervioso central;
Metotrexate;
Tumour cerebral

Pacientes y métodos: realizamos una revisión retrospectiva de 24 pacientes diagnosticados de linfoma primario del sistema nervioso central entre enero de 1990 y diciembre de 2010. Todos los pacientes fueron diagnosticados con resonancia magnética y confirmados quirúrgicamente.
Resultados: De los 24 pacientes analizados, 4 presentaban inmunodeficiencia. La media de edad era de 59,3 años (intervalo 13-79) y la relación entre varones y mujeres de 1 a 1,1. El deterioro cognitivo (33,4% de los pacientes) y la cefalea (22,5%) fueron los signos de presentación más frecuentes. El diagnóstico se realizó en 13 casos (54%) tras llevar a cabo una craneotomía y en los otros 11 (46%) mediante biopsia estereotáctica. La distribución histológica mostró que 22 casos (91,6%) eran linfomas tipo B, un caso un linfoma anaplásico de células gigantes y el otro correspondió a un linfoma de células T. La supervivencia media fue de 12,8 meses y a un año del 37,5%.
Conclusiones: Los linfomas cerebrales primarios se presentan alrededor de la sexta década de la vida y clínicamente se manifiestan con deterioro cognitivo, cefalea y déficits neurológicos focales. El 75% de los pacientes (18 casos) presentaban únicamente una lesión intracraneal y el restante 25% (6 pacientes) entre 2 y 4 lesiones. El estado clínico preoperatorio constituye el factor pronóstico más importante.

Introduction

Primary central nervous system lymphoma (PCNSL) is a non-Hodgkin lymphoma originating in the cerebrum, eyes, leptomeninges, or spinal cord with no evidence of systemic lymphoma at the time of diagnosis. These tumours typically originate in B-cells and differentiating them from systemic non-Hodgkin lymphomas is difficult, whether by using a microscope or immunohistochemistry. In contrast, secondary brain lymphomas develop due to extension or spread of a systemic lymphoma in the central nervous system (CNS).[1]

PCNSLs account for about 4% of all primary brain tumours and between 1% and 2% of all lymphomas. The incidence rate of PCNSL has grown slowly in the past decades due to an increase in life expectancy in the general population and the presence of increasing numbers of immunocompromised patients.[2]

The first description,[3] published by Bailey in 1929, referred to these tumours as 'perivascular sarcomas' since the cancer cells tended to surround blood vessels. In 1938, Yuile[4] named them 'reticular cell sarcomas', while Russell and Rubinstein[5] introduced the term 'microgliomas' in 1948. Their current name was coined by Henry et al.,[6] who in 1974 differentiated primary CNS lymphomas from systemic lymphomas. Rappaport[7] subsequently placed them in the group of non-Hodgkin lymphomas in his classification system.

We present a review of 24 patients diagnosed with PCNSL and treated in the last 21 years with a minimum follow-up time of 12 months. The study includes an analysis of patients' clinical and neuroradiological characteristics and aspects related to treatment and progress.

Patients and methods

We carried out a retrospective, descriptive study of patients diagnosed with primary brain lymphoma in our neurosurgery department between 1990 and 2010.

The study analyses patients' demographic and clinical characteristics, neuroradiology diagnostic techniques used, surgical and oncological treatments employed, and patient progress. The Karnofsky Performance Scale (KPS) was used to assess patients clinically. In addition, we completed a systemic study using bone marrow analysis, abdominal ultrasound, and computed tomography (CT) to determine the tumour stage in all patients.

Tumours were confirmed in all cases using stereotaxic biopsy or craniotomy. All patients were monitored a minimum of 12 months.

Results

The series contains 24 patients (13 males and 11 females) with a mean age of 54.7 years (range, 13—79). In patients with AIDS, mean age was 37.7 years, while in all other patients, mean age was 60 (Table 1).

Clinical presentation

The most common form of presentation was cognitive impairment in 8 patients (33.4%), followed by headache in 6 (25%), motor deficit in 5 (20.8%), and convulsions in the remaining 5 patients (20.8%). Four of the patients had AIDS.

According to the KPS, 14 patients scored between 90 and 100 at time of diagnosis, 6 scored between 80 and 70, and 4 patients scored between 60 and 50.

Diagnostic techniques

CT was used as the initial diagnostic method in all cases, and magnetic resonance imaging (MRI) was also used as a complementary method. CT results include 12 cases (50%) with hypodense areas, 8 (33.3%) with hyperdense areas, and 4 (16.7%) that were isodense. Contrast uptake was intense in 16 cases, including 3 with peripheral (ring-shaped) uptake; contrast enhancement was moderate in the remaining 8 cases.