CASE REPORT

MRI findings in chronic relapsing inflammatory optic neuropathy

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SUMMARY

Chronic relapsing inflammatory optic neuropathy is a steroid responsive recurrent optic neuropathy. We describe a patient who presented with recurrent episodes of painful visual loss over a period of 19 years and atypical findings on cranial MRI.

BACKGROUND

Subacute loss of vision accompanied by pain is mostly due to some form of inflammatory optic neuropathy, commonly due to demyelinating optic neuritis. At the onset, optic neuritis is usually associated with periorbital pain elicited by eye movement which worsens in synchrony with or preceding a reduction in vision. The pain tends to diminish as the visual symptoms become prominent. Chronic relapsing inflammatory optic neuropathy (CRION) is an exquisitely steroid responsive isolated recurrent optic neuropathy, associated with normal MRI studies of brain in majority.¹ Despite being an important differential to the other demyelinating illnesses (like multiple sclerosis and neuromyelitis optica) the exact pathogenesis and management options in CRION remains to be speculative, on account of its rarity.

CASE PRESENTATION

A 29-year-old female presented to our outpatient clinic with painful diminution of vision over a period of 10 days involving the left eye. Pain persisted beyond the onset of visual loss. Patient gave a background history of 11 similar attacks involving either of the eyes, but never simultaneous or sequential bilateral involvement over the past 16 years. Each episode began with local pain increasing on eye movement and visual loss over the next 7–10 days. The visual acuity during the episodes varied between

perception of light to finger counting at 1 m. The interval between episodes varied from 2 months to 3 years. Each episode remitted following administration of systemic steroids with complete recovery of vision. There was no history suggestive of any other cranial nerve, motor, sensory or autonomic system involvement. No relevant symptoms suggestive of connective tissue disease or autoimmune illness were noted historically. Examination revealed relative afferent pupillary defect in the left eye. There was reduced visual acuity in the left eye with finger counting at 1 m, with 6/6 visual acuity in the right eye on Snellen's chart. Fundoscopy revealed bilateral normal discs and retina. Rest of the neurological examination was unremarkable.

INVESTIGATIONS

On investigations, haemogram, serum biochemistry, chest x-ray, contrast-enhanced CT of the chest and abdomen were normal. Mantoux test, collagen workup (antinuclear antibody, antiphospholipid antibody, and rheumatoid factor) were negative. Serum ACE level was normal. Hepatitis B antigen, HIV serology were non-reactive. Cerebrospinal fluid examination was normal with no oligoclonal bands. Serum IgG neuromyelitis optica (NMO) antibody was negative. Visual-evoked response revealed prolonged P100 latency (in both eyes). Brain stem auditory response was normal. Optical CT demonstrated normal retinal nerve fibre layer thickness. Gadolinium (Gad)-enhanced cranial MRI done in 2010 showed asymmetrical enhancement of the left optic nerve (figure 1A) and non-enhancing hyperintensities in the right frontal lobe, bilateral medial temporal and occipital lobes and periventricular white matter on T2-weighted and fluid attenuated inversion recovery (FLAIR) images (figure 1B,C).

Figure 1 Gad-enhanced MRI brain showing (A) asymmetrical left optic nerve enhancement and (B) T2 fluid attenuated inversion recovery hyperintensities in the right frontal lobe and the periventricular region as well as (C) bilateral medial temporal and occipital lobes.

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Figure 2 Follow-up Gad-enhanced MRI brain done after 1 year showing (A) asymmetrical left optic nerve enhancement, (B) T2 fluid attenuated inversion recovery hyperintensities in bilateral (right>left) fronto-parietal and temporo-parietal regions and (C) patchy enhancement of left frontal and right medial temporal lesions.

Follow-up Gad-enhanced cranial MRI done in 2011 also revealed asymmetric enhancement of the left optic nerve (figure 2A), but with fresh hyperintense lesions in bilateral fronto-parietal and parieto-temporal regions in the cortical and subcortical location on T2/FLAIR images (figure 2B), with patchy enhancement in the left frontal and right medial temporal region (figure 2C).

DIFFERENTIAL DIAGNOSIS

In view of the recurrent steroid responsive attacks of optic neuritis, a differential diagnosis of other demyelinating disorders like multiple sclerosis, NMO spectrum disorder as well as other causes of secondary demyelination was duly considered. However, all the ancillary investigations were negative for the suspected aetiologies and even the gadolinium-enhanced cranial MRI showed atypical findings even on follow-up studies.

TREATMENT

For the present symptoms, patient was administered intravenous methylprednisolone 1 g/day for 5 days, followed by tapering doses of oral steroids over the next 1 month. Patient is currently maintained on oral azathioprine 100 mg/day with no further relapses.

OUTCOME AND FOLLOW-UP

Pain subsided over next few days and her vision improved to 6/6 over 2 weeks. As a steroid sparing agent, the patient was initiated on oral azathioprine (100 mg/day) following which she had no further relapses even after 1 year of follow-up.

DISCUSSION

Our case has features consistent with the entity of CRION with atypical findings on cranial MRI. Over the past 10 years, this is the second case of CRION from our registry (110 cases) of optic neuritis, the first one having normal neuroimaging.² Previously reported MRI abnormalities have been thickening, high signal intensity or enhancement involving the optic nerves in about 63% of patients.¹ CRION-like presentation has been associated with lupus, sarcoidosis and other systemic

autoimmune diseases.³ However, brain parenchymal MRI changes in CRION are unusual. Brain MRI changes previously described in a single patient out of a cohort of 15 patients were T2/FLAIR hyperintensities in the posterior parietal lobe.¹ Our patient had clinical features consistent with CRION without evidence of systemic illness. Cranial MRI changes in our patient included cortical and subcortical hyperintensities reminiscent of demyelination. The significance of these findings is subject to circumspection and may suggest CRION to be a part of spectrum of primary demyelination.

Learning points

- Chronic relapsing inflammatory optic neuropathy (CRION) as an entity should be diagnosed promptly as it may masquerade the other common pathologies involving the optic nerves like optic neuritis associated with multiple sclerosis or neuromyelitis optica.
- CRION may be associated with changes in cranial MRI simulating demyelinating disorders as is being illustrated in our case.
- The role of immunomodulators in prevention of relapses in CRION is still speculative as its exact aetiopathogenesis is unclear.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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