Image of the month: Primary central nervous system lymphoma mimicking Bell's palsy

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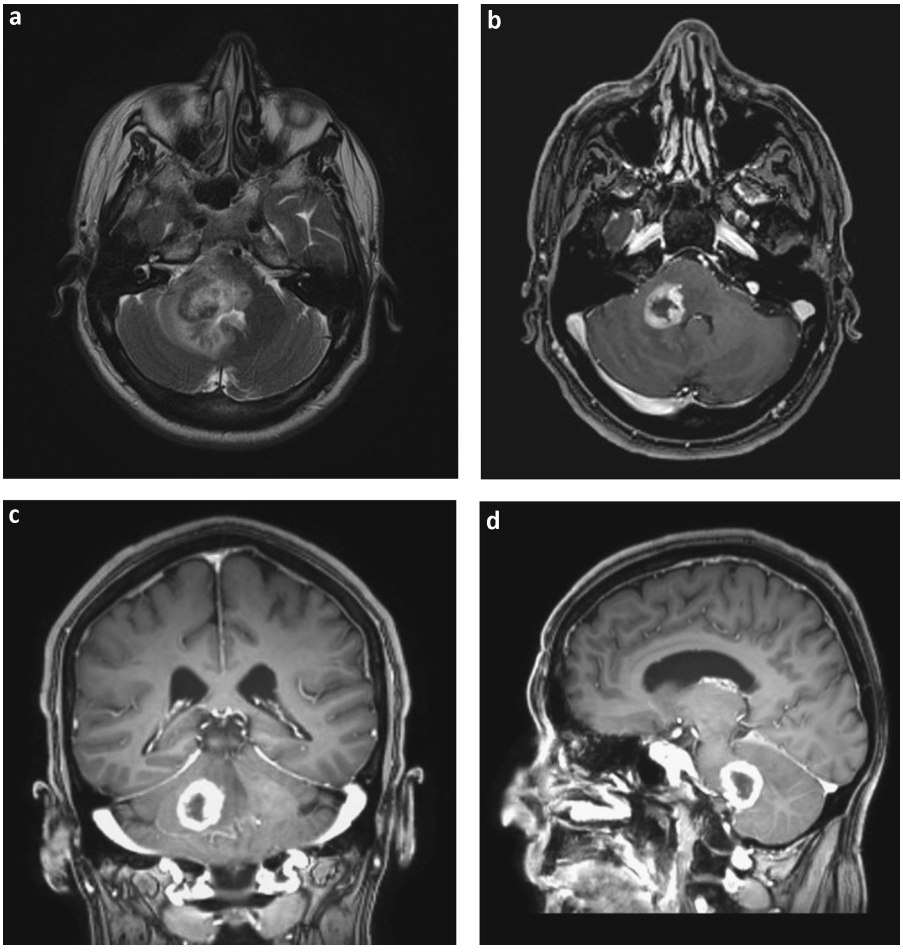

Fig 1. Magnetic resonance imaging of the head. a) Axial plane T2-weighted imaging near the onset of high-dose steroid therapy showing a lesion in the right cerebellar peduncle with oedema extending into the midbrain and pons. b, c, d) Post-contrast T1 stealth imaging the next day showing enhancement of the same lesion in the axial plane (b), coronal plane (c) and sagittal plane (d).

KEYWORDS: neurology, Bell's palsy, primary central nervous system T-cell lymphoma

DOI: 10.7861/clinmed.2022-0263

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Case presentation

A 65-year-old man presented to primary care with a sudden right-sided facial droop, dysarthria and mild right-sided facial numbness. His general practitioner initiated oral prednisolone 50 mg once daily for suspected Bell's palsy. This improved his symptoms temporarily, before they worsened after the course was completed. Thus, unusually, while awaiting a neurologist's assessment, his symptoms were controlled with low-dose prednisolone once daily.

When first seen by the neurologist, he had dysarthria, moderate right-sided lower motor neuron facial palsy, spinothalamic

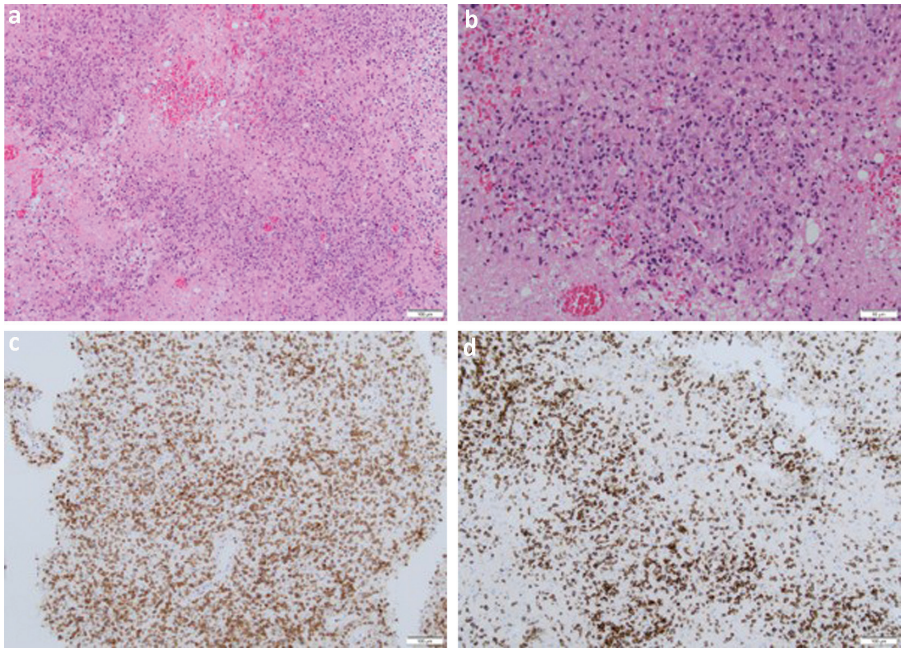

Fig 2. Histopathology slides. a) Haematoxylin and eosin stain (×10 magnification) showing a prominent granulomatous infiltrate within the white matter admixed with small lymphocytes and medium-sized blasts. Areas of necrosis are present, and an occasional mitotic figure is identified. No perivascular accentuation of the infiltrate noted. b) Haematoxylin and eosin stain (×20 magnification) showing similar features to (a). c) CD3 stain (×10 magnification) confirms the presence of numerous small CD3-positive T-lymphocytes throughout the infiltrate. CD5 staining shows a similar pattern. d) CD2 stain (×10 magnification) highlights the CD2-positive blastoid population among the infiltrate.

sensory loss over the right side of the face with preserved light touch and right-sided dysmetria. Examination of the limbs was normal.

Magnetic resonance imaging (MRI) showed a small lesion of the right cerebellar peduncle, for which the regional neuro-oncology multidisciplinary team meeting recommended re-imaging while he was off corticosteroids. Unfortunately, corticosteroid weaning led to a rapid deterioration over days, culminating in dense right-sided hemiparesis, facial weakness and severe ataxia, leaving the patient bedbound. He was, therefore, admitted to our unit where prednisolone 60 mg once daily was initiated, 6 weeks after symptom onset.

Repeat imaging from this time is shown in Fig 1. The lesion, now significantly enlarged and associated with oedema, was notably peripherally enhancing. In view of the differential diagnoses (infection, inflammation or neoplasm) and catastrophic decline, he was referred for biopsy. The initial histological report favoured necrotising vasculitis, while noting the presence of numerous T-cells.

In tandem with steroids, he was treated for vasculitis with pulsed intravenous cyclophosphamide, and gradually improved. Although he had positive anti-neutrophil cytoplasmic antibody (ANCA) serology, enzyme-linked immunosorbent assay (ELISA) for proteinase-3 (PR3) and myeloperoxidase (MPO)-ANCAs were negative (making the ANCA result non-significant). Indeed, a finalised histology report confirmed T-cell lymphoma (Fig 2). Whole body positron emission tomography – computed tomography (PET-CT) and testicular ultrasound ruled out systemic disease, thus confirming primary malignancy.

A four-cycle regimen of alternating MAT (methotrexate, cytarabine and thiotepa) and ICE (ifosfamide, carboplatin and etoposide) was initiated. By the start of this treatment, his symptoms had greatly improved; he was walking with a frame. The disease remained in remission 6 months on.

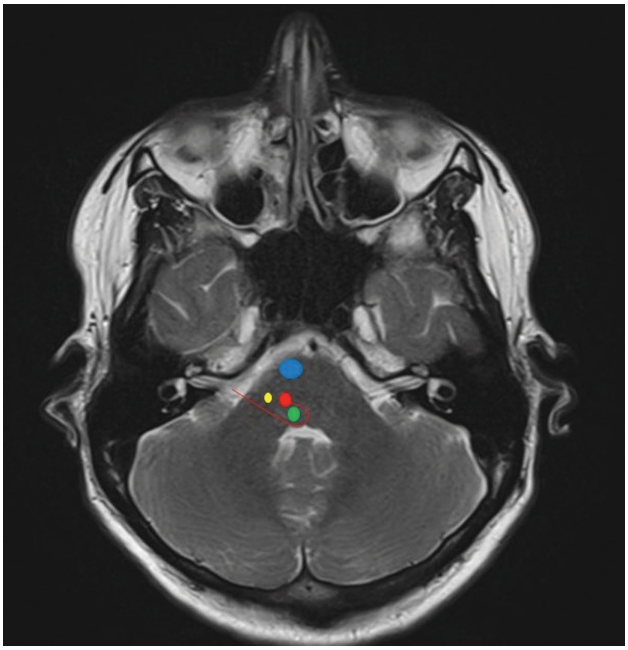

Fig 3. Axial plane T2-weighted magnetic resonance imaging of the head showing normal appearance of the lower pons and cerebellum. The right corticospinal tract (blue), facial nerve and nucleus (red), sixth nerve nucleus (green) and the ascending spinothalamic tract (yellow) are shown.

Discussion

Accounting for 3% of brain tumours, primary central nervous system (CNS) lymphoma is a rare disease, only very few cases of which are T-cell derived (the vast majority are diffuse large B-cell lymphomas).[1] This rarity compounds the interest of our case, whose initial presentation partially mimicked Bell's palsy, but that soon developed into a complex, disabling syndrome, affecting the cerebellum (ataxia), the facial nerve nucleus in the pons (lower motor neuron facial weakness), descending corticospinal fibres in the brainstem (hemiparesis) and ascending spinothalamic fibres (hemisensory loss; Fig 3). Our case emphasises the importance of distinguishing Bell's palsy from other causes of lower motor neuron facial weakness (and illustrates the difficulties of identifying the aetiology of some intracerebral lesions).[2] In retrospect, the dramatic response to steroids favours lymphoma, although biopsy (whose yield was not altered by steroid use, fortunately) was essential for securing the diagnosis. ■

References

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