Atypical ophthalmological presentation of neurocysticercosis in two children

Jitendra Jethani, MS, DO, DNB, P. Vijayalakshmi, MS, DO, and Mahesh Kumar, DO, DNB

Cysticercosis is an infestation by *Cysticercus cellulosae*, the larval form of the cestode *Taenia solium*. *C. cellulosae* is preferentially encysted in the brain, striated muscles, and subcutaneous tissue. Seizures, hydrocephalus, meningitis, and focal symptoms caused by large cysts are the principal manifestations. Neuroophthalmologic signs are common with the meningitic and hydrocephalic manifestations of the disease. The first ophthalmologic clues of cysticercosis are typically papilloedema, pupillary abnormalities, or nystagmus. We report an atypical presentation of neurocysticercosis in two patients, with chief complaints of vertical diplopia and nystagmus.

Case 1

A 10-year-old boy from the southern part of India presented with a 5-day history of vertical diplopia. There was no history of fever, headache, or any complaint suggestive of raised intracranial pressure. On examination, visual acuity was 6/6 in both eyes, and the anterior segments, pupils, and fundi were normal, with no evidence of proptosis. On cover test, an 8 to 10° left hypertropia was noted at near and distance with no change on head tilt. Examination of ocular movements revealed a fine, vertical jerk nystagmus in the primary position and in upgaze, with mild underaction of the left inferior rectus (IR) muscle. Convergence was normal. The separation of the images was found to be maximum in levoversion. Hess charting confirmed underaction of the left IR. Orbital ultrasonogram revealed no abnormal muscle thickness or any other pathology. The presence of upbeat nystagmus of acute onset with vertical diplopia led us to order a computed tomographic (CT) scan of the brain (Figure 1A), which revealed a tiny, ring-enhancing lesion with scolex and peri-lesional edema involving the left thalamic region and cerebral peduncle (along the course of the oculomotor nerve). Magnetic resonance imaging (MRI) (Figure 1B-C) confirmed the above findings and also the colloidal vesicular stage of cysticercosis. A small obstructive hydrocephalus, resulting in mild dilation of the third and lateral ventricles, was also noted. The patient was initially treated with intravenous steroids—Dexamethasone (Wockhardt Ltd, Mumbai, India), followed by oral prednisolone 1.5 mg/kg/day and albendazole (Glaxo Smith Kline, Pune, India) 15 mg/kg/day in two divided doses for 21 days. The patient was lost to follow-up but was symptom-free according to a written communication.

Case 2

A 6-year-old girl from the southern part of India presented with a 20-day history of double vision. The onset was sudden with no pain, headache, or vomiting; however, there was a history of fever 2 weeks before the onset of symptoms. After a detailed examination by an ophthalmologist and neurosurgeon, the patient was diagnosed with a pupil-sparing partial oculomotor nerve paresis (ptosis, exotropia, limited adduction, elevation and depression, and normal pupil). A CT scan of the brain revealed a cystic lesion with scolex in the substance of the midbrain, extending up to the third ventricle and aqueduct, with mild dilation of the ventricles. A radiological diagnosis of neurocysticercosis was made. The patient was given systemic albendazole 15 mg/kg/day in two divided doses for a period of 21 days, with a course of oral prednisolone 1.5 mg/kg/day which was tapered over 2 months. On subsequent visits, the patient showed improvement in ptosis and ocular movements and became symptom free.

Six weeks later, the patient presented with a recurrence of the diplopia. The clinical findings included a left exo-
tropia, restriction of adduction, elevation, and depression, and a dilated and sluggish pupil. Albendazole and prednisolone therapy were resumed. On follow-up, the patient was free from double vision with improvement in ocular motility, although the pupil remained dilated and sluggish. Less than a year later, the diplopia recurred with ocular movements showing limitation of adduction and depression. At this time, she was also noted to have upbeat nystagmus, more prominent in upgaze. The patient was subjected to thorough investigations for tuberculosis since intracranial tuberculosis was considered a differential diagnosis for a ring enhancing lesion. Antituberculous treatment with a two-drug regimen of isoniazid and rifampicin (Lupin, Aurangabad, India) was initiated, along with oral prednisolone 1.5 mg/kg/day. The patient became asymptomatic 8 to 12 weeks after starting the treatment. Six months later, the patient returned with a fourth recurrence of symptoms, with double vision increasing on down- and left-gazes. The patient was given oral praziquantel (Chandra Bhagat Pharma, Mumbai, India) 50 mg/kg/day in three divided doses along with oral prednisolone for 15 days. Within 15 days, she was totally free of symptoms. The corrected visual acuity in both eyes was 6/6. With 5 years of follow-up, she has remained asymptomatic except for upbeat nystagmus in upgaze.

Discussion

Cysticercosis is an episodic, relatively benign neurological disease. Neuroophthalmic signs are common in meningitic and hydrocephalic manifestations of the disease. The first sign to suggest the presence of a serious neurological disease is often the recognition of papilledema, occasionally associated with secondary abducens nerve paralysis, optic atrophy, pupillary abnormalities, or nystagmus. Cranial nerve involvement is otherwise surprisingly rare. The reported incidence of diplopia is 10.7% (6/86) in neurocysticercosis. For the diagnosis of neurocysticercosis, serological tests such as the enzyme-linked immunosorbent assay and enzyme-linked immuno-electro transfer blot have a low sensitivity, especially with small, isolated lesions. The evidence of a scolex on CT or MRI scan has been considered pathognomonic of neurocysticercosis. An MRI scan would have helped pinpoint the diagnosis earlier in the second case, but this was not available in our city at that time.

Upbeat nystagmus in children has been reported in Pelizaeus-Merbacher disease, Wernicke’s encephalopathy, and brainstem encephalitis; in peripheral disorders such as chronic middle ear infection; and secondary to organophosphorus poisoning. The nystagmus is thought to be due to the imbalance of the vertical vestibulo-ocular reflex and the otoliths. The close proximity of the lesion in both cases to the rostral interstitial nucleus of the medial longitudinal fasciculus and the interstitial nucleus of Cajal may have caused the nystagmus in this case.

Cysticercosis has long been recognized as a cause of the pretectal or dorsal midbrain syndrome, cerebrovascular complications, and internuclear ophthalmoplegia. The lesions may be solitary or multiple. Both of our cases had a solitary lesion in the midbrain.

In the first case, mild weakness of the IR muscle (inferior division of III nerve) caused symptoms of diplopia. The presence of vertical nystagmus was the clinical clue of a midbrain lesion. Contrast MRI and CT scans revealed a characteristic granuloma with an eccentric nidus suggestive of neurocysticercosis. The typical radiological findings and a remarkable clinical improvement with prednisolone and albendazole confirmed the diagnosis of a cysticercus cyst.

In the second case, the frequent recurrence of symptoms over a short time span resembles the case described by Kim et al. Their case presented with a complete oculomotor nerve palsy with pupillary involvement, with recurrence of symptoms on tapering the steroids. Our patient differed in that she was a young girl; she did not have a pupillary involvement initially, and she later developed persistent upbeat nystagmus. As with the patient of Kim et al, the recurrence in our patient could have been due to an inadequate dosage and duration of steroid therapy. Earlier recognition of the diagnosis would have avoided unnecessary antituberculous treatment, emphasizing the importance of neuroimaging studies in patients with acquired upbeat nystagmus.

References