Case Report:

Optic Neuritis in Childhood May Be an Early Presentation of Alarming Disease-Acute Disseminated Encephalomyelitis

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Abstract

We are reporting a 5-year old girl with isolated acute visual loss related to a bilateral edematous optic neuropathy after an episode of viral illness. MRI brain showed bilateral, multiple sub-cortical hyper-intense lesions indicating white matter disease, acute disseminated encephalomyelitis (ADEM) with bilateral optic neuritis. Treatment with pulse steroids resulted in favorable visual recovery. After 4 weeks follow-up, MRI images showed complete resolution of sub-cortical pathology. Bilateral isolated optic neuritis in childhood may be the only presenting symptom in ADEM which requires proper evaluation and early therapeutic management. Up to our knowledge, this case is the first reported case in Saudi Arabia having this presentation.

Key Words: Optic neuritis – Acute disseminated encephalomyelitis – ADEM – Child – KSA.

Introduction

OPTIC neuritis (ON) is an uncommon but a challenging condition in childhood and has rarely been reported in literature [1]. Presentation of ON most frequently occurs around 7 years of age. Severebilateral visual loss is observed in 77% of subjects, which may be associated with diplopia, painful extra-ocular movements and headache [2].

The mode of onset of visual loss is an important clue to the etiology. Rapid onset is characteristic of demyelinating and inflammatory causes. Causes of other neurological manifestations may be present if there is concurrent acute disseminated encephalomyelitis (ADEM) [3].

Diagnosis of ON is made mainly on clinical grounds. On fundoscopy, about two thirds of patients have disc edema and 17% have disc pallor [2]. However, some newer technologies include multifocal visual evoked potential and optic coherence tomography. Usually ON improves in 90% of cases over several weeks to near normal visual acuity [3]. However, it has been observed in many studies that often some degree of optic atrophy occurs, even with good return of visual acuity [4].

ADEM is an acute monophasic, inflammatory, demyelinating central nervous system disease. Its incidence is 0.2-0.8 per 100,000 children in USA and Canada and 0.7 per 100,000 children in Germany [4]. In addition to ON, it may be characterized by the sudden onset of fever, headache, confusion drowsiness and occasionally seizures [6].

The neurological signs, especially in ON, typically appear 2-30 days following a viral infection (usually of upper respiratory tract) or vaccination (especially against measles or rabies) in 50-75% cases [1,5]. Brain MRI usually shows bilateral multiple discrete areas (occasionally diffuse), deep white matter lesions but rarely also involves the gray matter, like thalamus and basal ganglia. These lesions are of increased signal on T2-weighted imaging [1].

Cerebrospinal fluid (CSF) analysis may show moderate mononuclear pleocytosis and moderate increase in protein [7]. In ADEM fatalities, periventricular lymphocytic infiltration with local myelosis is a hallmark finding on pathology specimens [5].

There is no standard therapy for ADEM. Corticosteroids have been commonly used. ON has been found to be more responsive to steroids if it
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Optic neuritis occurs in the setting of simultaneous neurological dysfunction, such as ADEM. The use of intravenous steroids was found to hasten the visual recovery but not the final visual outcome [3]. There are reports of successful use of heparin if visual deterioration occurs despite being on steroid therapy [3]. Plasmapheresis and immunoglobulins are other options if steroids fail. In cases of malignant brain edema, refractory to conservative measures, a difficult possible decision is craniotomy [7]. Outcome of ADEM has a wide spectrum with a 57%-89% chance of full recovery [4].

We aim to stress that bilateral isolated ON in childhood may be the only presenting picture in ADEM, which requires appropriate early evaluation and therapeutic management.

Case Report

Our patient, a 5 year old Saudi girl, was perfectly healthy till 3 weeks prior to her presentation, when she had an episode of gastroenteritis which was treated conservatively with fluids only. She recovered within a few days. Suddenly, one day when she woke up in the morning, she was unable to visualize, even near objects. She was asking for help and assistance for her activities. She was afebrile, did not lose consciousness, had no history of trauma or seizure and was not passing through any unusual stress. She was thriving well and acquired all her developmental milestones at appropriate age. Her family history was unremarkable for any chronic neurological, psychiatric or ophthalmological morbidities.

Clinically, she was healthy, conscious, and alert, with no apparent dysmorphism. Her vital signs were within normal limits. On ophthalmological examinations, right eye had complete loss of vision, fixed dilated pupil and severe papilledema on fundoscopy. Left eye had less severe findings. There was positive light perception; pupil had sluggish reaction to light and mild papilledema on fundoscopy. Both eyes had clear anterior segment and normal extra-ocular movements without any squint. The rest of her CNS examination was unremarkable.

Investigations showed normal CBC with ESR of 5mm after first hour. Her CSF study showed no WBCs or RBCs on microscopy with protein of 24mg/dl (normal <45) and with sugar of 91mg/dl. Full panel of serum electrolytes, liver function tests and renal profile were all normal. Brain MRI showed bilateral multiple subcortical hyper-intense lesions seen in the biparietal lobes on axial and coronal T2-weighted images (Figs. 1,2), indicating white matter disease, most likely ADEM. Both optic nerves were tortuous, slightly thickened and enhancing in a picture highly suggestive of optic nerve neuritis bilaterally (Fig. 3). Four weeks later, follow-up MRI brain showed complete resolution of subcortical pathology after steroid therapy (Figs. 4,5).

On the basis of history, clinical and radiological findings, she was diagnosed as a case of ON secondary to ADEM. She was treated with pulse steroid (methyl prednisolone 30mg/kg/day) for 5 days followed by oral steroid initiated at 2mg/kg/day and tapered over a period of 2 weeks. The child showed good response and started to move independently since the 4th day of treatment. Her visual acuity at time of discharge (7th day of admission) was 6/36 bilaterally. On follow-up 4 weeks later, visual acuity was 6/18 in right eye & 6/9 in left eye and fundus examination showed mild optic atrophy (right > left). At the same time, MRI brain revealed almost complete resolution of subcortical pathology (Figs. 4,5).

![Fig. (1)](image1)
![Fig. (2)](image2)
![Fig. (3)](image3)

MRI brain, T2-weighted images showing bilateral, biparietal subcortical hyper-intense lesions, indicating white matter pathology.
Four weeks later follow-up MRI images showing complete resolution of subcortical pathology after steroid therapy.

**Discussion**

Childhood ON differs from the adult type by the fact that it does not have any gender or racial predilection, usually bilateral and associated with optic disc swelling. The risk of subsequent development of multiple sclerosis (MS) is less (36% in 2 years follow-up) [2]. Initially, visual acuity is very poor and no light perception is common. Visual recovery is usually good and more complete but takes longer than in adult variety. Moreover, residual poor vision and optic atrophy are more common than in adult ON [8]. ON is a common feature of both ADEM and MS [8]. Bilateral involvement is more in favor of the earlier, which has been found in almost a quarter of childhood cases of ADEM [1].

Diagnostic differentiation between ADEM, MS and other white matter diseases may, at times, be difficult but it is of paramount importance for prognostic reasons [1]. The signs that increase suspicion for MS in a child include the presence of exclusively white matter abnormalities on MRI (especially if monolesional), optic neuritis, isolated myelitis, a recurrent or polyphasic disease course or post-adolescent age and disproportionately high oligoclonal antibodies in CSF [8].

In our patient, the viral infection episode (gastroenteritis) prior to presentation, presence of bilateral optic neuritis together with typical neuro-radiological findings increased the possibility for the diagnosis of ADEM, although MS could not be ruled out at that point. It supported the fact that isolated ON in children may be the only presenting monophasic clinical manifestation of the disease. In our patient, multiple discrete white matter lesions were more suggestive of ADEM than MS. Similarly, normalization of the MRI findings at one month follow-up was also in favor of ADEM [8]. Bilateral ON has been found to be associated with a higher risk of MS development and more likely to have an abnormal MRI (71%) than the unilateral ON group (33%) [2].

Generally, visual recovery in pediatric ON is not as likely good as in adults, but in ADEM, usually it has a favorable final visual outcome [8]. Our patient responded well to corticosteroids therapy, recovering vision almost completely. There is a lot of controversy in literature regarding the development of MS in this specific condition. Some researchers supported the finding that the risk of MS following pediatric ON is very low, especially when disc swelling is bilateral and it occurs in a child less than ten years of age. They thought that long-term complications are variable and depend upon the variety of ADEM [1]. On the other hand, some authors suggested that children with brain MRI abnormalities at the time of the diagnosis of optic neuritis have an increased risk of MS. However, they concluded that further larger collaborative studies are needed to define the prognosis for these children [4,6].

Despite the clinical and neuroimaging findings in our patient, long-term follow-up and serial imaging is necessary to rule out MS. Indeed, it has been stated that the absolute criterion for distinguishing MS from ADEM is the absence of relapses in the latter, but this criterion is not universally accepted as relapse has been reported in ADEM also, in some studies [1]. One of those found recurrence of ON in ADEM, 19% in same eye, 14% for the fellow eye and 30% for the either eye [10].
Conclusions:

This report describes the occurrence of mono-symptomatic, monophasic and bilateral optic neuritis in a child with neuroradiological findings suggestive of ADEM. This diagnosis should be considered whenever there is a temporal relationship between an infection or vaccination and monophasic optic nerve involvement, prompting appropriate neuroimaging, medical treatment and close long-term follow-up. It also suggests that, as adult ON is a forme fruste of MS, pediatric ON should be considered a forme fruste of ADEM.

References


