

Rare disease

A neurological presentation of intravascular B-cell lymphoma

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Intravascular B-cell lymphoma is a rare, aggressive subtype of diffuse large B-cell lymphoma that presents insidiously with symptoms relating to organ involvement. We present the case of a male in his late 40s who presented with fluctuating neurological symptoms including episodes of altered upper-limb sensation, seizures and psychotic phenomena. These symptoms and signs were associated with fleeting brain lesions on neuroimaging. A brain biopsy confirmed the diagnosis of intravascular B-cell lymphoma and he was treated with CHOP chemotherapy (cyclophosphamide, hydroxydaunorubicin, vincristine and prednisolone). Two years later, he remains well. Timely diagnosis and aggressive treatment provides an optimal chance of long-term survival so it is essential to recognise early disease characteristics.

BACKGROUND

Intravascular B-cell lymphoma is a rare subtype of diffuse large B-cell lymphoma characterised by proliferation of atypical B lymphocytes within the lumina of small blood vessels.^{1–3} It presents insidiously with symptoms relating to organ involvement and very occasionally, clinically isolated neurological manifestations. As the neurological differential diagnosis of intravascular B-cell lymphoma includes cryptogenic inflammatory central nervous system (CNS) disease steroids are frequently administered before a biopsy is undertaken, leading to temporary pathological resolution. It is therefore essential to recognise early disease characteristics to enable prioritisation of biopsy and early intervention. We present the case of a patient presenting with fluctuating neurological symptoms in which brain biopsy was vital to diagnosis and effective treatment.

CASE PRESENTATION

A male in his late 40s was diagnosed with probable ‘transient ischaemic attacks’ following three episodes of altered upper-limb and right-sided sensation, one of which was associated with an expressive dysphasia. An initial CT head was unremarkable, but 5 months later he suffered a seizure with prolonged postictal agitation. A further CT head demonstrated two hypodense areas in the right parietal and occipital lobes (figure 1A). He made a good recovery and subsequent neurological examination was unremarkable. However, serum lactate dehydrogenase was raised (477 IU/l) and although a cranial MRI head with gadolinium contrast showed resolution of the previous lesions a new large left frontal lesion had developed (figure 1B: T1 postgadolinium contrast, C: T2 fluid attenuation inversion recovery (FLAIR)). One week later he became aggressive with paranoid delusions and visual and auditory hallucinations. A repeat MRI head with gadolinium contrast showed temporal lobe involvement (figure 1D: T1 postgadolinium contrast, E: T2 FLAIR, F: T1 postgadolinium contrast, G: T2 FLAIR).

INVESTIGATIONS

A right temporal pole biopsy revealed atypical CD20, CD79a and CD5 positive cells within small leptomeningeal and intracerebral vessels, features consistent with intravascular B-cell lymphoma (figure 2). There was no bone marrow infiltration.

TREATMENT

The patient was treated with CHOP regime chemotherapy (cyclophosphamide, hydroxydaunorubicin, vincristine and prednisolone).

OUTCOME AND FOLLOW-UP

Following treatment, his cognitive function improved and his delusions decreased to mild paranoid thoughts. A CT head 4 weeks after treatment showed radiological quiescence of all lesions. Two years later, he remains well.

DISCUSSION

This case report describes the disease course and treatment response of intravascular B-cell lymphoma in a male who presented with fluctuating neurological symptoms associated with fleeting brain lesions.

The largest intravascular B-cell lymphoma case series are from Europe (n = 38) and Asia (n = 25) who report distinct clinical experiences. Median age at diagnosis was 70 and 66 years, respectively, with no clear sex predominance. Time from presentation to death was 25 months in Europe and 32 months when diagnosed antemortem but in contrast was only 7 months in the Asian series.^{4 5} In Europe most (95%) presented with non-specific symptoms including 55% with systemic symptoms such as fever and weight loss. Skin lesions were present in 39% and 34% had some neurological symptoms at presentation.⁴ In contrast, all Asian cases presented with febrile episodes and hepato/splenomegaly, none had skin lesions and only 16% had neurological symptoms.⁵ A retrospective analysis of 109 Asian patients found 25% of cases had CNS involvement at diagnosis and that the presence of

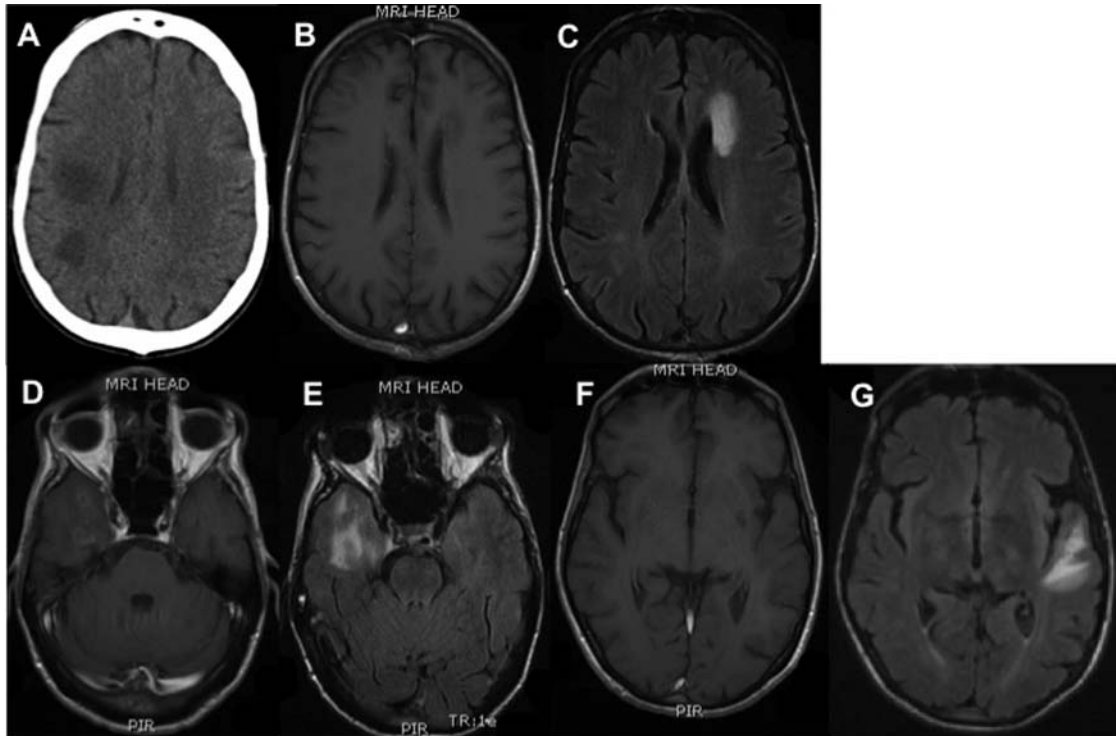


Figure 1 (A) CT head—2 hypodense areas in the right parietal and occipital lobes. March 2010—(B) MRI brain (axial T1 postgadolinium contrast), (C) MRI brain (axial T2 FLAIR) showing a new left frontal lesion. April 2010—(D, F) MRI brain (axial T1 postgadolinium contrast); (E, G) MRI brain (axial T2 FLAIR) showing temporal lobe involvement.

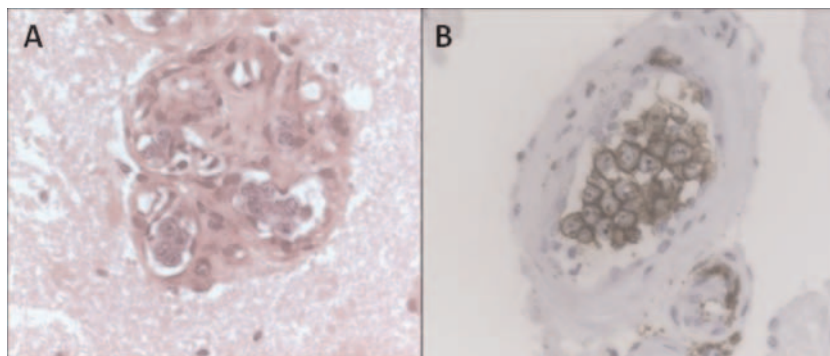


Figure 2 (A) Large, atypical intravascular cells in a cerebral blood vessel showing evidence of recanalisation (H&E). (B) B-cell lineage confirmed by positive staining with the CD20 antibody.

skin lesions was predictive of subsequent CNS recurrence.⁶

Our patient suffered from weight loss but no other additional symptoms documented in the European and Asian case series. His age at onset was younger than commonly

observed in intravascular B-cell lymphoma overall. Early brain biopsy was vital in securing pathological diagnosis and was undoubtedly a crucial factor in his long-term survival.

Competing interests None.

Patient consent Obtained.

Learning points

- ▶ Intravascular B-cell lymphoma presents insidiously with symptoms relating to organ involvement.
- ▶ Neurological presentation of intravascular B-cell lymphoma is rare.
- ▶ Early diagnosis and aggressive treatment provides the optimal chance of long-term survival.

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