SHORT REPORT

Idiopathic intracranial hypertension: is papilloedema inevitable?

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Arch Dis Child 2002;**87**:223–224

Headaches and papilloedema are key features of idiopathic (benign) intracranial hypertension (IIH). We describe three children in whom IIH was diagnosed in the absence of papilloedema. Recognition of atypical cases of IIH is important because pressure lowering treatment may be effective.

The most frequent presenting symptoms of idiopathic (benign) intracranial hypertension (IIH) are headaches, vomiting, and visual disturbances. Neurological examination is normal with the exception of papilloedema and the occasional sixth nerve palsy. Diagnosis is established by showing cerebrospinal fluid (CSF) of normal composition with raised CSF pressure in the absence of intracranial mass lesion or ventricular dilatation. An opening pressure of 20 cm CSF is generally considered to be the upper limit of normal in children.[1]

We report cases of three children in whom a diagnosis of IIH was considered despite the absence of papilloedema. In each case described, the absence of papilloedema was confirmed by an ophthalmologist. CSF pressure was measured by lumbar puncture performed under general anaesthesia. Carbon dioxide concentrations were normalised and patient position controlled prior to measurement of CSF pressure in order to minimise any artefactual pressure elevation.

CASE 1

A 12 year old boy presented with a four year history of almost daily frontal headaches with photophobia. He was normotensive, body mass index 28. Neurological examination, fundoscopy, visual fields, and visual acuity were normal. Cranial magnetic resonance imaging (MRI) revealed an empty sella. A diagnosis of IIH was considered. CSF pressure was raised on three separate occasions (opening pressures of 34, 51, and 56 cm CSF), CSF composition was normal. He was commenced on acetazolamide. Following an initial improvement of several months duration, his headaches recurred and CSF pressure was raised despite maximal dose of acetazolamide. He has been referred for consideration of insertion of a lumboperitoneal shunt. He continues to remain symptomatic but his vision remains unaffected.

CASE 2

A 3 year old boy with autistic spectrum disorder presented with a two year history of recurrent vomiting. He would clutch the back of his head prior to vomiting, which seemed effortless. He was normotensive. Neurological examination including fundoscopy was normal. There was no apparent visual field defect. Investigations to exclude metabolic or anatomical causes of vomiting were performed. On cranial MRI, isolated prominence of the optic nerve sheaths was noted. A diagnosis of IIH was considered. CSF pressure was raised (opening pressure 27.3 cm CSF, closing pressure 18.5 cm CSF). Following lumbar puncture his symptoms improved. He was commenced on acetazolamide and remains symptom free.

CASE 3

An 8 year old girl presented with a 20 month history of daily bifrontal headaches with no associated visual or somatic features. She was normotensive. Neurological examination, fundoscopy, visual fields, visual acuity, and cranial MRI were normal. CSF pressure was raised (opening pressure 31 cm CSF, closing pressure 15 cm CSF). Following lumbar puncture she complained of severe headache, which was worse on standing, and was presumed to be a low pressure headache. Acetazolamide was poorly tolerated and was therefore discontinued. Control of her headache has remained difficult. There has been no documented deterioration of vision.

DISCUSSION

These children were seen as part of a case mix presenting to a tertiary service in a regional paediatric neurology unit. In this unit classical IIH is currently diagnosed around 12 times per year.

Papilloedema is a usual finding in IIH. However, atypical cases in which papilloedema is asymmetrical, unilateral, or absent have been reported, particularly in adults.[1–3] Marcelis and Silberstein described the clinical and radiological findings in 10 adults diagnosed with IIH despite absence of papilloedema.[2] The authors discuss the possible mechanisms accounting for lack of papilloedema, including acquired or congenital optic nerve sheath abnormalities and resolution of papilloedema in chronic IIH. The latter is of potential relevance in the cases we have described in view of the duration of symptoms prior to diagnosis in these patients. Mathew *et al* described 12 patients in whom they diagnosed IIH occurring concomitantly with migraine type headaches; one of the patients was a child. The patients presented with chronic daily headache with migrainous features which had failed to respond to conventional antimigraine therapy; none of the patients had papilloedema.[3] Therefore, in children with chronic daily headache sufficient to interfere with normal function and resistant to conventional treatment, IIH should be considered even in the absence of papilloedema. The most serious consequence of IIH is visual loss. There is no evidence that IIH in the absence of papilloedema is associated with visual loss,[2 3] and the severity of papilloedema has been positively correlated with the risk of visual deterioration.[4] Despite this, recognition of atypical cases remains important because pressure lowering treatment may successfully control symptoms resistant to other modes of therapy. Non-specific radiological abnormalities can occur in IIH and may help

Abbreviations: CSF, cerebrospinal fluid; IIH, intracranial hypertension; MRI, magnetic resonance imaging

establish a diagnosis. These include empty sella (which has been more frequently reported among the subset of patients with IIH who do not have papilloedema[2]) and abnormalities of the optic nerve.[5 6] It has been suggested that these occur as a consequence of chronically raised intracranial pressure.[5 6]

In the first two cases described, the diagnosis of IIH was not considered likely prior to neuroimaging because of the lack of papilloedema. In case 2, alternative explanations for vomiting were initially sought. It is possible that this patient was suffering from headaches but unable to communicate this. Both case 1 and 2 had neuroradiological abnormalities which have been previously reported in association with IIH and CSF pressure greater than the accepted upper limit of normal. Both showed initial symptomatic improvement following lowering of CSF pressure, thus allowing confident diagnosis of IIH despite the lack of papilloedema. In the third patient the nature and timing of the headaches was consistent with raised intracranial pressure, and CSF pressure was raised in the absence of intracranial mass lesion.

Conclusion

It is important to be aware that papilloedema is not an absolute prerequisite for considering IIH in a child. Non-specific findings on cranial MRI may contribute to prompt diagnosis in atypical cases. The possibility of IIH should be considered in children with chronic headache, as medical or surgical treatment to lower the CSF pressure may be effective means of symptom control.

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Accepted 12 April 2002

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ARCHIVIST

Staphylococcal necrotising pneumonia

In most paediatricians' minds the term staphylococcal pneumonia is associated with a picture of severe bilateral disease in infants and young children, often with pleural effusion, empyema, pneumothorax or pyopneumothorax, lung abscesses, and pneumatoceles. That disease has been uncommon in the wealthier countries since the 1960s. Now an even more virulent form of staphylococcal pneumonia has been described from France (Yves Gillet and colleagues. *Lancet* 2002;**359**:753–9).

Staphylococcus aureus sometimes carries the gene for a toxin called Panton-Valentine leukocidin (PVL). In 1998 a review of data held by the French Reference Centre for Staphylococcal Toxaemia, in Lyon, showed that there had been eight cases of PVL-positive community-acquired staphylococcal pneumonia since 1986, six of whom had died. The *Lancet* report includes these eight cases, eight more cases from 1999, and 36 PVL negative cases also from 1999.

PVL positive *S aureus* pneumonia affected children and young adults (median (interquartile range) age 14.8 (5.4–24.0) years whereas PVL-negative pneumonia was a disease of the elderly (70.1 (59.2–81.4)). Three quarters of the PVL-positive cases died, average survival after admission being only 4 days. The disease often began with an influenza-like illness progressing within 2 days to very severe pneumonia. Common clinical features included high fever, hypotension, tachycardia, cyanosis, haemoptysis, and leucopenia. Pneumothorax or pneumatoceles did not occur. Radiologically there were multilobar alveolar infiltrates on admission, often with the later development of bilateral interstitial infiltrates consistent with acute respiratory distress syndrome. Ten patients had, or developed, pleural effusion.

Three of the 16 PVL positive patients had postmortem examinations that showed massive alveolar haemorrhage with necrosis of the interalveolar septa. There was also extensive necrosis of the mucosa of the trachea and bronchi with haemorrhagic foci.

Since the end of 1999 this laboratory has confirmed another 13 PVL positive cases and they also confirmed three fatal cases in the USA in 1999. This clinical picture seems to have been rare in the past but a similar illness caused the deaths of 385 American soldiers in 1918 soon after the influenza pandemic.