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Intrafourth ventricular neurocysticercosis

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Abstract :
We describe the clinicopathologic features and management of an example of a sixteen year old girl who presented with a fourth ventricular cysticercal cyst and had a successful outcome following an open excision of the cyst. The various surgical options for the management of this condition are discussed.

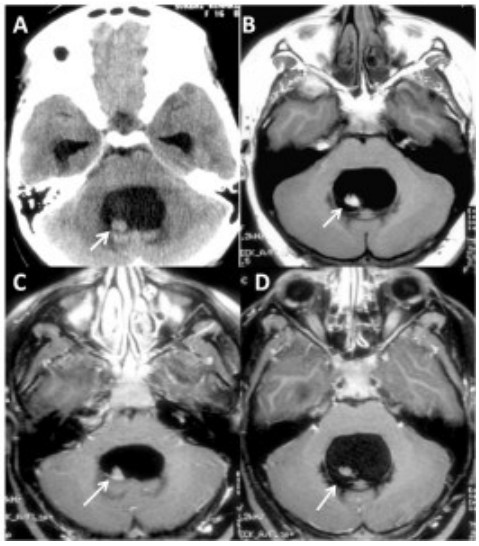
Keyword :
fourth ventricular cysticercal cyst, MRI, surgical options

Introduction:
Neurocysticercosis (NCC) is the most common parasitic infestation of the central nervous system. The pathological entity responsible for various clinical manifestations is the larval cyst of Taenia solium. The invasive oncosphere infects the brain through haematogenous spread after it pierces the intestinal wall. Commonly the oncosphere seeds the brain parenchyma. However, in 7-30% of patients the parasite reaches the ventricles through the choroid plexus, the occipital horn of the lateral ventricle and fourth ventricle being commonly involved due to the effect of gravity and cerebrospinal fluid (CSF) flow pattern. [2,3] Once the cyst reaches a size large enough to occlude the aqueduct or the fourth ventricular outlet, the clinical course may be rapidly progressive and frequently fatal as a result of acute obstructive hydrocephalus. [1] There is no consensus or clinical guidelines regarding the optimal management of fourth ventricular cysticercosis. Various modalities of treatment are reported for the excision of the fourth ventricular cysticercal cyst. We describe and discuss the clinicoradiological profile and management of a patient who presented with fourth ventricular cysticercosis.

Case report:
A sixteen year old girl presented with progressive holocranial headache associated with intermittent blurring of visionand projectile vomiting for 15 days.

She had imbalance while walking for 7 days. She had no other illnesses. General and systemic examination was normal. There were no subcutaneous nodules. Neurological examination revealed bilateral papilledema, signs of bilateral cerebellar dysfunction, gait ataxia and right lateral rectus paresis. Magnetic resonance imaging (MRI) of brain with contrast showed a cystic mass with a non enhancing wall measuring 4 x 3 x 3.5 cm in the fourth ventricle (Figure 1). There was an eccentric nodule in the left posteroinferior part of the cyst which was hyperintense on T1W and T2W images. The sagittal and coronal images (Figure 2) showed the cyst was occupying the whole of fourth ventricle with significant mass effect over the brainstem anteriorly with the hyperintense nodule seen in the inferior aspect of the cyst wall. The cyst caused the obstruction of the foramen of Megendie inferiorly and the aqueduct of Sylvius superiorly causing significant obstructive hydrocephalus resulting in dilatation of all the ventricles with periventricular CSF seepage. Serum enzyme-linked immunoelectro transfer blot (EITB) for cysticercal antibodies was positive. Afteradmission in the ward her headache worsened associated with deterioration in GCS score to 13/15. An emergency right frontal external ventricular drain was inserted, after which she regained normal sensorium and the headache improved. She then underwent midline suboccipital craniectomy and total excision of the fourth ventricular cyst. Intraoperatively the brain was lax and the cyst was obstructing the foramen of Magendie with no CSF flow appreciated. The cyst measured approximately 3 cm in size. It contained clear fluid and was not adherent to the walls of the fourth ventricle. The cyst was grasped and slowly delivered out of the fourth ventricle and excised as a single piece after which there was free flow of CSF into the cisterna magna.

PPreoperative sagittal and coronal



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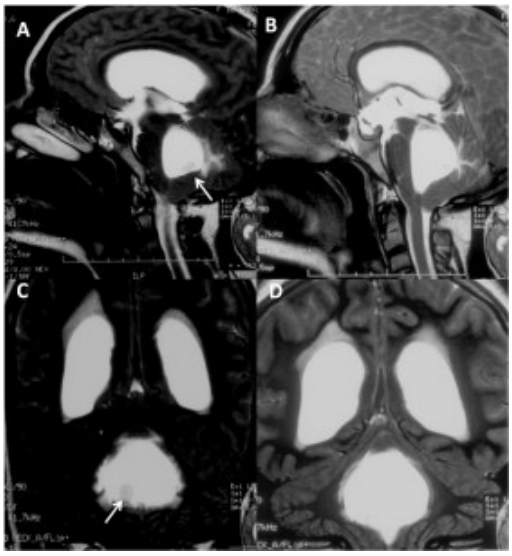
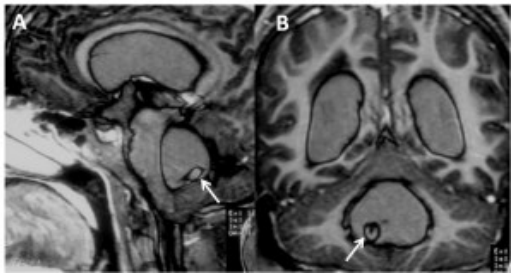
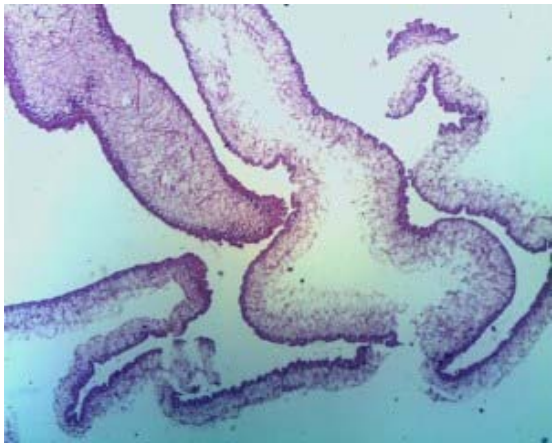
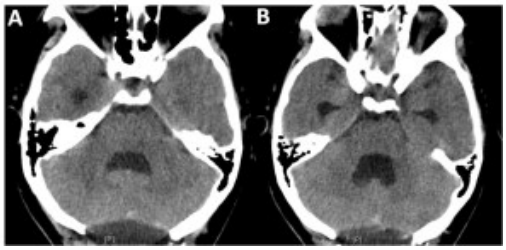






Cysticercal cyst wall

Histopathological examination showed (Figure 4) walls of the cysticercus with microvilli on the tegument and a layer of tegument cells beneath and loose stroma with few excretory channels in the lumen.



posteraive plain CT images

Postoperatively, the external ventricular drain was removed on the second postoperative day. Her headache and vomiting subsided and the signs of cerebellar dysfunction improved over the next two weeks. Postoperative computed tomography (CT) scan of the brain (Figure 2) showed total excision of the cyst with reduction in the size of the ventricles. Shunt was not considered in our case as there was no evidence of ependymal inflammation or scarring both radiologically and intraoperatively. At four month follow-up she was asymptomatic.

The causative role of cyst in causing hydrocephalus was considered in this case. The cyst measured 4cm on imaging with the small hyperintense nodule in the inferior aspect of the cyst wall. The cyst was occupying whole of fourth ventricle with significant mass effect over the brainstem and there was obstruction of the foramen of Megendie and aqueduct of sylvius resulting in obstructive hydrocephalus. The hydrocephalus was caused by the cyst as evident in the imaging and by the intraoperative finding of obstruction of the foramen of Megendie by the cyst wall. There was no evidence of inflammation associated with the cyst and there was reduction in the size of the ventricles in the postoperative images.

Patient improved significantly postoperatively and is doing well. Hence it is unlikely that the hydrocephalus occurred independent of this lesion. Lesionectomy alone was done in this case with total extirpation of the cyst by which patent CSF pathway was established through the foramen of Megendie. The intrafourth ventricular cysticercal cyst is seen in 3% of patients with NCC. [15]

Discussion:

Intraventricular form of infection is a life threatening condition with a rapidly progressive course.

Clinical features:

Presentation of an intraventricular form of NCC is that of symptoms related to obstruction of CSF flow, mass effect and those related to inflammation causing obstruction of CSF flow. [4] Our patient presented with subacute symptoms due to obstruction of CSF flow and mass effect, akin to that reported in patients with fourth ventricular cysticercal cyst. [4,5] Findings include decreased

alertness, papilledema, and focal motor deficits, including corticospinal tract signs and frontal lobe motor apraxia. The latter is the result of compression of frontal lobe long motor tracts in corona radiata and internal capsule that are caused by enlarged lateral ventricles, the ventricular expansion being maximal in the frontal horns. Our patient also had bilateral cerebellar dysfunction with gait ataxia. This is explained by the direct compression of the brainstem and midline cerebellar structures. 6 Abrupt CSF obstruction results in acute hydrocephalus, in which symptoms and signs include headache, diplopia, dizziness, vomiting, restlessness, seizures, respiratory changes, bradycardia, blood pressure elevation, and alteration of consciousness. In cases of intraventricular cysticercosis the sites of obstruction are the foramen of Monro, third ventricle, aqueduct of Sylvius, and fourth ventricle. [1]An asymptomatic fourth ventricular cyst may become symptomatic when the cyst larva dies; resulting in involution and inflammation. [6] The involutional cyst liberates antigenic substances that cause an inflammatory reaction throughout the ventricular system. This granular ependymitis fixes the cyst capsule to the ventricular wall with strong adhesions and fibrosis, resulting in development of hydrocephalus. Patients can present with increased intracranial pressure (ICP), meningoencephalitis with fever, alteration of consciousness, nuchal rigidity, focal neurological deficit, and inflammatory reaction detectable in the CSF. [23] Even with provision of appropriate treatment, mental deterioration, blindness, quadriparesis, and ataxia may occur. Complications include cerebral and brainsteminfarction due to angiitis, hypothalamic dysfunction, infections, repeated shunt failure, arachnoiditis, ependymitis, and ventriculitis. [23]

Imaging

In parenchymal form of NCC, CT imaging is helpful. However, in intraventricular NCC only MR imaging has been proven to be superior in differentiating the cystic masses of fourth ventricle. [7] One of the absolute diagnostic criteria for fourth ventricular cysticercosis is the demonstration of scolex within the cyst. [8] Scolex has a T1 weighted hyperintensity of fat and appears as a mural nodule within the wall of the cyst. In T1 weighted sequence, the cyst wall appears as a thin hyperintensity between the darkness of the cyst content and the CSF. On T2-weighted imaging, the cyst contents are isointense with the surrounding tissues, and the scolex is hyperintense. Cyst wall enhancement presumably indicates the presence of diffuse ependymal inflammation, representing the greater difficulty required to resect these lesions because of arachnoidal and ependymal scarring. [9] Fourth ventricle cysts may appear as neoplasms such as cystic medulloblastoma, astrocytoma, or ependymoma, with CSF obstruction and edema of adjacent brain. [9] Characteristics indicating diagnosis of a tumor over intraventricular cysticercal lesions include absence of T1 weighted hyperintense nodule within the cyst, the presence of contrast enhancement of the tumor, edema in the adjacent tissues, extension up and down the fourth ventricle and laterally into the prepontine cisterns. [9]

Laboratory studies The demonstration of the parasite on histology remains the gold standard. The serological tests to demonstrate cysticercal antibodies like enzyme-linked immunoelectro transfer blot (EITB) and the enzyme linked immunosorbent assay have shown poor sensitivity in patients

with solitary cysticercal lesion *with a positivity rate of only 18% to 49% in serum samples.* [21,11] *In a study of the 50 patients by Wilson et al*[11]*, thirty two patients with two or more lesions, 94% had detectable antibodies by* EITB compared with 28% of 18 patients with single lesions. However, in over 80% of patients with intraventricular cyst with inflammatory features of ependymal enhancement, EITB assays both are positive for NCC in serum and in CSF, regardless of the number or apparent condition of the cysts. [11] Immunodiagnostic tests may yield false positive reactions in tuberculosis, as well as in parasitic diseases such as echinococcosis. [12] Cerebrospinal fluid abnormalities are directly proportional to the degree of local inflammation and ventriculitis. In 50% of cases (with presumptive diagnosis based on imaging features) the CSF cell count, protein, and glucose are normal. [5] In the other 50% a moderate mixed pleocytosis, increased protein, and reduced glucose are seen.***Treatment of fourth ventricular NCC*** *The* treatment of fourth ventricular cysticercal cyst would be aimed at relieving the obstruction of CSF flow. Acute hydrocephalus usually requires an emergency ventriculostomy and subsequent resection of the cysts obstructing CSF flow, particularly those in the fourth ventricle. [5] The various therapeutic options for the management of fourth ventricular cysts are emergency ventriculostomy [4], placement of a ventriculoperitoneal shunt [6], endoscopic or open extirpation of obstructing cysts [5], antihelmintic medications (albendazole and praziquantel) and steroid therapy [15]Fourth ventricular viable cysts should typically undergo extirpation because they may cause brainstem compression even after insertion of a VP shunt. [5] Whether to choose endoscopic or open resection depends on the surgeon's experience; the less invasive endoscopic removal being preferred in most situations. Standard open neurosurgical approach for fourth ventricle cyst is midline suboccipital craniectomy and extirpation as done in our case. [5] Apuzzo et al [1] reported that none of the patients who underwent surgical extirpation and had no evidence of ependymitis required shunt in the mean follow up period of thirty eight months.Endoscopic techniques provide the least traumatic and safest means to excise intraventricular cysts.Different endoscopic techniques have been described for excision of fourth ventricle neurocysticercal cysts. The neuroendoscopic approach confers many advantages like avoidance of shunt placement due to the simultaneous creation of a physiological internal CSF diversion, easy access and navigation through the ventricular system within minutes, detection and excision of multiple intraventricular cysts, and minimal perioperative complications. [22]Despite its many advantages, neuroendoscopy has some limitations. In a patient with severe ependymitis and dense adhesions, endoscopic cyst excision can be difficult and hazardous. Intraventricular bleeding is another potential complication. [22] Involutional inflammatory fourth ventricular cysts are attached to the ventricular wall by strong adhesions and cannot easily be removed without damaging adjacent brain tissue and removal of the cyst does not typically obviate the need for a shunt.[1]Thus, when neuroimaging evidence of ependymitis exists, the preferred treatment is placement of a VP shunt without removal of the cyst. Ventriculoperitoneal shunt obstruction, either by cyst-related gelatinous material or by high CSF protein, is the most common complication. [14] Early shunt revision should be

considered if the patient experiences no improvement after shuntinsertion, and recurrent shunt obstruction by cyst material is another reason for excision of intraventricular cysts. [13]

Medical treatment

Although a recent report have noted success in using antihelmintic drugs without surgery in 33 patients with giant (5-cm-diameter) subarachnoid cysts, the role of primary antihelmintic therapy in fourth ventricular cysticercal cyst remains uncertain. [15] Both therapeutic failures and successes with praziquantel have been demonstrated in the treatment of an intra ventricular NCC. [16] There is an inflammatory reaction similar to that seen with the natural death of the larva when the cyst larvae die following antihelmintic therapy. [17] The scarring and granulomatous ependymitis may lead to acute hydrocephalus

Prognosis

In patients with fourth ventricular NCC who undergo insertion of a ventriculoperitoneal shunt or resection of cysts improvement typically occurs with resolution of the hydrocephalus. [17] Fourth ventricular NCC, however, can be fatal with a mortality rate of 13% in patients with acute hydrocephalus. [18] Because the mass effect of a cyst in the posterior fossa is less well tolerated than in supratentorial sites, the prognosis for patients with fourth ventricular cysts is guarded. Fourth ventricular cysts will also present in the inflammatory state, with edema and adhesion to the adjacent tissue making resection of the entire lesion difficult. Shunt surgery does not always ensure a good prognosis. [20]***Conclusion*** *A cysticercal cyst should be considered in the differential diagnosis of a patient with a fourth ventricular cystic lesion. Early recognition of this condition and its prompt excision can result in good outcomes obviating the need for a permanent CSF diversion procedure.*

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