Acute disseminated encephalomyelitis presenting as optic neuritis in a case of idiopathic thrombocytopaenic purpura

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DESCRIPTION

Acute disseminated encephalomyelitis (ADEM) is an uncommon monophasic inflammatory demyelinating disease that usually presents in children and young adults after viral infection. It is thought to be an autoimmune disorder of the central nervous system (CNS). Antiviral antibodies or a cellmediated response to the pathogen cross-react with the myelin autoantigens. Patients usually present with convulsions, paresis, altered sensorium and other signs of CNS involvement. Isolated optic neuritis as the only presenting feature of ADEM is a very rare and reported only in 2% of the cases. ²

We present a child with isolated visual loss related to optic neuritis, occurring after an episode of viral illness in a diagnosed case of idiopathic thrombocytopaenic purpura (ITP).

A 7-year-old boy presented with sudden onset diminution of vision in the right eye with pain on ocular movement of 1 week duration. There were no H/O convulsions, weakness, fever and headache.

The patient was a known case of ITP and had a history of viral fever 4 months ago. Visual acuity right eye (RE)—perception of light, accurate projection of rays. Left eye (LE)—6/6. Anterior segment examination RE showed a relative afferent pupillary defect. Fundus examination of the RE revealed disc hyperaemia and oedema, few peripapillary flame-shaped haemorrages which was suggestive of optic neuritis (figure 1). Fundus (LE) was normal (figure 2). Blood picture showed lymphocytosis,

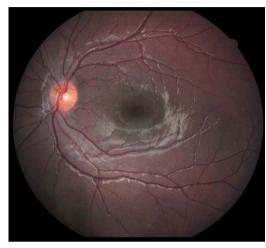


Figure 2 Left eye—normal fundus.

raised erythrocyte sedimentation rate and C reactive protein. Cerebrospinal fluid analysis showed pleocytosis and increased protein concentration. MRI showed features such as non-specific white matter enhancement and optic nerve enhancement (figure 3). Visually evoked potential showed prolonged latency suggestive of optic neuritis.

The patient was started on intravenous methyl prednisolone for 3 days and was then started on tapering doses of oral steroids under the guidance of paediatricians. The patient had thrombocytopaenic episodes which was promptly treated.

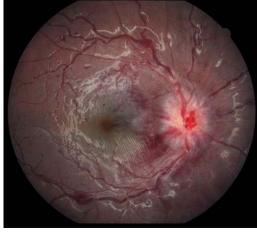


Figure 1 Fundus picture of the right eye showing disc hyperaemia, blurring of disc margins and peripapillary flame-shaped haemorrhages.

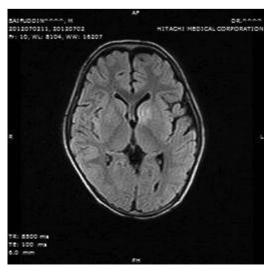


Figure 3 MRI of the brain showing features of acute disseminated encephalomyelitis.



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Figure 4 Resolving disc oedema.

After 3 months of follow-up the patient's vision improved to 6/36. Fundus picture showed mild persisting optic disc oedema (figure 4).

Contributors AJS was involved in data collection and compilation. SRJG was involved in drafting of the manuscript. NM and MS were involved in final approval of the manuscript.

Learning points

- ▶ Acute disseminated encephalomyelitis (ADEM) is an inflammatory syndrome affecting the central nervous system (CNS) that often is associated with parainfectious causes. Optic neuritis as an isolated manifestation is very rare.
- ➤ Visual prognosis is good in cases of optic neuritis in ADEM, once detected early and aggressively treated with intravenous steroids and immunoglobulins.
- ▶ Long-term CNS sequelae and disability was more common in patients who presented with optic neuritis. This alarms the need for an integrated approach as a team of ophthalmologists and paediatricians, for early detection, treatment and prevention of disability.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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