CASE REPORT

Neurocysticercosis: Giant intraparenchymal disseminated cyst. Clinical report case and literature review

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RESUMEN

La neurocisticercosis es una enfermedad parasitaria que afecta comúnmente al sistema nervioso central humano, es la principal causa de epilepsia adquirida y un problema de salud pública. Las formas son neurocisticercosis parenquimatosa, subaracnoidea, ventricular, medular y mixta. Las etapas que atraviesan los cisticercos incluyen: fase vesicular, fase coloidal, granular y la etapa calcificada. El objetivo de este caso es mostrar un quiste intraparenquimatoso diseminado gigante causado por la neurocisticercosis. El quiste intraparenquimatoso gigante o fase vesicular con la presencia de escolex dentro y fase coloidal son una presentación rara y poco frecuente. Los estudios de imagen e histopatológicos son los exámenes que nos dan el patrón de diagnóstico. La enfermedad tiene una buena evolución si se establece un diagnóstico oportuno y se lleva a cabo un tratamiento adecuado.

Palabras clave. Sistema nervioso central. Enfermedades parasitarias. Cisticercosis.

ABSTRACT

Neurocysticercosis is a parasitic disease which more common affection is the human central nervous system being the leading cause of acquired epilepsy and a public health problem. The neurocysticercosis forms are parenchymal, subarachnoid, ventricular, spinal and mixed. The stages that cross cysticerci to destruction include: vesicular stage, colloidal stage, granular and calcified stage. The aim of this case report was to show a giant intraparenchymal disseminated cyst caused by neurocysticercosis. The giant intraparenchymal cyst or vesicular stage with presence of scolex inside and colloidal stage that is a mixed presentation of neurocysticercosis, being rare and infrequent presentation, also imaging studies (cranial CT and MRI) and study histopathological are the exams that give us the diagnosis pattern, and which is confirmed by direct visualization; these have a good evolution if a proper diagnosis is established and carried out appropriate treatment.

Key words. *Central nervous system. Parasitic diseases. Cysticercosis.*

INTRODUCTION

Neurocysticercosis (NCC) is the most common parasitic disease of central nervous system (CNS) being the leading cause of acquired epilepsy and a public health problem.[1] Cysticercosis is endemic in Latin America, Sub-Saharan Africa and some regions of Asia including the Indian subcontinent, China, Korea and Indonesia. Approximately 2.5 million people in the world carry the adult *Taenia solium* and many more are infected with cysticercosis, the incidental finding of calcified granulomas are found in 10-20% of the general population in endemic areas, with an estimated 75 million Latin America people at risk of NCC, 400,000 of whom are symptomatic.[1,3] The World Health Organization (WHO) has reported that more than 50,000 deaths occur every year as a result of NCC.[1-4] *Taenia solium* is a zoonotic tapeworm whose definitive host is the human, harboring the adult form in the intestine, and whose common intermediate host is the pig, harboring the larval form or cysticercosis. The adult worm (intestinal tapeworm) develops after ingestion of infected pork with cysticerci and undercooked.[1-5]

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The types of NCC are parenchymal, subarachnoid, ventricular, spinal and mixed or disseminated.[1-6]

The stages that cross cysticerci to destruction include: vesicular stage, colloidal stage, granular stage (cellulose forms) and calcified stage.[1-6,8] The clinical manifestations of NCC ranging from asymptomatic or symptomatic and depend on the number, location, size and stage of the parasite, as well as host response degree. NCC can often affect the parenchyma, subarachnoid space, intraventricular system, ocular and spinal diseases can occur but are less common.[1-7] Clinically, it may present as epilepsy (70% of cases) that can be generalized or partial as the result of perylesional inflammation of the cysts, recurring as much as 50-70% of cases; altered consciousness, loss of vision or disturbances, headache, vomiting, papilledema, signs of meningeal irritation (< 2% of cases), arachnoiditis, insane confusing manifestations. There are 5 syndromes presentation of NCC [seizure, focal (medullary, meningeal or optoquiasmatic), psychic, intracranial hypertension, pituitary-diencephalic].[1-8] The diagnosis is based on diagnostic criteria and accuracy degree diagnostic of NCC (modified Del Brutto, *et al.)*.[4] Treatment of NCC includes cysticidal drugs, symptomatic measures and surgery. Since the presentation of this disease is not possible to standardize a single scheme treatment for all cases, this will depend on the number, location and viability of the parasites in the nervous system.[7,8] The aim of this case report was to show a giant intraparenchymal disseminated cyst caused by NCC.

CASE REPORT

A 42-year-old man, low socioeconomic status and poor hygienic-dietary habits, without any history relevant to the condition. He began his condition on May 21, 2013, when in a discussion of your company, presented focal seizure of the left arm, later became general; according to comments from colleagues describe them as tonic-clonic and retroversion look, without sphincter relaxation. Upon arrival at the service, he was completely asymptomatic, without motor deficit, the fundus was normal, the pupils were isochoric, abscense of facial paralysis, right side palmo-mental reflex positive, hyperreflexia of the left limbs, no sensory deficit, strength muscle is preserved and was symmetrical.

Admission vital signs

Blood pressure: 100/70 mmHg, heart rate: 80 beats per minute, respiratory rate: 18 per minute, temperature: 36.0°C. Laboratory tests on admission: Leukocytosis 15,200/UL with 8,500/UL neutrofilia. Single and contrasted cranial computed tomography (CCT) scan showed hypodense right frontal zone suggestive cystic process, very thin walls, no perilesional edema, mass effect to collapse the ventricle and divert lateral sulcus (Sylvian fissure); inside cystic cavity a hypodense area is observed corresponding to a parasitic scolex (Figure 1). Cerebrospinal fluid shows a predominance of mononuclear cells: 95%.

While staying, he progressing well without presence of seizures and anti-seizure scheme began with diphenylhydantoin (DFH) sodium 125 mg/8 h IV, Albendazole 400 mg/8 h (15 mg/kg/day) VO and praziquantel 50 mg/kg/day.

Later cranial NMR is made showing the contour cyst enhancement with contrast and the presence of other injuries inside the one is observed (Figure 2).

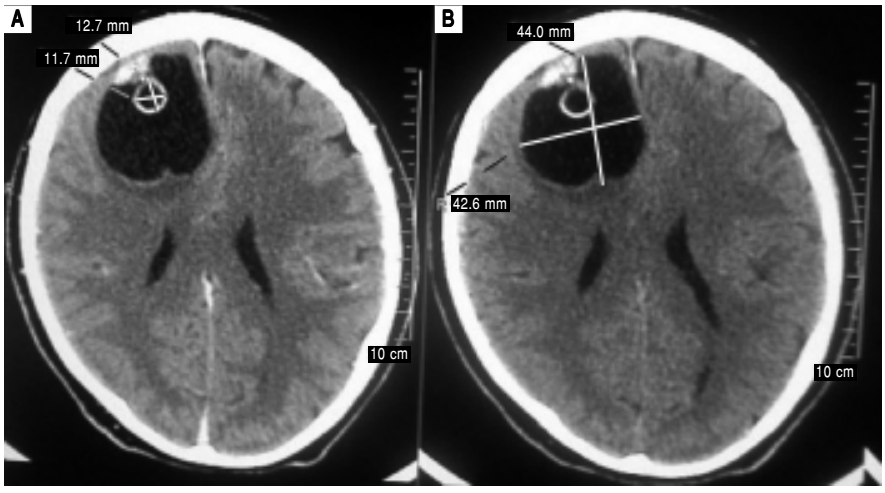

Figure 1. Cranial TC. Observe images 1 and 2 hypodense area suggesting a cystic process approximately 44.0 mm x 42.6 mm and intracystic scolex accompanied a colloid stage approximately 12.7 mm x 11.7 mm in the right frontal region.

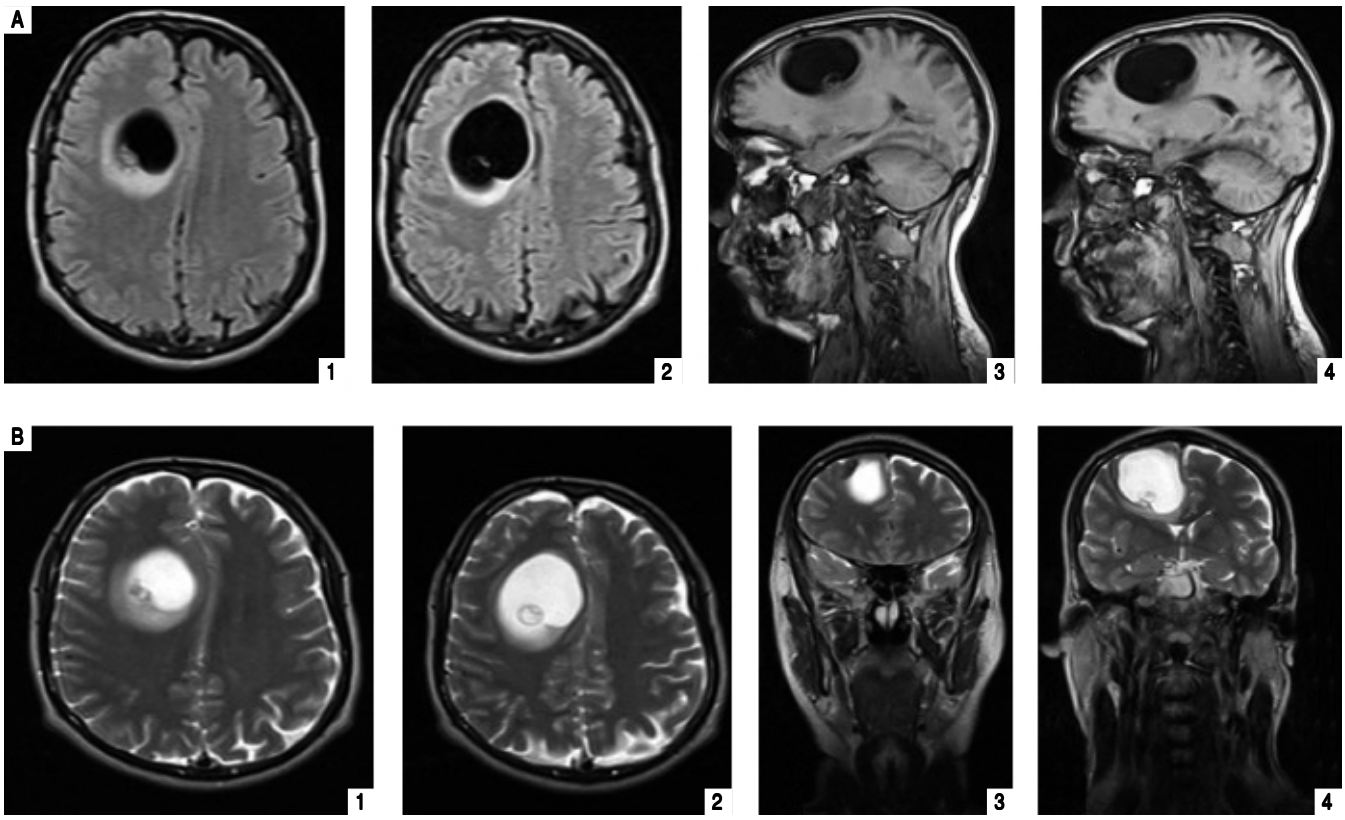

Figure 2. A. On top contrasted MRI T1 is observed, 1 and 2 axial images, 3 and 4 sagittal images: Note the contour cyst enhancement with contrast and the presence of other injurie inside. **B.** At the bottom MRI T2 is observed, 1 and 2 axial images, 3 and 4 coronal images. Hyperintense lesion with low perilesional edema and inside other cystic lesion.

Surgical (craniotomy) intervention was decided which took place without any complications, it was dissected and drained the contents of the capsule or vesicle, previous resection of the capsule or vesicle, intracystic material is extracted (scolex and other colloidal stadium cisticerci), which is sent to pathology (Figure 3).

After surgery he is admitted to the ICU service evolving favorably. Histopathological examination reported cysticerci vesicular stage, that inside had a colloidal stage of the parasite (Figure 4).

Another simple cranial CT is performed without evidence of cerebral edema in the midline unchanged in the right frontal zone, with post-surgery changes, image properly of cyst resection.

DISCUSSION

NCC is a parasitic disease that is acquired from eating pork infected with cysticerci and undercooked. Which makes us think of the high probability that the patient may have acquired Taenia because of his bad hygienic-dietary habits, and mainly unnoticed for a long time due to the pathophysiology of the parasite and the variability in the clinical presentation.[1-6]

Upon arrival, brought up the debut of single seizure (tonic-clonic seizure) right side palmo-mental reflex positive and left limbs hyperreflexia associated with predominantly neutrophilic leukocytosis, reason for which anti-seizure management started, steroid and cysticidal. Considering a late onset of seizures, simple cranial TAC is determined to take, which is one of the major methods for diagnosing neurocysticercosis as MRI.[1-6,9,10]

Due the presence of intracerebral giant cyst is decided to implement extractional craniectomy of giant intraparenchymal cyst approximately 44.0 mm x 42.6 mm and intracystic scolex, presence of a colloid stadium approximately 12.7 mm x 11.7 mm, which correlated a mixed form of cysticercus development, being a rare and uncommon presentation that is confirmed through microscopic and macroscopic exam, allowing the scolex identification

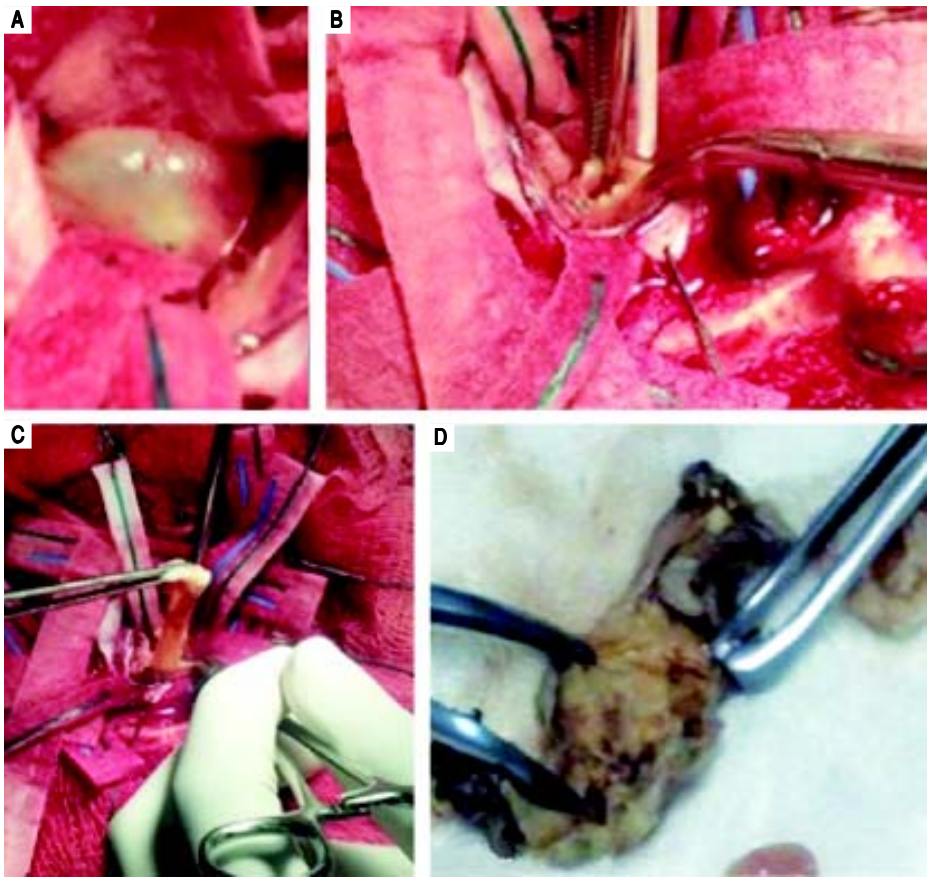

Figure 3. A. Shows thin, friable, transparent wall with vesicular stage; the cleavage site **(B, C)** is observed and removal of colloidal stage inside vesicular cyst, prior aspirated transparent liquid, **(D)** presents vesicular stage wall and colloidal form of neurocysticerci.

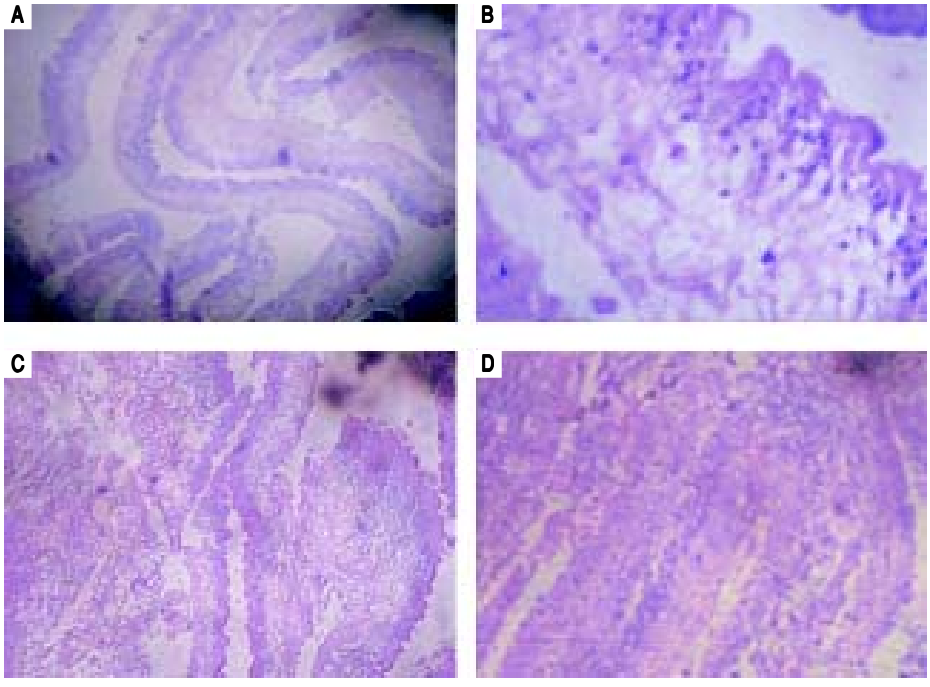

Figure 4. Tissue samples with hematoxylin-eosin staining. **A-B.** Cysticercus membrane is observed, hyalinization stage. Note the cuticle vanish trend that appears flattened in most cutting and transparency of the inner layers. Hyalinized and collagenous cisticerci membrane with granulomatous inflammatory and infiltrated adjacent reaction; cutting is typical of a vesicular cysticerci phase. **C-D.** Presence of connective tissue associated with cellular infiltration.

or its remains, and cause low perilesional inflammation in nervous system.[1,4-6,8]

Parenchymal neurocysticercosis is the most common form. This may manifest as a single lesion or as a massive parasitic infection, neurocysticercosis in the form of granular stage is the most frequent neurocysticercosis presentation, parenchymal cysticercosis can be large or small and deep or shallow, preferably being located in cerebral cortex and basal ganglia due the high vascularity of these areas. In literature is described isolated and rare cases of giant intraparenchymal neurocysticercosis, which consists a solitary cystic lesion larger than 4 cm, with septa and mural nodule inside the cyst. Currently literature there is a case report presenting a giant intraparenchymal neurocysticercosis cyst, mixed type, so we could be talking about of a first report of such case. Usually this injury requires surgery and should be distinguished from hydatid cyst, cystic necrotic tumor and porencephalic cyst.[5,9,10] Subarachnoid cysticerci may be small if is located in depth cortical sulci or can reach sizes greater than 50 mm if they are in CSF cisterns (cerebroespinal fluid).

Ventricular cysticerci can be small or large, they are located preferably in the fourth ventricle; these parasites can be found attached to the ependymal or floating freely in the ventricular chambers. Spinal cysticerci are located in the subarachnoid space or into the spinal parenchyma and its gross appearance is similar to cysts located in the brain.[1-5]

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The stages that cross cysticerci to destruction include: vesicular stage, colloidal stage, granular stage (cellulose forms) and calcified stage; each with its own histological, imaging and morphological characteristics.

Vesicular cyst is characterized by large, oval vesicle, with a translucent vesicular fluid, generally scolex it is not visible, although in most cases, a meticulous macroscopic review allows identification of the scolex or their remains, causing perilesional inflammation in nervous system and brain imaging findings are: cystic lesions without edema, no abnormal contrast enhancement; colloidal phase is characterized by cellular inflammatory reaction around the cyst, larva dies, begins to degenerate, and scolex disintegrates, the liquid inside becomes shady and the capsule becomes thick; as it was presented in this case; nodular stage or granular stage presents a cyst even more retracted and forms a granulomatous nodule that subsequently is going to calcify. Nodular stage is focal type, with well-defined borders without perilesional inflammatory reaction.[1-8]

According to histopathology reports was a giant cyst intraparenchymal vesicular type with one colloidal type cyst inside, founding the degenerated/destructed scolex causing an inflammatory reaction, which assumes that gave the pattern for the seizure presented.[5] The diagnosis is based on the criteria diagnosis and accuracy diagnostic degree of neurocysticercosis (Amended Del Brutto, *et al.)*.[4]

Our patient presented 2 absolute criteria, 3 major criteria, 2 minor criteria and 1 epidemiological criteria.[4]

Currently the choice immunoassay is EITB using partially purified antigen extract with 100% specificity and 94-98% sensitivity in patients with 2 or more cystic lesions; antibody detection by ELISA using CSF anticysticercus (in all patients with new onset of seizures or neurological deficits by neuroimaging) with sensitivity of 87% and 95% specificity.[2,3-6,10]

Drug treatment based on antiparasitic treatment, symptomatic measures and surgery was appropriate to this case, because of the variability of disease presentation is not possible to standardize a single treatment regimen for all cases. Depends on the number, location and viability of the parasites in the nervous system.[7,8]

Inactive NCC (calcifications) does not require antiparasitic treatment, but could help corticosteroids use in controlling some discomfort. Hydrocephalus secondary NCC can be tributary of ventricular peritoneal shunt valve (VPSV). Parenchymal active NCC usually requires treatment with albendazole or praziquantel. In general, it is preferred albendazole because it has more available and is inexpensive, penetrates better subarachnoid cysts and has not drug interactions with corticosteroids and anticonvulsants. Dose: 15 mg/kg/day for 7 to 15 days. It is best absorbed with fatty foods intake.[1,2,8,9]

Surgical therapy role in management of neurocysticercosis is limited to ventricular cysts neuroendoscopic resection, intraparenchymal giant cysts have confirmed that open surgery is the best choice.[3-4,9,10]

Differential diagnosis are necrotic hydatid cyst, cystic tumor and porencephalic cyst, brain abscess, basilar artery thrombosis, brain stem gliomas, craniopharyngioma, glioblastoma (GBM), intracranial epidural abscess, meningioma, neurosarcoidosis, oligodendroglioma, tuberculous meningitis.[9-10]

In conclusion, intraparenchymatous giant cyst or vesicular stage with presence of scolex inside and colloidal stage is a mixed presentation of NCC that according to the review literature, is the first case reported being rare and infrequent presentation, also imaging studies (cranial CT and MRI) and histopathology, are the exams that give us diagnosis pattern, which is confirmed by direct visualization; these have a good evolution if a proper diagnosis is established and carried out an appropriate treatment.

REFERENCES

1. Garcia HH, Del Brutto OH. Neurocysticercosis: updated concepts about an old disease.*Lancet Neurol* 2005; 4: 653-61.
2. Enseñat J, Martínez-Mañas R, Horcajada J P, De Juan C, Ferrer E. Dificultades diagnósticas y terapéuticas en la neurocisticercosis: presentación de 6 casos y revisión de la literatura. *Neurocirugía* 2007; 18: 101-10.
3. Saavedra H, Gonzalez I, Alvarado MA, Vargas V, Martinez M. Diagnóstico y manejo de la neurocisticercosis en el Perú. *Rev Peru Med Exp Salud Publica* 2010; 27(4): 586-91.
4. Del Brutto OH. Neurocisticercosis: actualización en diagnóstico y tratamiento, *Neurología* 2005; 20(8): 412-8.
5 Agarwal PP, Gaikwad SB, Garg A, Gupta V, Mishra NK, Mehta VS. Giant intraparenchymal neurocysticercosis: Unusual MRI findings. *Neurol India* 2004; 52(2): 259-60.
6. Alarcon F. Neurocysticercosis: it's an etiopathogenesis, clinical manifestations, diagnosis and treatment. *Rev Neurol* 2006; 43(Suppl. 1): s93-s100.
7. García HH, Pretell EJ, Gilman RH, Martínez SM, Moulton LH, Del Brutto OH, et al. A trial of antiparasitic treatment to reduce the rates of seizures due to cerebral cysticercosis. *N Engl J Med* 2004; 350: 249-58.
8. Odermatt P, Preux PM, Druet-Cabanac M. Treatment of neurocysticercosis: a randomized controlled trial. *J Neurol Neurosurg Psychiatry* 2008; 79(9): 978.
9. Umredkar A, Singla N, Mohindra S, Bal A, Gupta SK. Giant intraparenchymal neurocysticercosis: Report of surgical aspects two cases. *Neurology India* 2009; 57(6): 800-2.
10. Ramesh VG, Parthiban A. Giant parenchymal cysticercosis with unusual imaging features. *J Clin Neurosci* 2008; 15: 1404-6.