

Refractory Epilepsy In Neurocysticercosis

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Citation

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Abstract

The first consecutive 100 patients presenting uncontrolled epilepsy attending to epilepsy and neurocysticercosis clinic at Nelson Mandela Academic Hospital were selected for this study. CT scan and EEG test for all patients were performed. Schizencephaly, arachnoids cyst and racemose cysticercosis were identified as etiology of refractory epilepsy in eleven patients. The authors suggest considering those processes in the differential diagnosis of refractory epilepsy in HIV/AIDS patients

INTRODUCTION

Refractory epilepsy is not a common problem in patients with neurocysticercosis (NCC) therefore when epileptic patients do not respond well to the first line anti-epileptic drugs other causes of epilepsy apart from NCC should be investigated. In absent of ischemic stroke NCC related and other intracranial infections, cystic lesions should be included in the list of differential diagnosis.

NCC is a parasitic disease of central nervous system (CNS) caused by the larval stage (*Cysticercus cellulosae*) of the pig tapeworm *Taenia solium*. This is the most common helminthes to produce CNS infection in human being. The occurrence of acquired epilepsy or the syndrome of raised intracranial pressure in a person living in or visiting a region where taeniasis is endemic or even in one living in close contact with people who have taeniasis should suggest a diagnosis of cysticercosis; the NCC may remain a symptomatic for months to years and commonly a diagnosis is made incidentally when neuroimaging is performed^{1,2}. Active NCC is characterized by intraparenchymal cystic lesions which usually measure between 4 to 20 mm and is found on both cerebral hemisphere however isolate giant cystic lesions may be difficult to differentiate from other causes. Symptoms and signs are related both to the parasite which can show a different biological behavior from one place to another and to the inflammatory-immunological response of the host. Most patients under phenytoin or carbamazepine for a proper control of their seizures, responding very well^{1,2,3,4,5}.

Diagnostic criteria for NCC have been well-established,

based on these studies, categories of Absolute criteria (patonogmonic) is acceptable when the histological demonstration of the parasite from biopsy of the brain or spinal cord lesion is made, or cystic lesion showing the scolex on CT or MRI, or when sub retinal parasites can be visualized by fundoscopy examination; however in places where CT scan is not available, plain X rays of muscular tissues in the limbs showing "cigar shape" calcifications or plain skull X rays with intracranial calcifications (between 1 to 10 mm of diameter) can be useful to confirm the diagnosis. According to the International League Against Epilepsy (ILAE), cysticercosis is the most common cause of acquired epilepsy in the developing world, where prevalence rates of active epilepsy are twice those of developed countries^{3,4,5}. For more than 100 years, a cause-and-effect relationship between pathologic alterations in brain structure and seizure has been recognized. The precise mechanism by which seizures are produced is unknown but the association between structural pathology and focal onset seizures originating in or near the lesion is well accepted. It is presumed that seizures arise from neurons that lie adjacent to a lesion that is rendered by several possible mechanisms susceptible to spontaneous coherent hypersynchronous discharge. Nevertheless, how often structural cystic lesion cause epilepsy have been not well documented on the medical literature The main goal of this study is to identified cystic lesions in a series of patients with NCC presenting refractory epilepsy.

MATERIAL AND METHOD

The first consecutive 100 patients presenting uncontrolled epilepsy attending to epilepsy and NCC clinic from Nelson

Mandela Academic Hospital were selected for this study. CT scan and EEG test for all patients were performed.

SUBJECT INCLUSION CRITERIA

Patients suffering from partial onset (FS) or tonic-clonic generalized seizures (TCGS) according to the ILAE classification of epileptic seizures, whether or not secondarily generalized.

Diagnosed of epilepsy for no less than one year prior to be selected for this study

Epilepsy documented clinically and by paroxysmal activity on EEG.

Calcified NCC was confirmed by CT Scan of the brain in patients with multiple 2-10 mm intracranial calcifications.

The criteria used for identification of uncontrolled epileptic patients was established as follow: patient under regular treatment and good compliance taking 200mg of carbamazepine orally three times a day or patients taking 400mg of phenytoin orally at bedtimes and presence of at least and presence of at least eight FS with or without generalization or six TCGS per month.

Male/female subject, 13 to 75 years of age inclusive, weighted more than 50 kg

Patients on stable dose for at least two months before the selection.

Previous CT scan confirmed the free of primary or secondary brain tumors, progressive cerebral disease or any other progressively neurodegenerative disorder

Signed and dated written informed consent.

SUBJECT EXCLUSION CRITERIA

Pregnant ladies and females who are lactating.

Subjects whose seizures cannot reliably be counted on regular basis due to their fast and repetitive occurrence, severe or moderate mental retardation and illiterate peoples unable to report seizures.

History of stroke, cerebral schistosomiasis, current intracranial neoplasia, progressive cerebral disease or any other progressive neurodegenerative disease.

History of poor compliance, pseudoseizures, recurrent psychotic, major affective disorder or suicide attempts.

Normal CT scans of the brain and EEG

RESULTS

Eleven patients with NCC, uncontrolled epilepsy and intracranial structural lesions were identified. Among this group, 3 had arachnoids cysts (Figure 1). Schizencephaly was confirmed in two (Figure 2) Six patients presented signs of racemose NCC (Figure 3) and four of them with radiological signs of neuro-AIDS (Figure 4)

Figure 1

Figure 1: Shows arachnoids cyst and mixed NCC (Active and calcified)

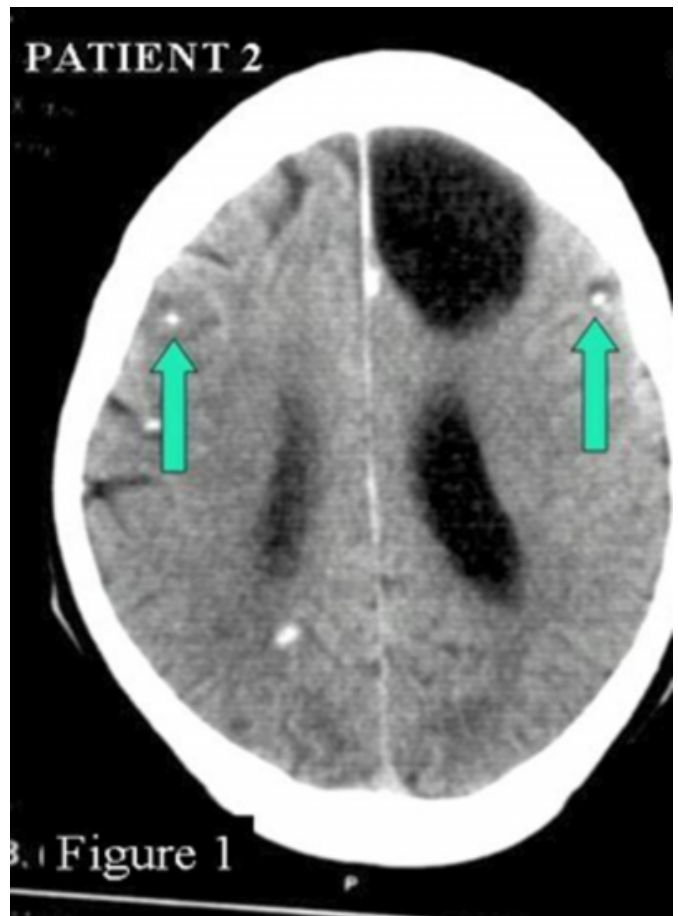


Figure 2

Figure 2: Shows radiological signs of schizencephaly and calcified NCC

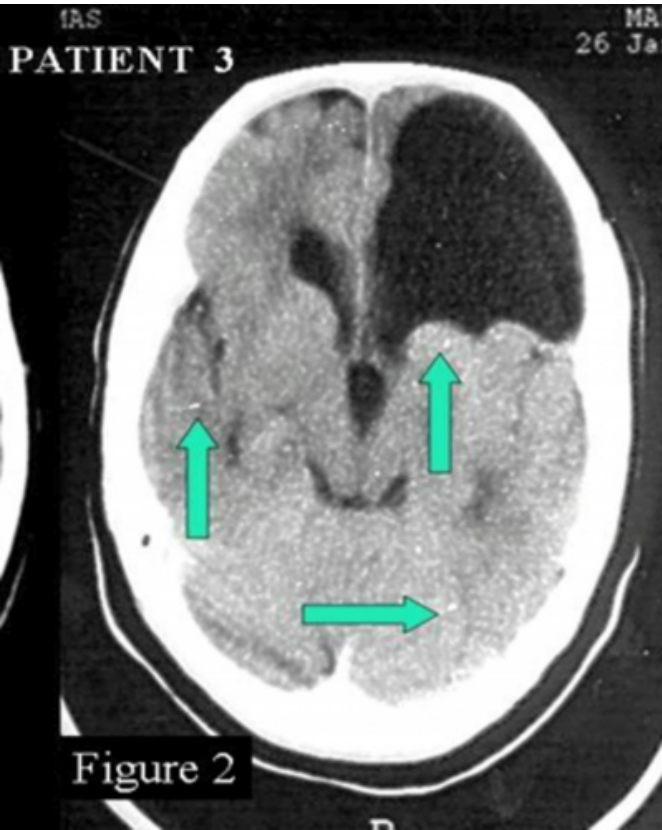


Figure 3

Figure 3: Typical radiographic presentation of racemose cysticercosis

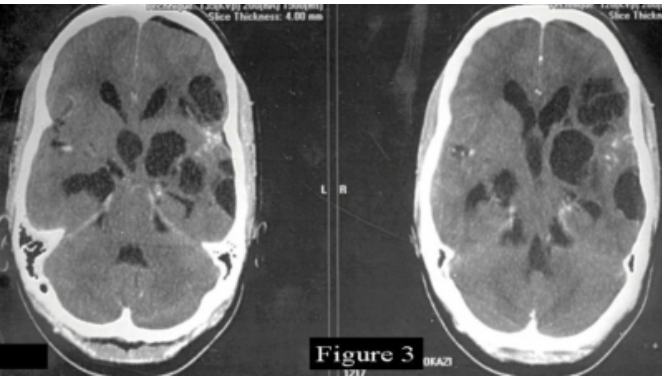
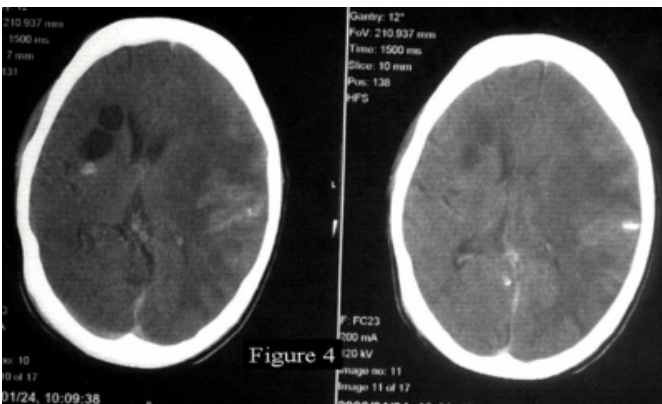


Figure 4

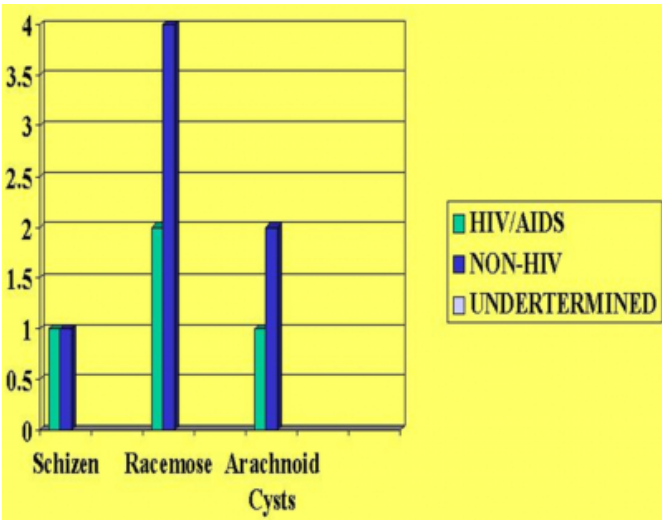
Figure 4: Shows radiological signs of racemose cysticercosis on the right cerebral hemisphere and signs of cerebral toxoplasmosis on the left in a patient in stage IV of HIV/AIDS



NCC and associated neuro-AIDS problems was commonest cause of uncontrolled epilepsy in this series (Table I).

Figure 5

Table 1: NCC and uncontrolled epilepsy



Six patients were male and five females. All patients were black. (98% of the population for this region is black). All patients were included in the age group of 21-30 years (n=5), 31-40(n=4) and 41-50(n=2)

COMMENTS

Because the shortage of this series and the poor statistical values, aspects like gender, race, and ages are considering for discussion on this study.

The spectrum of gray matter developmental abnormalities ranges from widespread gross deformities such as lissencephaly to small focal nodular gray matter

heterotopias, and all are due to defects in neuronal migration and organization that occur in uterus. These include gray matter migration abnormalities of the cortical mantle or cortical dysplasias (agyria, pachygyria, and polymicrogyria), abnormal location of gray matter or heterotopias (band, laminar, or nodular), schizencephaly, and hemimegalencephaly

Schizencephaly (Sc) and its different presentations are pathological process due to different disturbances of neuronal migration characterized by a cerebrospinal fluid-filled cleft, which is lined by gray matter. The cleft extends across the entire cerebral hemisphere, from the ventricular surface (ependyma) to the periphery (pial surface) of the brain. The clefts may be unilateral or bilateral and may be closed (fused lips), as in Sc type I, or separated (open lips), as in Sc type II. Patients presenting this uncommon disease show a variable outcome according to underlying brain lesion that can cause headache, several types of epileptic seizures, focal sensory-motor signs, and cognitive dysfunction^{6,7}. Clinical features of Sc are highly variable. Patients with unilateral clefts with fused lips may have mild hemiparesis and seizures but otherwise have normal development. When the cleft is open, patients present with mild-to-moderate development delay and hemiparesis; severity is related to the extent of cortex involved in the defect^{8,9}. Nevertheless, some patients only present epilepsy even in case of an associated NCC

Arachnoids cysts represent intra-arachnoids cerebrospinal fluid-containing cysts that do not communicate with the ventricular system and usually are not associated with brain maldevelopment. They constitute approximately 1% of intracranial masses, with 50-60% occurring in the middle cranial fossa. Cysts in the middle cranial fossa are found more frequently in males and on the left side. Most arise as developmental anomalies. A small number of arachnoids cysts are associated with neoplasm or occur as complications of adhesions following leptomenigitis, hemorrhage, or surgery¹⁰.

Arachnoids cysts often are an incidental finding on imaging and, usually, patients are asymptomatic even if the cyst is quite large. The most commonly associated clinical features are headache, calvarial bulging, and seizures, with focal neurological signs occurring less frequently. Controversy surrounds the treatment of arachnoids cysts. Some clinicians advocate treating only patients with symptomatic cysts while others believe that even in asymptomatic patients, cysts

should be decompressed to avoid future complications. The most effective surgical treatment appears to be excision of the outer cyst membrane and cystoperitoneal shunting¹⁰.

Arachnoids cysts can indent deeply into the hemisphere or invaginate into major fissures, displacing and flattening the underlying cortex. Compression severe enough to cause tissue necrosis is exceptional¹⁰.

In symptomatic patients, clinical features depend on the location of the arachnoids cyst. Cysts of the middle cranial fossa (50%) may compress the tip of the temporal lobe, displacing it in the occipital direction. This has been described as temporal lobe agenesis, although there is doubt as to the existence of a true temporal lobe agenesis. Pathologists believe that a malformation of the brain causing selective agenesis of the temporal does not exist. However, middle cranial fossa cysts are linked to ipsilateral chronic subdural hematomas. Rarely, they may communicate with the subdural space, forming a slitlike extension over the hemispheric surface¹⁰.

Unfortunately, information about refractory epilepsy and racemose NCC in HIV/AIDS patients was not available at the medical literature for us reviewed. However, at the present moment we are not able to propose a conclusion about these findings until other results from our research will be ready for analysis and discussion.

CONCLUSION

In the absence of ischemic stroke NCC-related or an associated intracranial infection, racemose cysticercosis in HIV/AIDS patients should be considered as an important cause of refractory epilepsy.

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