Intraventricular neurocysticercosis causing acute unilateral hydrocephalus

Neurocysticercosis commonly presents with seizures resulting from cystic or calcified parenchymal brain lesions. Here we describe a patient with intraventricular neurocysticercosis causing acute unilateral hydrocephalus and discuss diagnosis, imaging and management.

Case
A 30 year old female presented in a confused state after a three week history of headaches with features of raised intracranial pressure (collateral information). She was awake but disoriented with no focal neurological signs. Fundoscopy revealed engorged retinal veins. A contrast enhanced CT brain scan showed acute dilatation of the right lateral ventricle with midline shift (Figure 1B-D). The third and fourth ventricles were normal. This indicated obstruction at the level of the right foramen of Monro. No cause of the obstruction could be seen on the CT scan. A calcified granuloma was present in the left parietal lobe (Figure 1A).

An MRI was therefore done and showed a thin walled cyst in the occipital horn of the right lateral ventricle with a hyperintense scolex at the periphery of this cyst (long arrow, Figure 2A-D). Another cyst was seen in the right lateral ventricle close to the foramen of Monro (short arrow, Figure 2A, C). No parenchymal cysts were seen on the MRI scan.

Her blood investigations were unremarkable but of note was that the cysticercosis enzyme linked immunosorbent assay (ELISA) was negative.

Based on the imaging the diagnosis of intraventricular neurocysticercosis was made. Endoscopic third ventricular fenestration was therefore carried out and 3 thin-walled cysts were removed.

Immediately post surgery a noticeable improvement in her cognition was noted. The patient was treated with steroids and a course of Albendazole. CT and MRI brain scans repeated one week later showed significant improvement of the hydrocephalus (Figure 3A, B).
Neurocysticercosis (NCC) is the commonest central nervous system parasitic disease worldwide, comprising 10% of acute neurological cases in endemic areas. It is also the commonest cause of acquired epilepsy worldwide. In South Africa the Xhosa speaking people of the Eastern Cape have the highest prevalence of NCC.

Cysticercosis results from infection with the larval form of the tapeworm Taenia solium. Brain parenchyma is seeded through hemogenous dissemination. The cerebrospinal fluid (CSF) spaces are seeded via the choroid plexus. Intraventricular neurocysticercosis is an uncommon form of NCC and the literature in this regard, in particular from Africa is sparse.

Intraventricular neurocysticercosis occurs in 7-30% of patients with NCC. Intraventricular cysts occur singly or in multiples. Parenchymatous disease may be associated in 24% of patients with intraventricular neurocysticercosis. The fourth ventricle is the commonest site for intraventricular neurocysticercosis (53%), followed by the third ventricle (27%), lateral ventricles (11%) and the aqueduct (9%). Clinical presentation depends on the size of the parasites, their location inside the ventricular system, and the coexistence of granular ependymitis. Obstructive hydrocephalus develops in approximately 30% of all patients with NCC. The ependymitis caused by death of the larvae can lead to communicating hydrocephalus.

CT scan has a sensitivity and specificity of over 95% in the diagnosis of neurocysticercosis, however intraventricular cysts are poorly visualised on CT as they are of the same density as CSF. Their presence can be inferred from distortions of the ventricular system causing asymmetrical or obstructive hydrocephalus. Intraventricular cysts are better visualized by MRI as demonstrated in our case.

The cysticercosis ELISA is a simple and economical test but is neither sensitive nor specific to cysticercosis. For diagnosis of active neurocysticercosis the test shows 50% sensitivity with 70% specificity in serum and 87% sensitivity with 95% specificity in CSF. Hence this test is not reliable for the diagnosis of neurocysticercosis. This could explain why our patient’s serum ELISA was negative. The enzyme-linked immunoelectrotransfer blot assay (EITB) using purified extract of T. solium antigen detects the specific antibodies and has a sensitivity of 98% and specificity of 100%. This is the assay of choice but is not available in South Africa.

Treatment modalities of intraventricular neurocysticercosis include antihelminthic medication, microneurosurgical removal, ventriculoperitoneal shunting, and endoscopic management. Endoscopic excision of intraventricular neurocysticercosis and the creation of an internal CSF diversion for hydrocephalus has become the preferred management. Our approach to treating our patient was in keeping with this literature and had the successful outcome described above.

According to recent proposed diagnostic criteria for NCC our patient had one absolute criterion (cystic lesion with scolex on neuro-imaging), and hence a definitive diagnosis of neurocysticercosis.

Cysticercosis is endemic in South Africa. NCC is a common, treatable complication and typically presents with seizures. Unusual neurological manifestations with non-specific symptomatology and radiological features, like hydrocephalus with distorted, asymmetrical ventricles or unexplained acute obstructive hydrocephalus, should be recognised as possible manifestations of intraventricular neurocysticercosis. Brain MRI is the modality of choice to identify atypical presentations of NCC. Serological testing with cysticercosis ELISA has low sensitivity and specificity.

K Jivan, A Mochan, G Modi
Division of Neurology, Department of Neurosciences, University of the Witwatersrand, Johannesburg, South Africa

References