International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

A Case of Papilloedema in a Child: A Diagnostic Dilemma

Santosh Kumar

Consultant Ophthalmologist, Department of Ophthalmology, Military Hospital Jodhpur, Rajasthan, India

Received: 05/11/2015 *Revised: 25/11/2015* *Accepted: 30/11/2015*

ABSTRACT

Papilloedema is defined as swelling of optic nerve head (optic disc edema) bilaterally without reference to its underlying cause. The aetiology of this condition is diverse in nature and difficult to establish, especially in pediatric cases. Increased intracranial pressure is implicated as most common cause for this entity. Papilloedema is at times confused with papillitis, seen in optic neuritis. Recognition of papilloedema or papillitis is of great clinical significance, especially in pediatric setting as timely intervention can be done to prevent vision threatening complications like optic atrophy. Here, I report a case of bilateral optic disc edema in a child who posed a diagnostic dilemma and was managed effectively.

Keywords: Idiopathic intracranial hypertension, optic neuritis, papilloedema.

INTRODUCTION

Papilloedema is defined as a non-inflammatory passive swelling of the optic disc (optic disc edema), produced by raised intra-cranial pressure (ICP). It occurs as a result of interrupted axoplasmic flow in the optic nerve due to transmitted elevated cerebrospinal fluid pressure in the subarachnoid space around it. Papilloedema is not very common in pediatric age group and if at all it is detected, it should be managed energetically to prevent vision threatening complications like optic atrophy. On the other hand optic disc edema in Optic neuritis (ON) is called papillitis as it is inflammatory in nature. ON is generally confused with papilloedema as it has similar clinical presentation. I encountered a similar case which posed a diagnostic dilemma and was managed effectively.

CASE REPORT

A 6 yrs old boy was referred from pediatric department with complaints of decrease in vision- both eyes, painful eye movements, morning headache and vomiting for duration of last 1 week. He was a known case of seizure disorder on medication for last 3 years but recently he was off- medication for this disease as he has been seizure- free for last 2 yrs. On examination his visual acuity (VA) in both the eyes was 6/18 with no further improvement even after refraction. Ocular movements were full and free but painful. His pupils were reacting sluggishly though size and shape of pupil were normal. On fund us examination he had bilateral optic disc edema (Fig 1). Other relevant ocular examinations and physical examination parameters were within normal limits.

He was investigated for the causes of papilloedema/papillitis keeping in view common aetiologies in pediatric age group. MRI (Magnetic resonance Imaging) Brain reported evidence of thickened bilateral optic nerve with prominent perineural subarachnoid spaces (Fig 2), normal ventricles and no evidence of

intracranial space occupying lesion (ICSOL) or any haemorrhage. The relevant haematological investigations were normal. Since there was no intracranial pathology accounting for papilloedema, a diagnosis of Idiopathic Intracranial hypertension (IIH), a rare cause was suggested. Papillitis in children generally have bilateral presentation and since decreased vision and painful ocular movements were not explained by IIH, optic neuritis was also added as differential diagnosis.

He was prescribed oral acetazolamide 125 mg thrice a day for one week in view of IIH. After one week he had mild relief from headache and vomiting but no improvement in visual acuity. Since there was no significant improvement, we planned to start with steroids. He was admitted in the hospital and given inj Methyl prednisolone 250 mg IV dissolved in 100 ml of saline once a day for three days after doing necessary investigations. In the mean time, oral acetazolamide was continued. After a week of this treatment he had complete relief from his symptoms and his unaided VA improved to 6/9. He was prescribed spectacle and with this, his VA was 6/6. The optic disc edema had also decreased (Fig 3) and he was discharged from hospital. He was advised follow- up at nearest eye hospital after a month as the child had to go back to his native place.

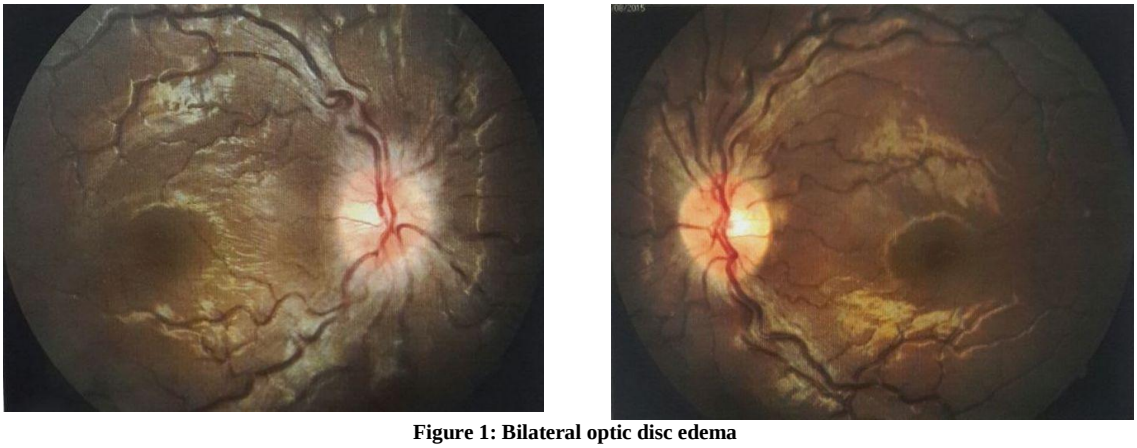

Figure 1: Bilateral optic disc edema

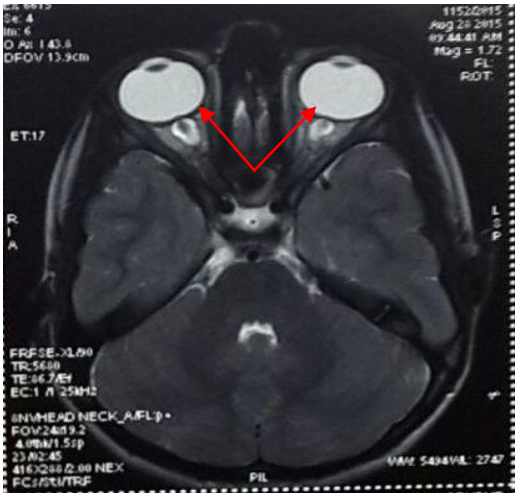

Figure 2: Thickened bilateral optic nerve with prominent perineural subarachnoid spaces (red arrow)

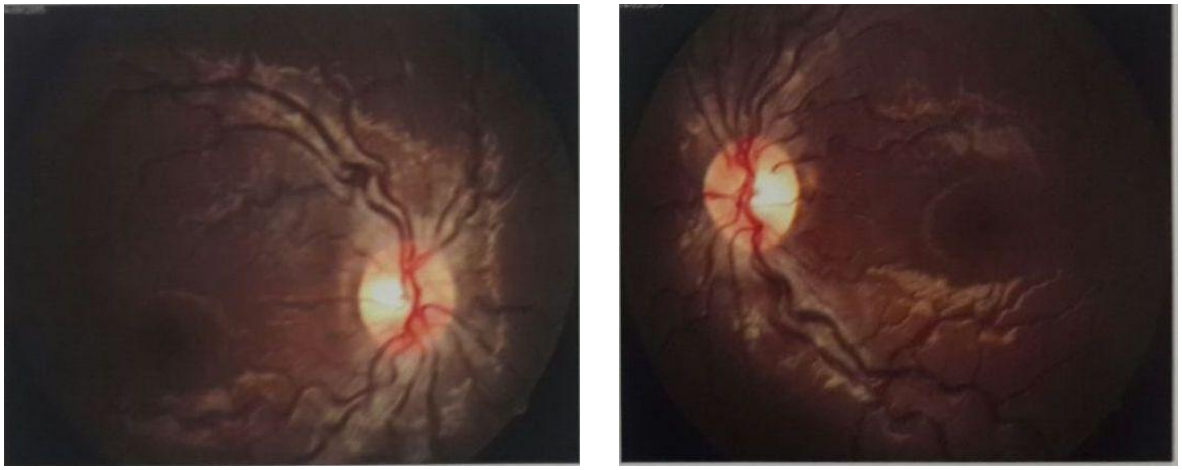

Figure 3: Optic disc edema resolved

DISCUSSION

Papilloedema may be observed at almost any age, but it is relatively uncommon in infants, because the fontanelle are not obliterated and the cranial sutures are not fused at this age; therefore they bulge and split leading to increase in head circumference rather papilloedema. [1] Possible conditions causing papilloedema in a child include intracerebral mass lesions, cerebral hemorrhage, head trauma, meningitis, hydrocephalus, impairment of cerebral sinus drainage, anomalies of the cranium, and idiopathic intracranial hypertension (IIH). [2] These conditions should be ruled out in a child presenting with optic disc edema. In this case also, MRI brain was done to rule out these conditions.

Optic disc edema in a child poses a difficult situation for the clinicians as there is a long list of conditions which can cause this. To diagnose the exact aetiology and rule out others is a herculean task as most of times child will not cooperate for the examination as well as investigations. In this case also a differential diagnosis of IIH and optic neuritis was made after ruling out other causes, based on MRI report. The presenting complaints of the child ie decrease in vision and painful ocular movements were suggestive of optic neuritis but morning headache and vomiting were in favour of IIH. In cases of IIH, lumbar puncture has been suggested to evaluate the ICP in adults but in pediatric population no clear cut guidelines exist and more over the consent for the procedure were not given by the parents of the child.

Idiopathic intracranial hypertension (IIH), also known as pseudotumour cerebri (PTC) is characterised by signs and symptoms of increased intracranial pressure (ICP) in the absence of space-occupying lesion. [2] IIH mainly occurs among obese women of childbearing age. [3] The exact prevalence of this condition in pediatric population is not known though few cases have been reported. Similar to adult patients, children are at risk of development of permanent visual loss. [4] Children with IIH usually complain of headaches and may have vomiting, blurred vision and horizontal diplopia. The presence of vomiting, headache and normal MRI Brain report in this case also goes with the features of IIH.

On the other hand, optic neuritis (ON) in pediatric patients is considered as a different entity in comparison to adults. It is normally bilateral and occurs after a viral infection. Due to presence of bilateral optic disc edema, it is confused with papilloedema. Morales et al in their study has shown that ON has good prognosis in pediatric age group. [5,6] ON is less often associated with multiple sclerosis in children than it is in adults, although approximately one third of children with

ON still develop multiple sclerosis. [7] This case also had good visual recovery and MRI Brain was not suggestive of any multiple sclerosis plaque.

A conservative approach was followed in this patient. That is the reason he was initially treated with acetazolamide, which is relatively safer in children. Since there was no major improvement in VA of child, a course of intravenous steroid was given. The patient improved dramatically following this. Oral steroid was not given after intravenous methyl prednisolone, as recommended by Optic neuritis treatment trial (ONTT) study. [8] ONTT gives the treatment protocol for the optic neuritis in adult population. Similar studies for pediatric population are not available and applying the recommendations of ONTT on children may not be justified.

CONCLUSION

Optic disc edema in a child poses a difficult diagnostic dilemma for the clinicians. However meticulous and systematic approach can clinch the diagnosis. Management should be started immediately to prevent vision threatening complications.

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How to cite this article: Kumar S. A case of papilloedema in a child: a diagnostic dilemma. Int J Health Sci Res. 2015; 5(12):424-427.

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