CASE REPORT

Neuro-Behçet's disease mimicking a cranial tumour

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SUMMARY

Behçet's disease affects many systems and has been described as a multisystem disorder. In Behçet's disease, neurological involvement is responsible for morbidity and mortality, and can sometimes mimic other neurological disorders. We present a case of neuro-Behçet's disease that mimicked a cranial tumour.

incidence of neuro-Behçet's disease (NBD) in BD ranges from 1.3% to 59%.^{3–6} Since NBD can sometimes mimic other neurological disorders, diagnosing BD may be difficult. We present a case of neuro-Behçet's disease mimicking a cranial tumour.

CASE PRESENTATION

A 47-year-old woman was admitted with a diagnosis of hemiplegia and dysarthria. She had no genital ulcers or uveitis, but had recurrent oral ulcers. In addition, her daughter and sister had ulcers in the mouth and genital area. The patient was human leucocyte antigen-B51 homozygous and positive for the b5101 subtype. Lesions with minimal peripheral oedema were detected as

BACKGROUND

Typical Behçet's disease (BD) presents with recurrent oral and genital ulcers and uveitis.¹ Hulusi Behçet first described the disease in 1937. BD can affect many systems and has been described as a multisystem disorder.² Neurological involvement in BD is responsible for morbidity and mortality. The

Figure 1 The (A) coronal T2-weighted images, (B) fluid attenuated inversion recovery imaging and (C,D) contrast-enhanced T1-weighted MRI, showing lesions with peripheral rim-like enhancement and minimal peripheral oedema in the left cerebral peduncle and left temporal lobe.

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hyperintensities at the left cerebral peduncle and left temporal lobe on T2-weighted (T2W) and fluid attenuated inversion recovery imaging (FLAIR) cranial MRI (figure 1A, B). After administering contrast, the lesions showed peripheral rim-like enhancement (figure 1C, D). There was no skin or joint involvement suggestive of sarcoidosis on physical examination. Hypercalcaemia and hypercalciuria were absent. The blood ACE level and chest radiograph were normal. Consequently, sarcoidosis was ruled out.

Routine blood and urine cultures were negative. There were no leucocytes in the cerebrospinal fluid (CSF) and glucose levels were normal. No growth was seen on CSF culture. Herpes simplex virus PCR and anti-HIV were negative. These findings ruled out bacterial infection and viral encephalitis.

There were decreased *N*-acetylaspartate (NAA) levels and increased choline on MRI spectroscopy spectral analysis (figure 2). These findings were considered to indicate a tumour.

Given the clinical history and lesion location, NBD was considered in the differential diagnosis. To avoid invasive procedures, we first decided to treat the BD medically. The corticosteroid treatment resulted in marked clinical improvement and the lesions became smaller and no contrast enhancement was seen on follow-up MRI 4 months later (figure 3A–C).

TREATMENT

The patient was given intravenous pulse corticosteroid therapy with methylprednisolone 1 g/day for 3 days and then oral methylprednisolone 1 mg/kg/day for 2 months, oral azathioprine 50 mg two times per day for 1 year and colchicine 0.5 mg three

times per day for life. The steroid treatment was stopped after decreasing the dose gradually.

OUTCOME AND FOLLOW-UP

After 4 months of medical treatment and rehabilitation, the patient was able to walk unaided and her speech was normal. The lesions were smaller on follow-up MRI.

DISCUSSION

NBD can mimic multiple sclerosis (MS), sarcoidosis, infection and malignancies, especially lymphomas. MRI is commonly used to diagnose NBD. Unfortunately, malignancies, abscesses and MS are sometimes indistinguishable on MRI.⁷

The lesions in NBD may be in the parenchyma or other locations. Non-parenchymal lesions have a better prognosis than parenchymal lesions.^{8–11} Parenchymal lesions are seen as unilateral or bilateral hyperintense areas on T2W MRI and the lesions have a minimal mass effect.¹² The lesions are bilateral in approximately one-third of the cases, and hemispheric lesions are uncommon.⁸ NBD is seen most often at the mesodiencephalic junction and in the pontobulbar area. White matter lesions are seen in nearly 10% of the patients on MRI and they mimic MS lesions.^{13 14} In our patient, the distribution of the lesions and symptoms were not suggestive of MS.

With treatment, prominent regression occurs and only small hyperintensities may remain. Brainstem and spinal cord atrophy may be seen on MRI with disease progression.¹² The treatment of the NBD includes immunosuppressive drugs such as corticosteroids, azathioprine, mycophenolate mofetil, methotrexate,

Figure 2 Spectral analysis showing an elevated choline peak and decreased *N*-acetylaspartate on MRI.

Figure 3 The (A) coronal T2-weighted images, (B) fluid attenuated inversion recovery imaging and (C) contrast-enhanced T1-weighted images showing a marked reduction in the size of the lesions and lack of contrast enhancement.

interferon α and antitumour necrosis factor agents.^{9 15–17} Corticosteroid, azathioprine and colchicine were used in our case, and resulted in marked improvement clinically and radiologically.

When an intracranial mass is seen on MRI, and if there are findings consistent with BD in the clinical evaluation, NBD should be considered in the differential diagnosis of the mass lesion.

Learning points

- ▶ Neuro-Behçet's disease can sometimes mimic other neurological disorders.
- ▶ Neuro-Behçet's disease may respond well to treatment.
- ▶ When an intracranial mass is seen on MRI and there are clinical findings consistent with Behçet's disease, neuro-Behçet's disease should be considered in the differential diagnosis of the mass lesion.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Reminder of important clinical lesson

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