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CASE REPORTS

Meningeal relapse by follicular lymphoma as a single mass

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Abstract Follicular lymphoma is the second most common lymphoma throughout the world. Its course is usually indolent. Affection of Central Nervous System by a follicular lymphoma is usually as primary disease, and secondary affection is usually due to high-grade transformation. In this case-report we describe a young patient who presented a follicular lymphoma which secondary affected the central nervous system without high grade transformation.

Keywords Follicular lymphoma · Central nervous system relapse · Low-grade lymphoma · CHOP-rituximab

In 1993, a 32-year-old white man, a habitual consumer of alcohol and marijuana, was diagnosed with stage IIIA2 follicular lymphoma, affecting the axillary, retroperitoneal, mesenteric, iliac and inguinal nodes. He was initially treated with six cycles of CVP achieving complete remission. One year later he presented with submandibular adenopathy, with cytology positive for lymphoma. A subsequent computerised tomography (CT) scan showed retroperitoneal and coeliac node involvement. He refused further treatment and follow-up was not performed. In March 2007 he presented with headache and altered mental status of two months' duration. Physical examination was normal, except for a firm subcutaneous nodule in the right temporoparietal zone. Laboratory tests, including lactate dehydrogenase (LDH) and beta-2-microglobulin, were normal, and he was negative for HIV-1. CT body scan showed no adenopathies or visceral involvement. A brain CT scan (Fig. 1) detected a 70×35 mm hyperdense right parietal mass with vasogenic oedema, and an extracranial mass with similar radiologic properties. Magnetic resonance imaging (MRI, Fig. 2) showed that the lesions were hypointense in T2 and that the parietal bone was located between the two lesions. A biopsy of the extracranial mass led to a diagnosis of non-Hodgkin's follicular lymphoma, with fewer than 2 centroblasts per high-power field. The tumour was positive for CD20, BCL-2, BCL-6, CD10 and CD23, and negative for CD3, CD30, CD5 and cyclinD1, with a Ki67 proliferative index of 20%. The patient refused local radiotherapy. He was treated with CHOP-rituximab, which resulted in a partial response after 1 cycle, with clinical improvement.

Follicular lymphoma is the second most common lymphoma in the USA and Western Europe, accounting for approximately 30% of all non-Hodgkin's lymphomas [1]. Follicular lymphoma usually affects older adults, in the sixth decade and older, with a slight female predominance. Despite its indolent course, with an overall survival of over 8 years, most patients relapse and eventually die of the disease, although up to 20% of patients can achieve spontaneous remissions [2], which are usually transient.

The central nervous system (CNS) can be affected by lymphoma, usually as primary disease. CNS involvement usually consists of multiple, discrete, deep seated brain masses near the ventricles, but it can also affect the dura as a single mass stemming from a meningioma, or diffusely through the white matter as "lymphomatosis cerebri". Secondary central nervous system lymphoma (SCNSL) is an additional clinical entity, with a rate dependent on the histology of the primary lymphoma [3]. In patients with lymphoblastic or Burkitt's lymphomas, the rate of SCNSL can reach 78% in patients who did not receive CNS prophylaxis, and 19% in patients who received CNS prophylaxis. In-

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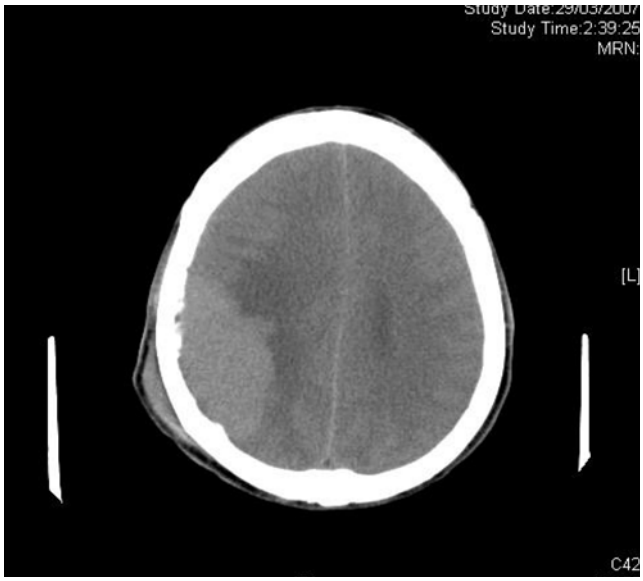

Fig. 1 Brain CT scan. Hyperdense right parietal intra and extracranial mass

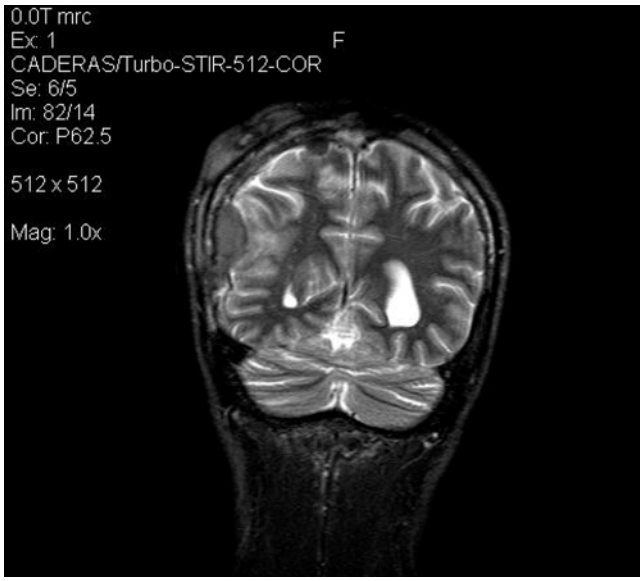

Fig. 2 Brain MRI (coronal turbo short tau inversion recovery (STIR) imaging) showing both lesions

creased LDH and involvement of more than one extranodal site have been shown to be independent predictors of CNS recurrence in patients with high-grade NHLs [4]. Early leptomeningeal disease can be detected by assessing cerebrospinal fluid (CSF), either by cytology or flow cytometry [5]. Low-grade lymphomas usually do not affect the CNS [6]; in the largest clinical series to date, CNS involvement has been observed in fewer than 3% of patients. Follicular lymphoma has been found to account for about 20% of patients with CNS involvement, which may be due to transformation into high-grade NHL [7]. Significant risk factors for low-grade lymphoma CNS relapse are B-symptoms and involvement of the bone marrow or skin [8]. Generally, however, CNS prophylaxis is not recommended and study of CSF is not needed [9].

The case described herein is striking for several reasons. The first is the relatively young age of the patient (32 years). The second is the occurrence of spontaneous remission, lasting 14 years, after an early relapse that occurred one year after first-line chemotherapy. The third is CNS relapse. To our knowledge, a similar case, with local aggressiveness presenting as an intracranial bulky mass breaking through the parietal bone and no other distant sites, has not yet been described in the literature.

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