‘Killer’ open ring in the brain

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DESCRIPTION
A previously well immunocompetent man in his 60s presented with a 3-month history of subacute altered mental state and seizures. On clinical examination, he was confused but had no focal neurological deficits. Cerebrospinal fluid (CSF) examination showed minimal cells with an elevated protein of 1.1g/dL. Epstein-Barr virus (EBV) PCR was positive in the CSF but other bacterial, viral serology and CSF cytology were negative. An extensive panel of autoimmune and paraneoplastic encephalitis antibodies tested negative. MRI brain showed multiple brain lesions with an incomplete (open-ring) pattern of enhancement (figure 1). These were hyperintense on T2-weighted images with no restricted diffusion. The patient was treated for ‘tumefactive’ inflammatory-demyelinating pathology with corticosteroids and plasma exchange but there was no improvement in his symptoms. He developed haemophagocytic lymphohistiocytosis and further increase in size of the brain lesions. A brain biopsy revealed extranodal natural killer/T-cell lymphoma (NKTL). Blood EBV titres were markedly elevated at up to 310 000IU/mL and EBV-encoded RNA was demonstrated in the brain tissue. Unfortunately, he did not respond to immunotherapy treatment with nivolumab and died about 6 weeks after presentation.

As evidenced in this case, we caution against over-interpreting the specificity of the ‘open-ring’ enhancement pattern, with no restricted diffusion, as a sign of inflammatory aetiology.1 2 The lack of response to immunosuppressive treatment and EBV positivity were unusual for an inflammatory disease process and prompted histological confirmation with a brain biopsy. NKTL is a highly aggressive and rare form of non-Hodgkin’s lymphoma associated with EBV infection.3 It occurs more commonly in the Asian population and typically affects the upper aerodigestive tract such as the nasopharynx and sinuses. Central nervous system involvement is extremely rare and often heralds a poor prognosis.4

Figure 1 (A) Axial T2-weighted images (T2W) with hyperintense lesions in the left parieto-occipital region, splenium of the corpus callosum (arrows), and bilateral frontal white matter. (B) Apparent diffusion coefficient showed no corresponding restricted diffusion; and on (C) CT scan, the lesions were hypodense. (D) Coronal Fluid Attenuated Inversion Recovery (FLAIR) image: there were also hyperintense lesions in the right thalamus, left basal ganglia and left temporal lobe. (E) Axial and (F) sagittal T1W post gadolinium demonstrated incomplete ring enhancement of the left parieto-occipital and splenial (arrow) lesions.

Learning points
► Extranodal natural killer/T-cell lymphoma is a rare haematological malignancy that is associated with Epstein-Barr virus infection and can present with central nervous system manifestations.
► ‘Open-ring’ or incomplete ring enhancement should not be deemed diagnostic of ‘tumefactive’ inflammatory pathology; and due considerations should be given to neoplasms.

REFERENCES