CASE REPORT

Acute onset blindness: a case of optic neuritis and review of childhood optic neuritis

Sithara Ramdas,[1] Danny Morrison,[2] Michael Absoud,[1] Ming Lim[1]

[1]Department of Paediatric Neurology, Evelina London Children's Hospital at Guy's and St Thomas' NHS Trust, London, UK
[2]Children's Eye Department, Guy's and St Thomas' NHS Trust, London, UK

Correspondence to
Dr Sithara Ramdas,
sithara.ramdas@nhs.net

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SUMMARY

Optic neuritis (ON) is an acquired disorder of the optic nerve due to inflammation, demyelination or degeneration. We report a child who presented with acute onset bilateral visual loss who, following a diagnosis of ON, was treated and had excellent visual recovery. Paediatric ON is considered to be different clinical entity to adult ON. Although in children ON is usually parainfectious or postinfectious, it can be the first presenting feature of multiple sclerosis or neuromyelitis optica spectrum disease. In this paper, we discuss the literature on treatment of ON and prediction of risk of recurrence.

BACKGROUND

Optic neuritis (ON) is an important cause of unilateral or bilateral acute onset visual loss in children. It can occur in isolation or in association with systemic autoimmune conditions. ON has an incidence of two per million children per year.[1] Although ON in children is usually parainfectious or postinfectious, it can be the first presenting feature of multiple sclerosis (MS) or neuromyelitis optica spectrum disease (NMOSD). Unlike adults there is no established treatment consensus in children though treatment with corticosteroids is recommended.[2]

CASE PRESENTATION

A 7-year-old girl presented with acute severe deterioration of her vision over the course of 3 days. She was born in the UK to Egyptian parents and prior to this, she had no visual symptoms. She had noticed slight blurring of her vision 3 days previously but had not mentioned this to her parents. On the day before presentation, she told her dad that she could not watch TV properly 'as everything was blurry'. On the day of her presentation, her teacher called the parents from school as the girl could not read or write and was unable to see the lines in her exercise book. Parents had brought samples of her writing from that week (figure 1),

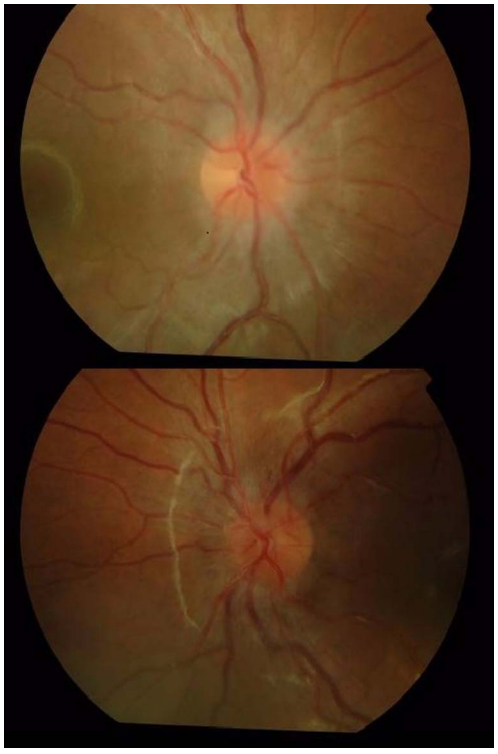

Figure 2 Right and left retinal photographs of posterior pole of the eye, 2 days after presentation. The optic disc is central in the image. There is mild disc swelling and loss of the clear disc margins with softening of the edges.

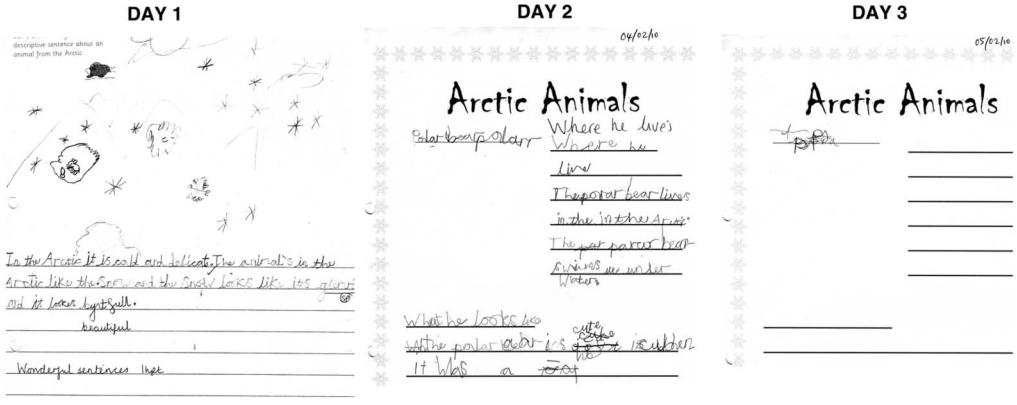

Figure 1 Sample of patient's handwriting showing deterioration over 3 days.

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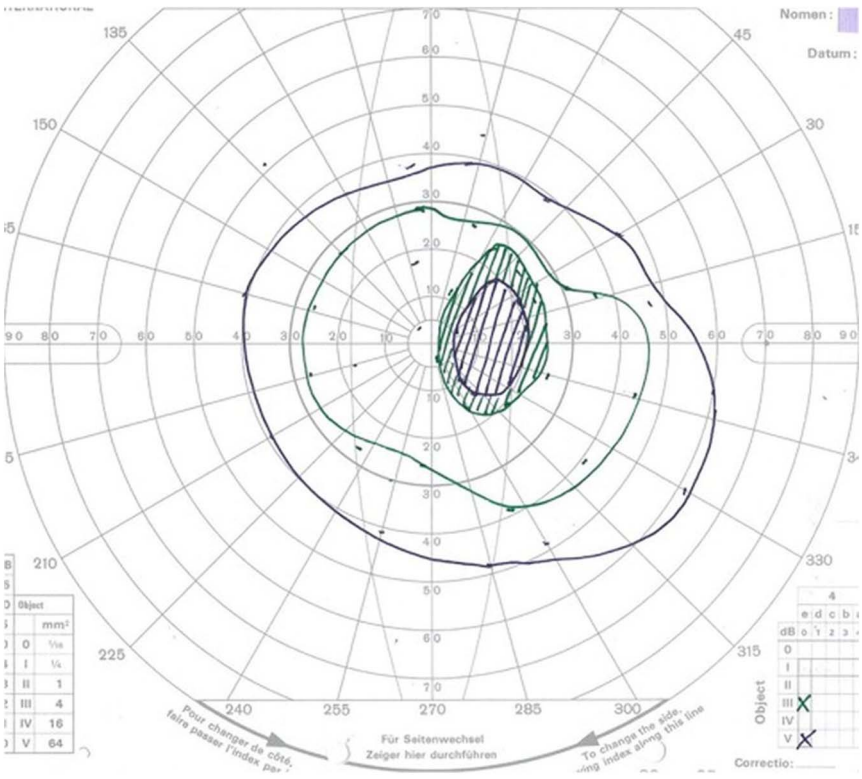
A



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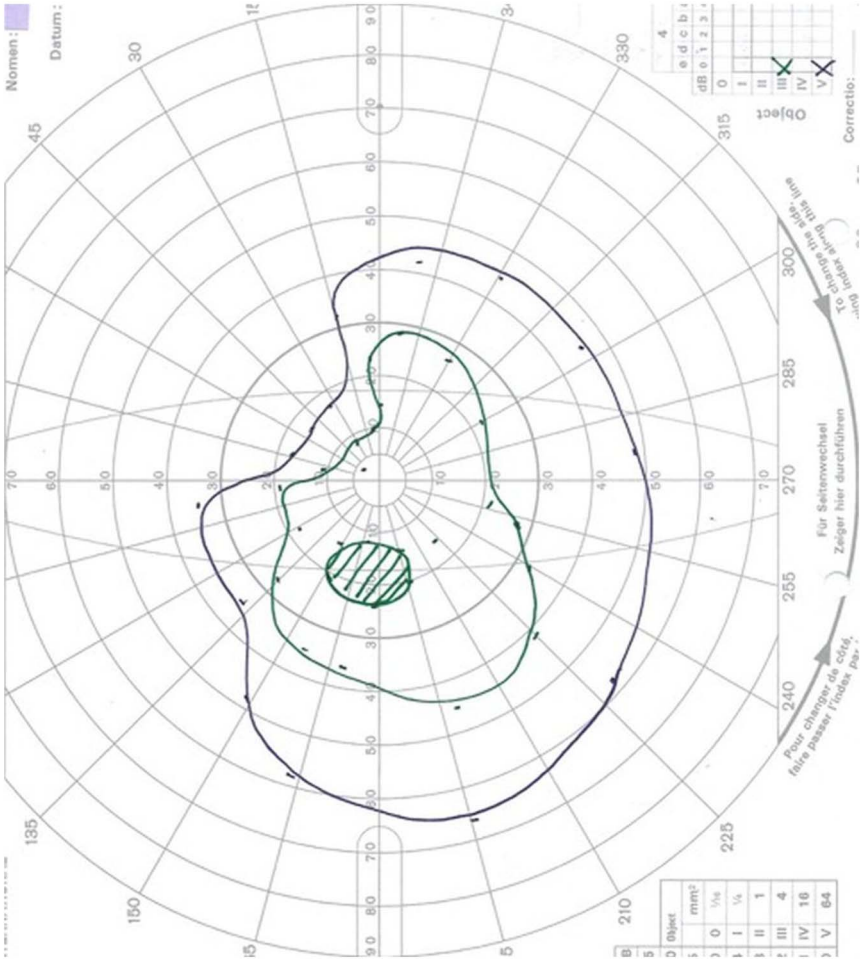

Figure 3 (A and B) Goldmann visual fields in the acute phase showing enlarged blind spot plus constriction in both eyes. The green isopter is III4e, and the purple is V4e.

showing progressive deterioration of her handwriting over the course of 3 days. There was no history of preceding illness. There was no family history of autoimmunity and there was no relevant social history.

On examination, right visual acuity was perception of light only, and on the left counting fingers at 20 cm with minimally preserved colour vision. Acuity on formal testing was 1/60 bilaterally (log MAR 1.0). Conjugate eye movements were full range and pain free.

No conjunctival congestion or lid swelling was noted. Funduscopy demonstrated bilateral optic disc swelling (figure 2). A relative afferent pupillary defect (RAPD) was noted in the right eye. Colour vision (Ishihara) at presentation was right 0/17 and left 1/17 (ie, test plate only). At presentation, near visual acuity was right: not possible, left N48.

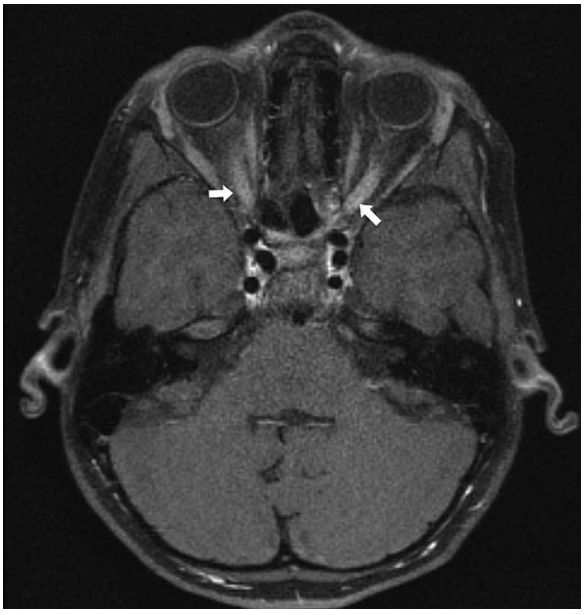

Figure 4 MRI brain with gadolinium showing bilateral postcontrast enhancement of optic nerves.

Goldmann visual fields on day 2 of admission shows enlarged blind spot, plus constriction in both eyes (figure 3A, B).

Rest of the neurological examination was normal. Coordination and gait could not be adequately tested due to her marked visual impairment, but no significant cerebellar signs were observed.

INVESTIGATIONS

Blood inflammatory markers were normal. Blood analysis was also negative for viruses, mycoplasma and Lyme serology. Autoantibodies and aquaporin antibodies were negative. Cerebrospinal fluid (CSF) analysis showed normal cell count and protein, negative microscopy and virology. Oligoclonal bands were negative in the CSF.

A gadolinium-enhanced, fat suppressed MRI (figure 4) showed bilaterally abnormal optic nerve enhancement in keeping with acute bilateral ON.

DIFFERENTIAL DIAGNOSIS

A number of conditions can present with acute onset visual loss and these must be considered in the differential diagnosis. Table 1 lists the differential diagnosis and appropriate investigations to be considered.

TREATMENT

She was treated with a standard regime of intravenous methylprednisolone, followed by oral prednisolone that was weaned over 4 weeks.

OUTCOME AND FOLLOW-UP

Her visual acuity after 10 days was 6/60 bilaterally, and it gradually improved with time. At follow-up, near visual acuity was right N5, left N5 (ie, normal). Colour vision (Ishihara) at follow-up (5 months later) was right 13/13, left 13/13. She had normal Goldman visual fields at 18 months follow-up in both eyes (figure 5A, B).

Her neurological examination remained normal.

In summary, our patient had idiopathic ON from which she had made a full recovery with no relapses at 5-year follow-up.

Table 1 Differential diagnosis of ON in children (adapted from Shams and Plant[3])

		Diagnostic aids
Corticosteroid-responsive optic neuropathy	Sarcoidosis, systemic lupus erythematosus, Behçet's syndrome NMOSD	Serum ACE ANA, dsDNA, lupus anticoagulant, anticardiolipin antibody Aquaporin 4 antibody
Other inflammatory conditions	Postinfection, postvaccination, neuroretinitis, acute disseminated encephalomyelitis	History Blood/CSF analysis MRI brain
Compressive optic neuropathies	Primary optic nerve tumours, gliomas, meningioma, craniopharyngioma, arterial aneurysms	MRI brain and orbits MRA/angiogram
Infections	Tuberculosis, Lyme disease, toxocariasis or helminthitis (usually visible retinal/optic head lesion)	Serology
Infiltrative	Leukaemia Lymphoma	MRI brain Blood/CSF analysis
Nutritional	Vitamin B12 deficiency	Vitamin B12 level
Toxic	Ethambutol toxicity	History
Genetic	Leber hereditary optic neuropathy Biotinidase deficiency	DNA analysis Serum biotinidase
Factitious visual loss		Diagnosis of exclusion

NMOSD, neuromyelitis optica spectrum disease; ON, optic neuritis.

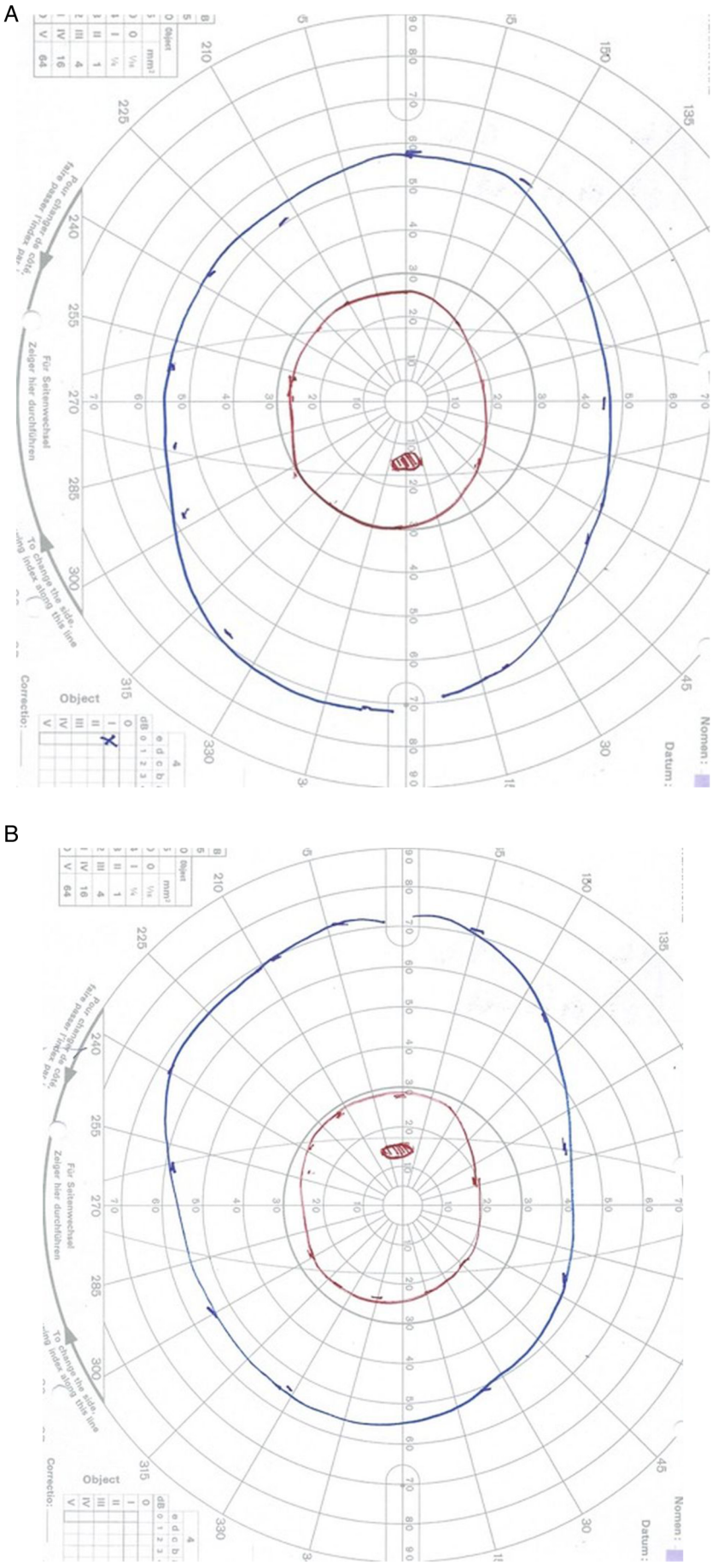

Figure 5 (A and B) Normal goldmann visual fields in both eyes at 18 months follow-up. The blue isopter is I4e, and the red is I2e. The red is the smaller of the two targets.

DISCUSSION

Clinical presentation

The common presentation is acute or subacute onset bilateral or unilateral visual loss. The diagnostic criteria for ON are acute or subacute loss of vision and ≥1 of: RAPD (unilateral cases), visual field deficit or scotoma, impaired colour vision, optic disc oedema or abnormal visual evoked potentials. MRI is not necessary for diagnosis. Visual acuity at presentation is usually severely affected, 6/60 or worse in 70–100%.[4–7] In addition to decreased visual acuity, children can have orbital pain/painful eye movements, decreased colour vision and contrast sensitivity, photopsia and a spectrum of visual field defects.

Average age of presentation is 8.6–11.8 years.[4 8–13] Bilateral involvement may occur simultaneously or sequentially and is seen in 40–70% of cases.[4 10–14] Younger children (<10 years of age) are more likely to have bilateral ON.[14] A history of preceding upper respiratory tract infection is reported in 30–70% of cases[7 15–17] and there may also have preceding headache/supraorbital pain or painful eye movements.[10 18]

Investigation

MRI brain and orbit with contrast should be performed in all cases not only to diagnose ON but also to exclude any other intracranial pathology and look for any other signs of demyelination. Blood and CSF analysis must be performed to look for infection and inflammation. Visual evoked potential can be useful in very young children. Optical coherence tomography can provide specific diagnostic clues to differentiate MS–ON and NMOSD–ON as discussed below.[19 20]

Treatment

Unlike the ONTT (Optic Neuritis Treatment Trial) study in adults, there have been no clinical trials in children looking into treatment of ON. Current clinical practice vary from a wait and watch approach for unilateral and mild bilateral cases to treatment with corticosteroids in severe ON.[8 9 21] The regime commonly described in literature is a course of intravenous methylpredisolone 30 mg/kg/day for 3–5 days followed by oral prednisolone at 1 mg/kg/day with slow wean over 4–6 weeks. It is important to recognise that the response to treatment can vary due to underlying aetiology and these must be investigated appropriately.

Prognosis

Children with idiopathic ON, even those with severe visual loss, have very good recovery of visual function. The visual recovery spontaneously begins at 2–3 weeks and can continue up to 2 years. Visual recovery to 6/12 or better is seen in 70–85%.[4 6 22 23] A single series[5] reported that patients with unilateral disease had an excellent prognosis (100% had better than 6/12) compared to those with bilateral disease (only 50% ≥6/12 and 35% >6/60). Colour vision defects commonly persist.[4] Younger children (<6 years) have better visual prognosis than older children.[22] After an episode of ON, mild pallor may persist in the temporal optic disc along with RAPD and perception of Uhtoffs phenomenon.[7]

Long-term risk prediction

In children with first episode of ON, it is important to be able to distinguish whether an episode of ON is due to MS or NMOSD as the therapeutic options differ and in fact interferons can worsen NMOSD.[24–27]

MS versus NMOSD

The two consistent risk factors for MS following an episode of ON include positive MRI brain lesions at presentation of ON[7 15] and presence of CSF oligoclonal bands.[18 28 29] It remains unclear if the age of onset of ON influences the future risk of MS.[8 14]

Risk for NMOSD is significant if the child has positive Aquaporin 4 IgG antibodies at presentation of ON.[24 30] Optic nerve involvement in NMOSD in comparison to MS is more likely to be recurrent, often simultaneously bilateral, involves the chiasm and cause altitudinal visual field defects with severe residual visual loss.[31–33]

A review by Bennett *et al* has shown that the retinal nerve fibre layer (RNFL) is significantly reduced in patients with NMOSD with ON compared to healthy controls and that RNFL thinning may be an early phenomenon.[34] NMO–ON affects the entire peripapillary RNFL with particular involvement of the superior and inferior quadrant in comparison to MS–ON where temporal quadrant is more affected.[34] Macular thinning is more severe in NMO–ON than in MS–ON, in keeping with the poorer visual recovery observed in ON with NMOSD. Patients with NMOSD are also more likely to develop microcystic macular oedema by ocular coherence tomography in affected eyes.[34 35]

More recently, MOG antibodies have also been identified in children with ON, recurrent ON and NMOSD syndrome who are Aquaporin 4 antibody negative.[36 37] Although the precise predictive value of these antibodies remains to be determined, these children appear to have a non-MS course of demyelination.[37]

Learning points

- Optic neuritis is rare in children but early recognition is important for diagnosis and prompt treatment.
- Most children have very good recovery of visual function.
- Optic neuritis could be the first presentation of serious underlying disorder like multiple sclerosis or neuromyelitis optica spectrum disease; so, detailed investigations of the first presentation and appropriate follow-up are important.

Contributors SR and ML were responsible for conception of the case report. SR drafted the initial manuscript. DM, MA and ML revised the manuscript critically.

Competing interests None declared.

Patient consent Obtained.

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