Gliomatosis cerebri mimicking viral encephalitis in a 4-year-old child

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DESCRIPTION

A 4-year-old boy presented with an afebrile right-sided focal seizure. MRI of the brain demonstrated an infiltrating fluid attenuated inversion recovery (FLAIR) hyperintense abnormality in the left frontal, temporal and occipital white matter, involving the temporal lobe cortex, without enhancement, microhaemorrhage or diffusion restriction (figure 1).

The findings were interpreted as infectious encephalitis and intravenous acyclovir was started. Cerebrospinal fluid analysis revealed normal protein and white cell count, and was negative for viral, bacterial and mycobacterial agents.

After initial improvement the focal seizures recurred. Repeat imaging after 28 days showed progression of the FLAIR abnormality with extension into both thalami (figure 2). The lesion was biopsied and histology showed diffusely infiltrating grade III astrocytoma. The

patient was treated with radiotherapy but died 6 months later.

Gliomatosis cerebri (GC) is an infiltrating glial tumour involving at least three cortical lobes.¹ Presentation in children is uncommon: to our knowledge only eight cases in children younger than 5 years have been described.^{1–3} It typically presents with non-specific symptoms and signs including seizures, hemiparesis, ataxia, lethargy and symptoms of intracranial hypertension.^{1 2} GC may mimic more common disorders, such as encephalitis and acute disseminated encephalomyelitis, resulting in delay in diagnosis and treatment.^{4 5} Although prognosis is poor and GC is universally fatal,^{1 2} there is evidence that treatment confers some short-term survival benefit.¹ It is important, therefore, to include GC in the differential diagnosis in cases of widespread infiltrating brain lesions when the clinical features are atypical or unexpected, and to consider brain biopsy in those cases.

Figure 1 Axial fluid attenuated inversion recovery (A and B) and coronal T2-weighted (C) MRI obtained at initial presentation demonstrate an extensive, hyperintense abnormality centred on the white matter of the left temporal lobe (asterisk) and extending into the white matter of the left frontal lobe (white arrow) and left parietal lobe (open white arrow). The cortex of the left mesial temporal lobe (black arrow) and left parasagittal occipital lobe (open black arrow) are also involved. Diffusion-weighted imaging (D) does not demonstrate diffusion restriction in the lesion. Postcontrast T1-weighted MR sequence (E) does not demonstrate enhancement. There is no microhaemorrhage on susceptibility-weighted SWAN sequence (F).

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Figure 2 Axial fluid attenuated inversion recovery (A and B) and coronal T2-weighted (C) MRI performed 4 weeks from initial presentation. There has been significant progression of the lesion, with extension into the left parietal lobe (open white arrow), left occipital lobe (open black arrow), both thalami (white arrow heads) and left insular cortex (black arrow head). White matter biopsy (D and E; H&E stain, original magnification $\times 600$) demonstrates pleomorphic astrocytes and multiple mitotic figures (black arrow). Immunohistochemical staining (F; original magnification $\times 400$) demonstrates expression of glial fibrillar acidic protein. Histopathology is consistent with WHO grade III anaplastic astrocytoma.

Learning points

- Gliomatosis cerebri is an infiltrating glial tumour, which is rare in children.
- Appearance on MRI is typically of a contiguous, fluid attenuated inversion recovery hyperintense abnormality involving at least three cortical lobes with diffuse mass effect.
- It presents with non-specific symptoms and signs and may mimic more common disorders, such as infectious encephalitis and acute disseminated encephalomyelitis.

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Competing interests None.

Patient consent Obtained.

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