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Racemose Neurocysticercosis: Medical and Surgical Treatment

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Abstract

Introduction: Neurocysticercosis is a parasitic infection of the central nervous system. The racemose variant tents to invade the subarachnoid space and it is considered malignant; It has increased morbidity and mortality and often shows a decreased response to medical treatment alone.

Case Reports: A 23yearold male farmer with a 1 month of headache, seizures and mild language impairment. Imaging and serology confirmed racemose neurocysticercosis and responded well to medical treatment. A 48-year-old indigenous woman with long history of headache, seizures, worsening right hemiparesis, confusion, and incoherent speech. Imaging was compatible with racemose neurocysticercosis but did not respond to medical treatment. Due to worsening symptoms and lesions growth, surgery was performed. She had motor function improvement, but language and cognitive function remained altered.

Conclusion: In Racemose Neurocysticercosis, due to its fatal complications, medical treatment alone may have limited effect; an early surgical treatment can be a valid consideration in selected patients. However, previous neurological status may influence outcome.

Keywords: Neurocysticercosis; Racemose Neurocysticercosis; Surgical Treatment; Medical Treatment; Adult

Abbreviations

NCC: Neurocysticercosis; CNS: Central Nervous System; *T. solium: Taenia solium*; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; CSF: Cerebrospinal Fluid; AEDs: Anti-Epileptic Drugs; SCG: Solitary Cysticercus Granuloma; DWI: Diffusion Weighted Imaging; VP: ventriculo-Peritoneal

Introduction

Neurocysticercosis (NCC) is a parasitic infection of the central nervous system (CNS) caused by the *Taenia solium* (*T. solium*) [1]. The disease is endemic in Central and South America, Asia, and Africa [2].

The International Task Force for Disease Eradication of the World Health Organization declared *T. solium* potentially eradicable in 1992. However, NCC is still considered as the major cause of acquired epilepsy in developing countries (30%) [3]. *T. solium* is not only the cause of serious zoonotic disease, but also causes great burden due to health care costs, disability, and economic losses [3,4].

Racemose NCC refers to a severe variant of extraparenchymal NCC (intraventricular, subarachnoid space and cisterns) presenting as multiple cystic multilocular membranes (cluster of grapes).

Extraparenchymal location allows the cysts to grow to a large size (4 - 12 cm) causing mass effect and they usually lack scolex, contrast-enhancement or edema Spinal and ocular location have also been reported. [5]. Racemose NCC is associated with increased morbidity and mortality, as well as a decreased response to medical treatment [1,6,7].

The diagnosis is often a challenge. It can be confused with tumors or other infections [1,8]. Neuroimaging supported by serology, clinical findings, and epidemiological context play a fundamental role in the diagnosis and monitoring of this disease [1,7,9]; however, epidemiology is often scarce or not always considered [3].

The treatment is complex as there are no standardized guidelines [8]. The usual management involves long courses of antiparasitic drugs with corticosteroids and anticonvulsants. However, if big multiple growing cysts are present, medical treatment alone may not be satisfactory and surgical intervention should be considered [7,10-12].

Case Reports

Case 1: A 23-year-old male farmer from the amazon region presented to the emergency room with a complaint of 1 month of worsening headaches and seizures. His neurologic exam showed mild language impairment.

A head computed tomography (CT) scan demonstrated a multilobulated hypodense mass in the left Sylvian fissure and surrounding arachnoid space and parenchymal infiltration with edema. A primary tumor was suspected, and corticoids were started. Subsequent magnetic resonance imaging (MRI) was performed which demonstrated the presence of non-enhancing, cystic lesions highly suspicious of Racemose NCC (Figure 1 and 2). The electroencephalogram reported several outbreaks of paroxysmal epileptiform activity in a left frontotemporal focus.

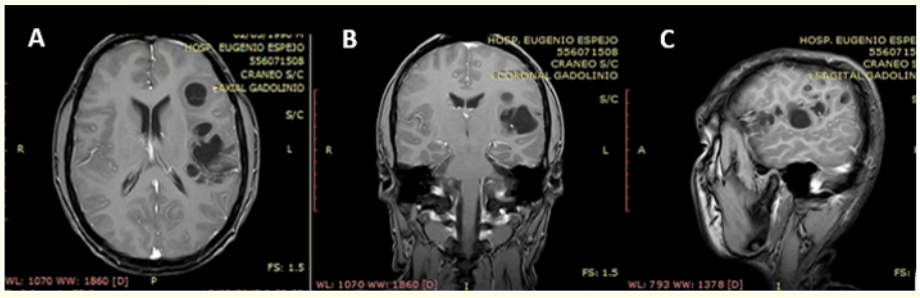

Figure 1: Brain contrast-enhanced MRI. A: Axial view; B: Coronal view; C: Sagittal view.

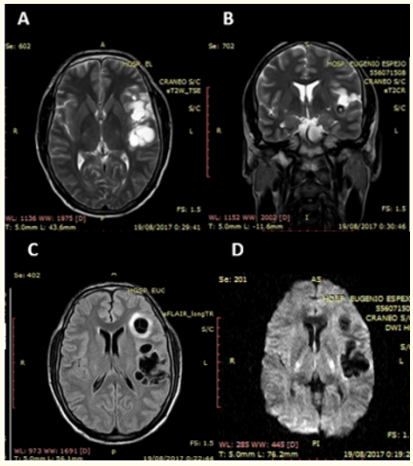

Figure 2: Brain MRI. A: T2, axial view; B T2, coronal view; C: FLAIR, axial view; D: DWI, axial view.

Due to epidemiology analysis (sex, age, occupation, geographical region), imaging findings, and clinical setting, this case was considered highly suspicious of racemose NCC, and medical treatment was established using a long course of albendazole, corticoids, and antiepileptic drugs with good response at 6 months follow up. The diagnosis was confirmed by the Polymerase Chain Reaction test in cerebrospinal fluid (CSF). At 6 month follow-up, the patient reported headache and seizure control and language improvement.

Case 2: A 48-year-old indigenous woman from the Andean region, presented to the emergency department with a 6-month history of severe headache and seizures. Neurological examination showed right hemiparesis (4/5), confusion and incoherent speech.

Brain MRI revealed multiple conglomerate cystic lesions of different sizes in the left fronto-temporo-parietal region compromising the Sylvian cistern and causing midline shift (Figure 3).

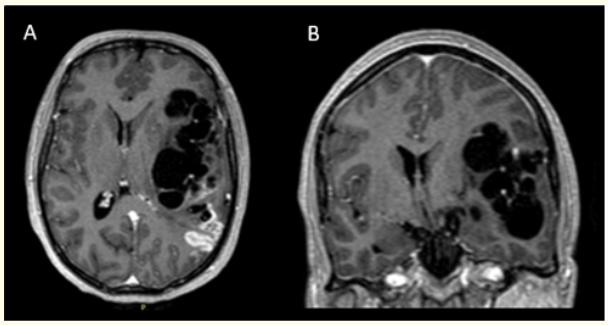

Figure 3: Contrast MRI. A: Axial view; B: Coronal view.

After epidemiological consideration, imaging findings, and patient's symptoms, racemose NCC was diagnosed and medical treatment with albendazole 400 mg every 12 hours for 30 days, antiepileptic drugs, and an intravenous corticosteroid was started during hospitalization. Electroencephalogram revealed an abnormal Theta Delta activity in the left fronto-temporal region. The patient was discharged after 15 days showing seizure control and better motor and language function.

However, after 20 days, the patient was admitted again due to seizures, worsening right hemiparesis (2/5), and dysphasia. New brain contrast-MRI confirmed medical treatment failure and worse midline shift as a result of increased number and volume of cysts (Figure 4).

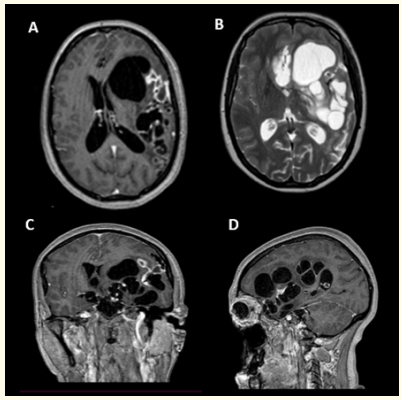

Figure 4: Brain MRI. A: contrast T1, axial view; B: T2, axial view. C: Contrast T1, coronal view; D: Contrast T1, sagittal view.

Medical staff considered surgery as an option to alleviate ICP by removing the cysts via left pterional approach with no major complications. Several dull, translucent whitish membranes were resected; the histopathology confirmed the diagnosis. Immediate post-op CT scan showed complete lesions removal, small pneumocephalus, and no bleeding in the surgical site (Figure 5).

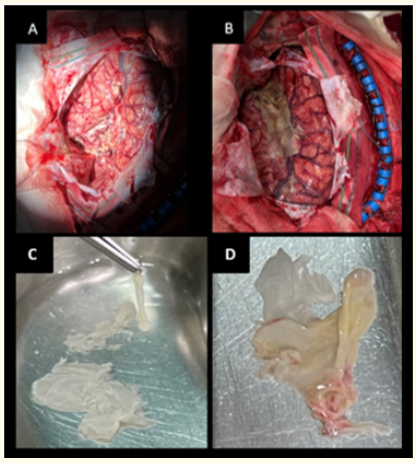

Figure 5: Surgical treatment. A, B: Left pterional approach showing Sylvian fissure. C, D: Cyst membranes confirming diagnose.

The patient continued medical treatment (albendazole, corticoids, AEDs) and showed motor function improvement and seizure control. Unluckily the chronicity of the disease caused severe and irreversible brain damage so, language and cognitive functions remained impaired.

Discussion

NCC is a common intracranial parasitic disease in developing countries and represents one-third of reported seizures and epilepsy cases [3,6,13]. At least 18 countries in Latin America have NCC; it is still considered a major public health problem [3,8]. The author's home country, Ecuador, is endemic, especially in the Andean region where NCC represents 10% of the epilepsy cases, and 25% of those attributed to an identifiable isolated seizure [3,4]. In 2013, the Ecuadorian Ministry of Public Health highlighted 67 new cases of NCC (0.42 cases per 100,000 people). Most of these cases correspond to hospitalized patients in the public health system, which suggests a great underestimation of this pathology [3]. The

racemose form of NCC is rare and has been found in 3.5% of NCC patients [12].

NCC generally invades the brain parenchyma (60 to 90%), subarachnoid space and ventricles. The intracranial complications and clinical manifestations vary from person to person [6,13].

The standardized treatment is still controversial and so far, medical treatment is the primary choice. The data regarding the surgical management (especially for the extraparenchymal cases) is non-conclusive [13,14].

Clinical setting

The clinical presentation of NCC is heterogeneous as it depends on different factors: the number of lesions, location, lesion stage, and the host immune response [6,11,15]. It can range from asymptomatic to serious neurological complications such as seizures or epilepsy (79%), headache (40%), focal neurological deficit (16%), elevated intracranial pressure (ICP) (12%), and ataxia, visual disturbances, cranial nerve palsy, meningeal symptoms, and mental status alterations (less than 10%) [8,12,16]. In addition, brainstem dysfunction, cerebellar ataxia, sensory deficits, involuntary movements, stroke, extrapyramidalism, dementia, and other rare clinical manifestations such as intrasellar cysticercosis and pseudotumor cerebri have been reported [16].

Racemose NCC generally comprises meninges, ventricles, or subarachnoid space. The oncospheres reach the ventricles through the choroid plexus; ventricular and meningeal forms may develop elevated ICP due to meningitis, adhesions, or ependyma inflammation (parasite death) causing hydrocephalus [6,17].

Cerebrovascular disease due to arteritis (cerebral infarction, transient ischemic attacks, and cerebral hemorrhage) have been described. One series showed 54% of arteritis in angiography (the middle and posterior cerebral arteries were the most affected) [6].

The intraventricular or cisternal location allows an exaggerated cystic growth as seen in our cases [6,12]. These forms are considered malignant, and when there is hydrocephalus secondary to meningitis, the mortality can reach to 50% at 2 years after CSF diversion [6,15]. These forms require rapid and aggressive intervention given their rapidly progressive course and are often associated with a poor prognosis. It can occur in conjunction with the parenchymal form or in isolation [6,16].

Diagnostic

The clinical recognition of this disease is complicated, and neuroimaging continues to be fundamental; the arrival of new imaging techniques and subsequent confirmation by serology has made possible an earlier and more accurate diagnosis [3,6,17].

Very often, the diagnosis of racemose NCC is not clear on routine MRI and can be confused with tumor lesions as in our first case which can delay treatment onset. Diffusion weighted imaging (DWI) is necessary to distinguish these cysts from the more common epidermoid cysts for example [7].

Due to the different clinical features, the variability in the interpretation of immunological tests, and the unforeseen findings in neuroimaging, Del Bruto., *et al*. reviewed the clinical, immunological, imaging, and epidemiological criteria for the diagnosis of NCC and proposed two degrees of diagnostic certainty: definitive and probable [8,11,17].

Furthermore, the pathophysiological classification of NCC into active, transitional, and inactive forms allows to correlate clinical manifestations and imaging which may facilitate medical and surgical management [9].

Treatment

The usual management of NCC involves long drug courses. However, if massive amounts of cysts are present, medical treatment alone may not be sufficient and surgical intervention can be required [6,10,12]. Nevertheless, when lesions have caused severe and irreversible brain damage, neurological outcome improvement after surgery may be limited as shown in our second case.

Medical treatment

In most patients, symptomatic therapy alone is indicated. This usually consists of analgesics, antiepileptic drugs (AEDs), and steroids. AEDs may be stopped after the resolution of a solitary cysticercus granuloma (SCG) with a low risk of seizure recurrence. However, in patients with calcific or multiple granulomas, AEDs are used for several years due to a high risk of recurrent seizures. Steroids are usually used for brief periods of one or two weeks, especially in the first week of cysticidal drugs [7].

Cysticidal drugs (praziquantel and albendazole) are known to

destroy live larval cysts whether in the parenchyma, ventricles or the subarachnoid spaces and possibly speed up the resolution of granulomas; In our first reported case, patient showed good response to medical treatment alone and good neurological outcome at follow-up [6,7,11]. However, this drug therapy is debatable and there is no agreement on its indication or evidence for the ideal duration [7].

Compared to praziquantel, albendazole is the preferred drug in racemose NCC due to its CSF penetration, lower cost, side effects, and the ability to be used with steroids [1,7].

There has been controversy as to whether these drugs modify the natural history of NCC: a) cysts usually die within a short time and treatment may be unnecessary, b) scarring due to inflammation after medication could worsen the long-term prognosis of seizures and c) increased inflammation due to cysts death [6,11].

However, some authors as Proaño., *et al*. reported that intensive pharmacological treatment can be effective in neurocysticercosis with giant cysts and that surgery may be necessary only when there is a risk of death due to acute obstruction [18].

Surgical treatment

Surgery is recommended when the diagnosis is uncertain. Additionally, a group of international experts in cysticercosis published a consensus regarding the NCC surgical approach in the following situations [6,7,11,12,19]:

1. Extraparenchymal NCC, intraventricular cysts, hydrocephalus due to racemose cysts, or hydrocephalus due to ependymitis: patients with focal neurological deficits and raised ICP due to large parenchymal cysts; in such situations, surgery might reverse the deficits and reduce ICP.

2. Spinal cysticercosis (intra-extramedullary).

3. Large parenchymal colloidal cyst or subarachnoid racemose cyst causing mass effect: large cysts can exert pressure on the adjacent neurological structures.

4. To confirm the diagnosis: often, the NCC diagnosis is not secured without a biopsy as the imaging can mimic an abscess or tumor.

5. Refractory epilepsy: epileptogenic scar around a cortical cysticercal lesion (inflammation from a degenerating cyst).

Craniotomy and excision of the cyst frequently lead to a good outcome. The cysts are often easily removed but, in some cases, they can be densely adhered to neurovascular structures making surgery more challenging [6,7,19,20]. For intraventricular cysts, the endoscopic approach is preferred as it is minimally invasive. For incompletely excised cysts, continuing medical treatment with steroids and albendazole is recommended [6,7,21].

Surgical complications for NCC are like those for any other intracranial pathology. Though endoscopic surgery is generally safe and complications (intraventricular hemorrhage, fornix damage, CSF leak, seizures, and meningitis) occur in less than 5% of patients [7].

The outcome following surgery may depend on the form of the disease for which the surgery was performed, chronicity, and the previous neurological status. For intraventricular cysts, large parenchymal cysts, large racemose cysts causing mass effect and for biopsy or excision of a SCG, the outcome is generally excellent with over 95% of patients making an uneventful recovery. Some series have found that 75% of patients who underwent surgery had improved at 3-year follow-up. However, shunt surgery for hydrocephalus (cysticercotic meningitis) is complicated by frequent shunt revisions (blockages or infection) in approximately 68% of patients; mortality of 50% on long-term follow up has been reported [6,7].

Conclusion

The treatment of this parasitosis is very complex since there are no standardized guidelines; However, the authors agree there are three fundamental pillars: symptomatic treatment, pharmacological treatment, and surgical treatment [7,20]. Every patient should be individualized depending on the location of the lesions, the stage of evolution of the parasite and the clinical setting (clinical response to primary treatment and previous neurological status) as the authors decided with the presented cases [7,19,22,23].

Surgery is infrequently used in the management of patients with NCC [7] but some studies have reported better results when pharmacological and surgical treatment are combined [24,25]. Though, the authors suggest that timing till surgery or medical treatment, may influence the outcome.

For parenchymal cysticercosis, the surgical goal is to remove the lesion, treat the medically intractable epilepsy and establish the diagnosis to establish subsequent treatment.

In extraparenchymal NCC and its fatal complications (hydrocephalus, arachnoiditis, ependymitis), medical treatment has risks and surgical treatment is an option to consider. For subarachnoid cysticercosis with a higher failure rate of a ventriculo-peritoneal (VP) shunt alone, surgery can help to remove the cyst and relieve the hydrocephalus at the same time. In intraventricular NCC associated with hydrocephalus, the effect of a VP shunt by itself seems unsatisfactory, and craniotomy for cyst resection is necessary [13,14,28].

Conflict of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

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Volume 2 Issue 8 August 2021