Sir,

An atypical presentation of Wernicke's encephalopathy in an 11-year-old child

Wernicke's encephalopathy has classically been described in adult alcoholics as the triad of altered mental status, ocular abnormalities, and ataxia. It is caused by thiamine (vitamin B1) deficiency and is potentially fatal. It is important to highlight the fact that the classical triad is not always typical and that awareness of this condition by ophthalmologists is vital, so cases will not be missed, and early treatment can be instigated. We report a case of Wernicke's encephalopathy in a child where ocular symptoms were predominant and onset of blurred vision actually prompted her admission to hospital despite illness for the preceding few months.

Case report

An 11-year-old girl was referred to hospital complaining of a 2-day history of blurred vision bilaterally. She described this as a central haziness, and on questioning, her parents revealed that she had an 11-week history of vomiting, poor appetite, lethargy and weight loss. Problems began after an episode of laryngitis 3 months previously during which she had choked on a sausage and subsequently felt too afraid to eat. She stated 'I'm scared I will choke and die' and had been vomiting up anything she tried to ingest at a frequency of 5–6 episodes a day. Her weight had dropped 20 kg over the 11 weeks. Clinical examination revealed she had upbeat nystagmus in the primary position with right and left gaze paresis. Visual acuity was 6/60 right eye and 6/24 left eye with a bedside Snellen chart and there was no relative afferent pupillary defect. She was able to see the Ishihara colour test plate with the left eye only and no plates were seen with the right eye. Both optic discs were mildly swollen nasally with peripapillary intraretinal haemorrhages (Figure 1a). Her gait was unsteady with abnormal finger-nose pointing on the right side and she also was noted to have subtle short-term memory problems. Her full blood picture, serum lactate, and B12 and folate levels were normal. Serum potassium was low

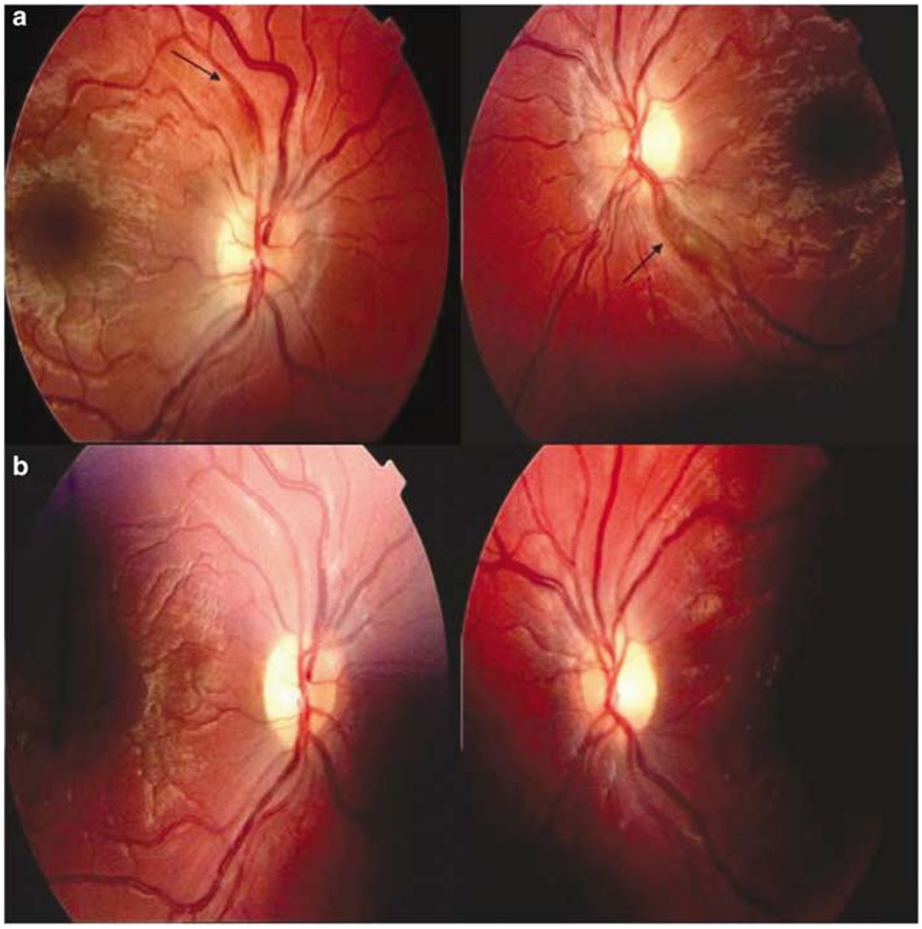

Figure 1 (a) Fundal photographs showing nasal disc swelling and intraretinal haemorrhages (see arrows) (taken a day after first dose of thiamine treatment before complete resolution of findings). (b) Photographs 1 week after treatment with thiamine showing resolution of the fundal findings.

at 2.8 consistent with persistent vomiting. Urgent enhanced CT scan of brain was normal and lumbar puncture and CSF analysis revealed no organisms or tumour cells, normal protein, glucose, and cellular counts. She subsequently had an MRI scan of brain, which showed typical midline increased signal intensity in the dorsal mid-brain, thalami, and mamillary bodies (Figure 2). The diagnosis of Wernicke's encephalopathy was confirmed and she was treated with intravenous thiamine. The following day vision had improved to 6/12 right eye and 6/24 left eye and gaze had improved dramatically with much less nystagmus. At review 2 weeks later, her ataxia had resolved, vision was 6/12 in each eye, and discs and fundi appeared normal with full colour vision (Figure 1b).

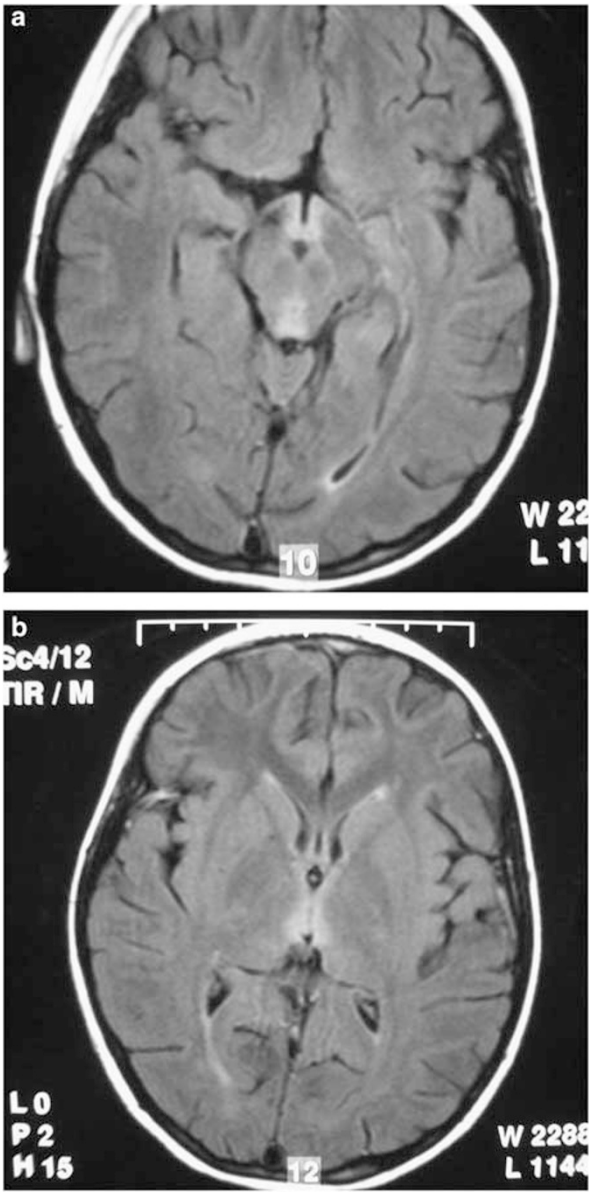

Figure 2 Axial fluid-attenuated inversion recovery (FLAIR) images showing increased signal in the (a) mamillary bodies and dorsal midbrain and (b) thalami.

Comment

Severe thiamine deficiency is known as beriberi and cerebral beriberi is also known as Wernicke's encephalopathy. Wernicke's encephalopathy in children has previously been described in the neurological and radiological literature. It has occurred in various conditions leading to malnutrition such as after autologous bone marrow transplantation,[1] chronic gastrointestinal disease,[2] prolonged starvation,[3,4] and malignancy[5]. The case reported in this paper is unusual in that it was the blurred vision that precipitated her attendance to the hospital as an emergency, and the ophthalmological features that predominated to help clinch the diagnosis.

The presentation of Wernicke's encephalopathy can be extremely variable and only about 16% of patients present with the classical clinical triad of symptoms and 80% of patients have not been diagnosed before death.[6] In the paediatric population, the classical triad was present in only 21.4% at onset.[5] Mortality in Wernicke's encephalopathy has been reported from 10 to 20%, therefore prompt recognition of the condition and early treatment are vital.[7]

Fundal findings including disc swelling and intraretinal haemorrhages have been described rarely in Wernicke's encephalopathy.[1,2,4,5,8–10] The mechanism of the development of disc swelling in this condition is poorly understood. It has been postulated that the nerve swelling is due to a Wernicke's encephalopathy-associated optic neuritis and that the haemorrhages are secondary to the disc oedema.[8] This would correlate well with the findings in this child as she described bilateral central scotomata, found to be associated with poor colour vision, and this gradually resolved with resolution of the oedema and haemorrhages after thiamine treatment. Her reduced vision and scotomata would be consistent with an optic neuritis-like condition. A further theory involves the suggestion that the pathological findings of necrosis of nerve cells and myelinated structures in Wernicke's occur in the optic nerves and cause the resultant oedema.[8] Kulkarni *et al*[11] highlight the fact that, although optic disc swelling and optic neuropathy are not commonly described, these findings should not prompt the clinician to exclude Wernicke encephalopathy. They reported a similar case of thiamine deficiency in a young woman who had gastric bypass surgery for obesity, who also presented with similar ophthalmic features. They postulate that retinal haemorrhages could also have been caused by vomiting,

but mainly peripapillary distribution would be unusual if that was the case.

In this child, malnutrition was precipitated by an episode of choking and a subsequent fear of this occurring again. A similar case has been reported previously in the literature in a 10-year-old boy with good recovery of neurological function 1 month after treatment.[3]

Acknowledgements

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Sir,

Increased and persisted subretinal haemorrhage after photodynamic therapy for choroidal neovascularization secondary to angioid streaks

Increased subretinal haemorrhage after photodynamic therapy (PDT) for choroidal neovascularization (CNV) secondary to angioid streaks

Angioid streaks are discontinuities in thickened and calcified Bruch's membrane.[1] CNV has been reported to occur in patients with angioid streaks[2] and, because of macular involvement and central visual loss, the visual prognosis is usually poor.[1,2] The treatment efficacy of PDT for angioid streaks with CNV remains under evaluation.[3–5] In this study, we report a case of increased subretinal haemorrhage after PDT for CNV secondary to angioid streaks with a final disciform scar formation.

Case report

A 52-year-old male was referred to our clinic due to image distortion of his right eye for months. The fluorescein angiography disclosed an extrafoveal CNV lesion secondary to angioid streak (Figure 1a and b). The greatest linear dimension (GLD, including the CNV, the area of leakage, and areas of blocked fluorescence in the lesion) of the CNV was 700 μm. He was known to suffer from angioid streaks, which had led to visual loss in his left eye due to disciform macular scar. The best-corrected Snellen visual acuity (BCVA) was 6/8.6 in his right eye and 6/60 in the left eye. Verteporfin PDT was performed on the right eye to treat the extrafoveal CNV. Unfortunately, increased subretinal haemorrhage was noted 2 months after PDT and it persisted for 20 months during follow-up. The CNV was progressively enlarged to 3000 μm associated with subfoveal involvement