CASE REPORT

Tumefactive demyelination: an unusual cause of a spontaneously resolving homonymous hemianopia

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SUMMARY

A 21-year-old man presented to eye casualty complaining of altered vision associated with headache and vomiting upon waking. Clinical examination was unremarkable except for a right-sided homonymous hemianopia. The MRI scan of the brain revealed a space occupying lesion within the occipital lobe and MR spectroscopy highlighted this to be inflammatory in nature, most likely a tumefactive demyelinating lesion (TDL). Lumbar puncture displayed positive oligoclonal bands. The patient was managed conservatively and made a full recovery, with normal visual fields recorded after a 3 month follow-up. This is a case of a TDL manifesting itself as an unusual cause of homonymous hemianopia; misdiagnosis of TDL is common and potentially damaging to the patient.

BACKGROUND

A tumefactive demyelinating lesion (TDL) is defined as a solitary demyelinating lesion greater than 2 cm.¹ TDLs pose a diagnostic challenge and are frequently misinterpreted on both clinical and radiological grounds, as intracranial neoplasms. It is important to be aware of the differential diagnosis of tumefactive demyelination, in order to direct appropriate investigation, consider steroid treatment and avoid unwarranted surgical intervention.

CASE PRESENTATION

A 21-year-old man, with no medical history woke up with symptoms of headache, vomiting and altered vision. Ophthalmic examination was unremarkable except for visual field testing, which confirmed a right-sided homonymous hemianopia (figure 1A,B). Visual acuity of 6/5 OD+6/6 OS unaided and normal colour vision were recorded. The rest of the neurological examination was normal.

INVESTIGATIONS

Routine blood tests were normal. Inflammatory markers (erythrocyte sedimentation rate/C reactive protein (ESR/CRP)) and an autoimmune screen including autoantibodies, extractable nuclear antigen, double-stranded DNA, anti-neutrophil cytoplasmic antibody, as well as serum ACE, were also normal. MRI scans of the brain revealed a 35×20 mm space occupying lesion adjacent to the occipital horn of the left lateral ventricle within the occipital lobe (figures 2A,B and 3A,B). MR spectroscopy showed prominent lipid/lactate peaks with a reduction in the N-acetylaspartate (NAA) ratio and slight increase in the choline ratio, indicating an inflammatory demyelinating lesion (figure 4). Visually evoked potentials were normal but lumbar puncture displayed positive oligoclonal bands.

Figure 1 Full Field 120 Screening Test of patient's (A) left and (B) right visual field showing a right-sided homonymous hemianopia on the day of presentation.

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Figure 2 (A) Axial and (B) sagittal T1-weighted post-IV gadolinium MRI demonstrating the left occipital lobe lesion with a lack of contrast enhancement and lack of adjacent mass effect.

DIFFERENTIAL DIAGNOSIS

The initial differential diagnosis to consider would be an intracranial neoplasm. Another differential diagnosis would be an intracranial abscess. However, our patient did not display any signs of sepsis and there was no background history of immunosuppression. Normal inflammatory markers and normal autoimmune screen reduce the likelihood of these symptoms being secondary to a systemic inflammatory disorder. Finally, the acute onset of visual symptoms could point towards a vascular cause. However, taking into account the age of the patient and the radiological findings and positive oligoclonal bands on lumbar puncture, one would consider demyelinating disease as a more likely aetiology.

TREATMENT

The patient was managed conservatively with observation and no treatment.

OUTCOME AND FOLLOW-UP

Repeat MRI scans of the brain showed a reduction in the size of the lesion and repeat visual field tests 3 months later indicated that the right-sided homonymous hemianopia had completely resolved (figure 5).

DISCUSSION

Clinical presentation of TDLs is often determined by the size and location of the lesion.² Signs and symptoms can therefore vary, ranging from motor symptoms, headaches and confusion, to seizures and optic neuritis.³ This wide clinical spectrum means TDLs pose a diagnostic challenge to the clinician with differential diagnoses such as intracranial neoplasm, abscesses, inflammatory disorders and demyelinating disease need to be considered.⁴ Treatment can also differ greatly for these conditions. Incorrect diagnosis of TDLs based on MR scans and histopathological tests may lead to potentially harmful surgical intervention or radiation therapy, since the biopsy results from

Figure 3 (A) Unenhanced axial T2-weighted MRI demonstrating the left occipital lobe lesion with minimal surrounding oedema. (B) Coronal fluid-attenuated inversion recovery MR sequence demonstrating the left occipital lobe lesion. Dwivedi RA, et al. *BMJ Case Rep* 2013. doi:10.1136/bcr-2013-009363

Figure 4 MR spectroscopy trace displaying prominent lipid/lactate peaks with a reduction in the N-acetylaspartate ratio and an increase in the choline ratio, indicating an inflammatory demyelinating lesion.

these lesions can display features of glial neoplasm such as hypercellularity and atypical reactive astrocytes.⁵ With MR scans, TDLs and gliomas are difficult to distinguish as both can show variable contrast enhancement, non-specific ill-defined borders, extensive perilesional oedema, mass effect and central necrosis.⁶ Proton MR spectroscopy is therefore a useful tool in helping to differentiate between TDLs and other intracranial disease processes.⁵ MR spectroscopy identifies changes with active inflammatory/demyelinating processes and axonal injury.⁷ These include changes in the resonance levels of choline, which indicates myelin breakdown in the active demyelination process, changes with lactate that reflect inflammation, as well as changes with lipids and NAA that point towards cell membrane breakdown and axonal injury.⁸ The study by Butteriss *et al*⁸ states that acute multiple sclerosis plaques exhibit a pattern on MR spectroscopy of an increase in chemical resonance levels of lipids, lactate and choline combined with a reduction in the levels of NAA. The MR spectroscopy results for our patient were in keeping with this as they showed prominent lipid/lactate peaks with a reduction in the NAA ratio and a slight increase in the choline ratio, hence indicating an inflammatory demyelinating lesion (figure 4). With regard to treatment, high-dose corticosteroid therapy has been shown to reduce the size of TDLs and improve the associated clinical symptoms.⁹ In our case, treatment with steroids was deferred until a more definitive diagnosis could be reached. Over the next 4 weeks, while diagnostic tests were being conducted, the patient was monitored

symptomatically. His symptoms had improved subjectively during this interval and he continued to be observed without treatment until his visual field loss recovered. Our case is unique in that the clinical symptoms resolved completely without any steroid therapy or other forms of pharmacological treatment and such a case has not yet been published.

Learning points

- ▶ Tumefactive demyelinating lesion (TDLs) pose a diagnostic challenge to the clinician as they can mimic sinister intracranial pathology.
- ▶ Correctly identifying a TDL is important for averting misdiagnosis of an intracranial neoplasm and alerting the clinician that further demyelinating episodes may result in diagnosis of multiple sclerosis.
- ▶ Homonymous hemianopia can be an unusual clinical manifestation of a TDL.
- ▶ The diagnostic ability of MR spectroscopy is useful in distinguishing between TDLs and other intracranial pathology such as glioma.
- ▶ Although steroid therapy can be used to treat TDLs, the spontaneous recovery of our patient's visual field and a reduction in the size of the TDL was possible without any treatment.

Figure 5 Binocular Esterman visual field test showing complete recovery of the right-sided homonymous hemianopia three months later.

Contributors All four authors have contributed towards the conception and design of this case report, the acquisition of data and their interpretation, and in the drafting and writing of this case report.

Competing interests None.

Patient consent Obtained.

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