Clinical Radiology (2008) 63, 97–100

CASE REPORT

A novel radiological appearance of meningeal extranodal marginal-zone B-cell lymphoma

D. Butteriss[a,*], J. Lewis[b], A. Barborie[c], C. Soh[d]

Departments of [a]Neuroradiology, [b]Oncology, and [c]Pathology, Newcastle General Hospital, Westgate Road, Newcastle-upon-Tyne, Tyne and Wear, UK, and [d]Department of Neuroradiology, Hope Hospital, Salford, Manchester, UK

Introduction

Primary meningeal low-grade lymphoma is an uncommon entity that typically presents with symptoms and imaging characteristics similar to those of meningioma. We report a case with a more acute onset and previously undescribed imaging appearances. Confirmatory histology is included.

Case report

A 64-year-old woman presented to the acute stroke service, via her general practitioner, complaining of sudden-onset left arm and leg weakness. She had a medical history of previous myocardial infarction with ongoing angina and hypertension. She had undergone several previous operations, including excision of a facial cutaneous squamous cell carcinoma. She was an ex-smoker of 1 year having previously smoked approximately 30 pack-years.

Neurological examination revealed left arm and leg weakness of 2–3/5, with no other significant findings, and computed tomography (CT) was performed to confirm a clinical diagnosis of right hemisphere infarct. Pre and post-contrast enhanced CT examinations of the head demonstrated patchy periventricular low-attenuation changes consistent with small-vessel disease that were not felt to be responsible for the new neurological symptoms, and also an area of non-enhancing gyral swelling in the right parietal lobe (Fig. 1). In view of the clinical presentation, a diagnosis of subacute infarct was considered, but the appearances were atypical and differential radiological diagnoses of low-grade neoplasm and focal encephalitis were also made. In view of the radiological uncertainty, magnetic resonance imaging (MRI) was performed. This showed an area of apparent gyral swelling confined to the right parietal lobe, without signal change in the cortex or underlying white matter. There was no evidence of haemorrhage or of altered diffusivity and no enhancement after gadolinium administration (Fig. 2). At this stage the radiological appearances were felt to be atypical of an infarct, but the clinical condition of the patient improved and an interval MRI examination was planned. However, during this interval the patient became more unwell with increasing left-sided weakness and headaches. The headaches were occipital, at their worst in the mornings, but not associated with nausea or vomiting. There was no papilloedema on fundoscopic examination. Interval MRI examinations did not reveal any change in the appearances of the right parietal abnormality.

After a sudden deterioration the patient was admitted to the neurosurgical unit bed-bound with severe left hemiparesis, and having had a transient episode of aphasia. Further imaging revealed no change, and a probable diagnosis of low-grade astrocytoma was made. After discussion at the regional neuro-oncology multi-disciplinary meeting, the decision was made not to treat empirically. The neuropathologist requested that the surgeon include some meningeal tissue in the biopsy as it may be helpful in ascertaining a histological diagnosis. Subsequently surgical biopsy of the right parietal lobe and overlying meninges was performed.

* Guarantor and correspondent: D. Butteriss, Department of Neuroradiology, Newcastle General Hospital, Westgate Road, Newcastle-upon-Tyne, Tyne and Wear NE4 6BE, UK. Tel.: +44 191 233 6161; fax: +44 191 219 5037.

E-mail address: dbutteriss@doctors.org.uk (D. Butteriss).

0009-9260/$ - see front matter
doi:10.1016/j.crad.2007.05.019

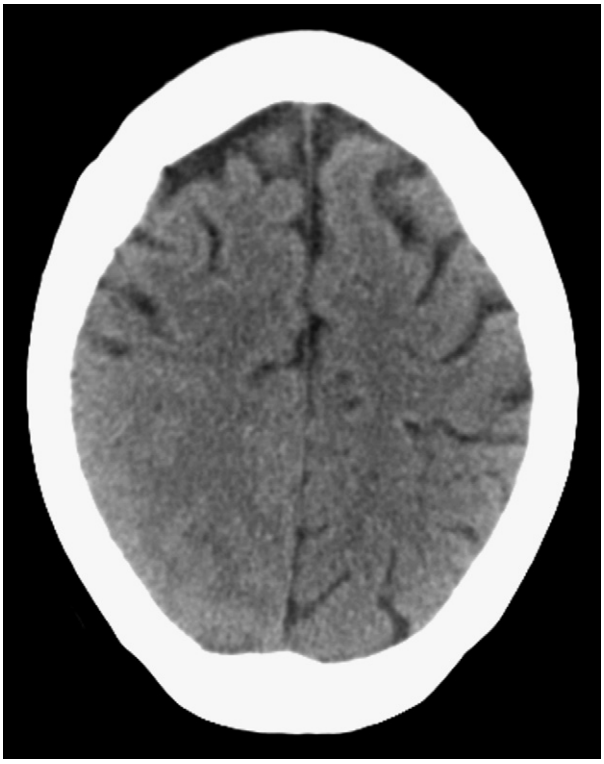

Figure 1 Representative section of non-contrast enhanced axial CT through the rostral cranium showing diffuse swelling of the right parietal lobe with effacement of the sulci, but preservation of the local grey/white matter differentiation.

Pathological examination revealed a dense meningeal infiltrate of small lymphoid cells. A few cells were noted in the Virchow—Robin spaces, but there was no parenchymal involvement. The infiltrate was predominantly mature B cells expressing BCL2, CD20 and CD79a, the Ki67-positive proliferative fraction was below 10%. There was no expression of CD5, CD10, CD23, BCL6, or CD1 (Fig. 3). Scattered T cells were also present. Due to this immunophenotype a diagnosis of extranodal marginal zone B-cell lymphoma was made.

Further investigations to exclude systemic lymphoma, including an MRI examination of the whole spine, CT examinations of the chest, abdomen, and pelvis, and a bone marrow aspirate were all negative.

The patient was referred to the oncology department for treatment. Although it is extremely rare to find low-grade primary non-Hogkin's lymphoma (NHL) in the brain involving the meninges, case reports in the literature suggest that this is a radiosensitive tumour, as one would predict. There is a tendency with cerebral NHL for the disease to diffusely involve the parenchymal tissue, thus she was treated with fractionated whole-brain radiotherapy with a boost to the right parietal region. The whole brain received 28 Gy in 14 fractions, and subsequently, 12 Gy in six fractions to the right parietal region. Oral dexamethasone was administered during the radiotherapy period. There was a rapid symptomatic improvement within 1 week of commencing radiotherapy, and she was discharged home on completion of the course. At this stage the patient was mobile and capable of self-care. There has been no relapse at approximately 10 months.

Discussion

Primary low-grade meningeal lymphoma is an uncommon condition that should be distinguished from both the more common high-grade primary cerebral lymphoma, and also from systemic lymphoma with meningeal involvement. While up to 9% of patients with systemic NHL may demonstrate meningeal deposits, only 1% of primary lymphomas affect the central nervous system (CNS). Most of these are high-grade primary neoplasms, and both of these conditions have a poor prognosis. In contradistinction, however, primary meningeal lymphomas are usually low-grade, respond well to treatment, and have a favourable prognosis.

Fifteen cases of primary meningeal low-grade lymphoma have been described in the literature, 13 were mucosa-associated lymphoid tissue-type lymphomas, and two were follicular lymphomas.[1—8] All affected the dura, except for one intraventricular lesion,[4] and one within the cerebello-pontine angle.[2] Radiologically the majority of these lesions have appeared as focal enhancing masses indistinguishable from meningiomas, and the correct diagnosis has only been made on histological examination after biopsy or excision. The cerebello-pontine angle lesion was radiologically diagnosed as a cranial nerve schwannoma,[2] and another lesion demonstrated biparietal dural thickening and was radiologically identified as bilateral subdural haematomata.[7]

The present case is unique in that there was no radiologically apparent dural abnormality, no pathological enhancement post-gadolinium, and the abnormality appeared to be parenchymal, although there was no parenchymal attenuation change on CT, or signal abnormality on any MRI sequence.

In keeping with the indolent nature of the neoplasm, these conditions characteristically present insidiously and non-specifically with signs and