Transient cortical blindness as a manifestation of solitary cysticercus granuloma

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SUMMARY

Neurocysticercosis is recognised as a significant cause of neurological morbidity in endemic regions. The wide range of pleomorphic and non-specific neurological manifestations of neurocysticercosis must be kept in mind by physicians, as the disease has shown resurgence in developed countries. When an atypical presentation of an unusual tropical disease occurs in non-endemic regions, the diagnosis is often missed. We describe a case of a 4-year-old girl who presented with a history of transient bilateral loss of vision with headache and vomiting. Brain MRI revealed the presence of a single cysticercus granuloma in the occipital lobe. A diagnosis of symptomatic occipital lobe seizure secondary to neurocysticercosis was made. She was given a course of albendazole. There was no recurrence of symptoms at 3 years follow-up. Occipital seizures that are associated with ictal amaurosis closely mimic basilar migraine. Such cases benefit from neuroimaging in order to rule out the underlying structural causes.

BACKGROUND

Since neurocysticercosis (NCC) can present with a wide range of neurological presentations, it is important for a physician to be aware of such atypical presentations in order to avoid untimely delay in diagnosis and management.

CASE PRESENTATION

A four-and-half-year-old Indian girl was brought to the emergency department by her parents after she had developed transient loss of vision in both eyes, which lasted for 2 min. This was followed by headache and several episodes of vomiting. The episode of blindness was reported by both the patient and her parents. However, by the time, she was assessed by the physician in the hospital, there was no visual deficit noted. There were no associated abnormal movements or posturing. There was no history of fever, head trauma or similar episodes in the past.

Her medical history was significant for an atrial septal defect and an episode of simple febrile convulsion that she experienced 1 year ago. Although the child was born in the rural parts of India, she was residing in Dubai with her parents. They usually visited their town every summer and their last trip was 1 year prior to presentation. She was not in contact with anyone who was ill. Her immunisations were all up-to-date. There was no family history of any neurological illness.

On examination, she was afebrile, fully conscious and oriented. Her anthropometric measurements were within normal range. There were no dysmorphic features present. There was no neck rigidity. There was no sign of any neurological deficit.

Ophthalmological examination was normal. Besides a grade 2 systolic murmur that was auscultated in the precordium, the other systemic examination was unremarkable.

INVESTIGATIONS

All routine blood investigations were normal. Stool examination was negative for any cyst or ova. Blood culture, stool culture and urine culture were negative. Toxoplasma serology, HIV serology and tuberculin skin test were all negative. Serological test for cysticercosis antibodies in blood (by enzyme-linked immunoelectrotransfer blot) was also negative.

Brain CT showed a subcortical hypodensity in left occipital lobe with perifocal oedema causing obliteration of high convexity cortical sulci. Subsequently, a brain MRI with contrast was then conducted for further evaluation. It showed single-focus hyperintense lesion in left occipital lobe with surrounding oedema (figures 1 and 2). The lesion showed perilesional enhancement after contrast administration (figure 3). These findings were highly suggestive of a single cysticercus granuloma. Brain EEG showed low brain threshold for epileptic discharge. Echocardiography revealed a small ostium secundum atrial septal defect.

DIFFERENTIAL DIAGNOSIS

- ▶ Migraine with visual aura
- ▶ Idiopathic childhood occipital epilepsy of Gastaut
- ▶ Tuberculoma
- ▶ Brain tumours

TREATMENT

The episode of transient blindness in our patient was due to the presence of a single cysticercus granuloma in the occipital lobe that triggered an occipital seizure. The spread of seizure activity to both the occipital lobes caused transient bilateral blindness. This is termed as ictal amaurosis and is a known presentation of occipital lobe seizures. Although the serological result was not in favour of cysticercosis, based on the clinical and radiological findings, a presumptive diagnosis of single cysticercus granuloma was made.

She was therefore given a 7-day course of albendazole, along with a short course of prednisolone and phenytoin.

OUTCOME AND FOLLOW-UP

A repeat brain MRI with contrast was repeated 2 months later which showed a complete resolution of the lesion.

The child was seen again 3 years after the first presentation. There was no further recurrence of

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Figure 1 Axial T2-weighted MRI of the patient before treatment. It shows a single-focus hyperintense ring lesion in the occiput, with surrounding oedema.

symptoms during this interval and there was no neurological abnormality detected during clinical examination.

DISCUSSION

NCC is the most common parasitic disease of the central nervous system, which is caused by the larval stage of the pork tape worm-*Taenia solium*.¹ It has received attention as an important cause of epilepsy and neurological morbidity in endemic regions, where pig-rearing is practiced in substandard conditions.²⁻³ However, because of the ease of international travel, it is becoming increasingly prevalent in industrialised countries, where the disease is seen predominantly, but not exclusively among the immigrant population.⁴⁻⁵ Risk factors for acquiring cysticercosis include residence in an endemic region or contact with a tapeworm carrier.

Figure 2 Sagittal T2-weighted MRI of the patient before treatment. It shows a single-focus hyperintense ring lesion in the occiput, with surrounding oedema.

Figure 3 Axial T1-weighted MRI after contrast administration. There is perilesional enhancement of the lesion.

Del Brutto *et al*⁶ have suggested a diagnostic criterion for NCC based on clinical, neuroimaging, serological, histopathological and epidemiological criteria. The presence of neuroimaging findings suggestive of NCC, resolution of cystic lesion after cysticidal therapy, history of travel to disease-endemic region and the suggestive clinical picture fulfils the criteria for diagnosis of NCC in our patient.

NCC is an intriguing disease as its manifestation depends on many factors like number, stage and site of cysticerci, as well as the host inflammatory response.⁷⁻⁸ This explains the wide range of unusual symptoms with which it can present.⁹ In contrast to adult patients, the most common radiological finding in children with NCC is a single parenchymal cyst.¹⁰

Seizures are considered the most common manifestation of NCC due to parenchymal involvement and presence of cysticerci in the epileptogenic cerebral cortex.¹¹ Several case reports of unusual presentations of NCC-like extrapyramidal disease,⁹ hemiballismus,¹² dorsal midbrain syndrome¹³ and nominal aphasia¹³ are reported in the literature. Therefore, NCC should always be kept in mind as a differential diagnosis when a child from an endemic region presents with any neurological impairment, behavioural disorder or learning disability.¹⁴

Visual loss in cases of cysticercosis can be multifactorial. Chang *et al*¹⁵ analysed 23 patients with visual loss due to cysticercosis. Almost half of the cases were due to optic neuropathy as a result of papilloedema. Other causes included were chiasmal damage and retrochiasmal lesions. Prasad *et al*¹⁶ have discussed a case of cysticercal encephalitis which caused permanent visual impairment.

However, a transient binocular visual loss is a different clinical entity. Its presence virtually always indicates the occurrence of a transient dysfunction of the visual cortex. This occurs in migraine, cerebral hypoperfusion, head trauma, posterior reversible encephalopathy syndrome and seizure activity. In our case, transient bilateral blindness with headache is explained by occipital seizures that are triggered by the presence of solitary cysticercus granuloma in the occipital lobe. Elementary visual hallucination, ictal blindness and ictal headache are the recognised features of occipital lobe seizures.¹⁷⁻¹⁸ The pathophysiological basis of ictal blindness in cases of occipital seizures is poorly understood; however, a bilateral visual loss is postulated to be due to bi-occipital spreading of seizure activity.¹⁷ Such

manifestations closely mimic symptoms that physicians mistake for migraine with visual aura.¹⁹ In contrast to popular belief, a recent study by Rozen *et al*²⁰ concluded that binocular blindness with migraine headache is, in fact, a very rare occurrence, which accounted for only 1.6% of migraine patients referred to a headache specialty clinic. Menon¹⁸ reported two other cases of symptomatic occipital lobe epilepsy due to NCC that presented with visual hallucination and headache. They were misdiagnosed and treated for 2 years as migraine with visual aura. Both patients became seizure-free with appropriate treatment of the underlying disease and epileptic seizures. Sharma *et al*²¹ reported four other patients with single cysticercus granuloma in the occipital lobe who presented with migraine-like visual hallucinations.

Recognition of cases with occipital epilepsy remains a diagnostic challenge. Jobst *et al*²² demonstrated that less than two-thirds of patients with the former diagnosis had posterior scalp EEG changes. It is also important to note that even distinguishing symptomatic occipital epilepsy from other epileptic phenomenon like idiopathic childhood occipital epilepsy of Gastaut is difficult and experts recommend neuroimaging to rule out underlying structural causes for seizure.¹⁹

There are ongoing studies that are evaluating the role of sero-diagnostic techniques in patients with NCC. Serological tests have limited sensitivity especially in cases with a single parenchymal cyst.²³ Therefore, a negative serology does not exclude the diagnosis in patients with a compatible clinical presentation and neuroimaging findings.

Treatment issues regarding NCC are controversial and treatment of patients should be individualised according to the clinical scenario. This is due to differences in the infection process as well as the host response.²⁴ In general, eradication therapy with cysticidal agents benefit symptomatic patients with multiple, live (non-calcified) cysticerci. A meta-analysis by Abba *et al*²⁵ found that administration of cysticidal therapy in patients with viable lesions reduced the number of lesions. Another study by Kalra *et al*²⁶ concluded that the combination of albendazole and dexamethasone increases resolution of NCC lesions and reduces the risk of recurrence of seizure among children presenting one or two ring-enhancing lesion on CT. The role of steroids is to suppress the inflammatory response induced by destruction of live cysticerci. On the other hand, Singhi *et al*²⁷ reported that although eradication therapy is recommended in children with multiple lesions, many children with single lesions will improve without antiparasitic therapy. Nevertheless, the time duration taken for spontaneous resolution of single cysticercus lesion is highly variable and can even take a few years.²⁸

NCC is predominantly a treatable and preventable condition. Thus, the possibility of its diagnosis should be kept in mind during the diagnostic work-up of any child with symptoms suggestive of a neurological, cognitive or personality disorder, especially among those who have travelled from an endemic region.

Learning points

- ▶ Consider a diagnosis of occipital lobe seizure in a patient who presents with acute-onset of cortical blindness.
- ▶ Consider a diagnosis of neurocysticercosis (NCC) in any child from endemic region who presents with sudden onset of neurological, cognitive and personality disorders.
- ▶ Emphasise on the role of brain imaging in suspected cases of occipital lobe epilepsy to rule out structural lesions.
- ▶ Understand the limitation of serological tests in cases of NCC.

Competing interests None.

Patient consent Obtained.

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