

A Case of Neurocysticercosis, Currently an Uncommon Cause of Epilepsy in Portugal

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Abstract

Neurocysticercosis (NCC), endemic in developing countries, where it is an important cause of epilepsy, is the most frequent parasitic disease of the central nervous system. Clinical manifestations may vary, with seizures being the most common form of presentation, such as in our report. In Europe, the disease is currently rare, but with the increasing number of immigrants from endemic countries, there has been an increase in the number of people with NCC, with Portugal registering 357 NCC hospitalized cases between 2006 and 2013 (mean of 45 cases per year). This is a case report of a 7-year-old child, from Angola, who presented to the emergency department in post-seizure status, with neuroimaging being decisive in establishing the diagnosis of NCC. Although uncommon in Portugal, it is important to consider this diagnosis in a child who presents with a non-febrile seizure, especially if the child emigrates from a high-prevalence area or has had a long stay in endemic regions.

Keywords: Epilepsy, Neurocysticercosis, Neuroimaging, Parasitic disease, Taenia Solium

Introduction

Neurocysticercosis (NCC), the most common parasitic disease of the human central nervous system, is caused by the larval stage of the pig tapeworm, *Taenia solium*. Endemic in most developing countries, represents an important cause of seizures in children in those countries, being the most common preventable cause of epilepsy, estimated to contribute up to 30% of epilepsy cases.[1-3] In developed countries, the incidence of the disease has been increasing, probably related to the growing number of travelers and populations immigrating from endemic areas, with Portugal registering 357 NCC hospitalized cases between 2006 and 2013 (mean of 45 cases per year).[2,4] This case report describes a rare cause of seizures in a developed country, the treatment of which remains a challenge for European health professionals, where the disease is still rare but increasing.

Case Report

A 7-year-old Angolan child, living in Portugal for about one year with a foster family was admitted to the Emergency Department in post-convulsive status.

The episode lasted around a minute and was accompanied by loss of consciousness, tonic-clonic movements in the right hemibody and upward rolling of the eye, with spontaneous resolution, and post-critical drowsiness. Denied loss of sphincter continence. She had a previous history of malaria attacks in Angola, and no other relevant personal history.

On day 2 of admission, a secondarily generalized tonic-clonic seizure and a subsequent focal motor seizure on the left upper limb were observed, stopping after IV midazolam, with no other associated symptoms. Physical examination including neurologic examination was unremarkable.

Analytical studies were performed upon admission and revealed a white blood count of 9.80×10^9 , hemoglobin 12.8 g/dL, platelet count $279 \times 10^9/L$, and creatinine kinase 250 U/L. Liver and kidney function, blood ionogram, and gas analysis were in the normal ranges. Brain Computed Tomography (CT) scan on admission revealed hypodensity of the right parasagittal parietal white matter, with a rounded area of approximately 7 mm of adjacent maximum diameter (Fig.1).



Fig 1: Brain CT scan revealed hypodensity of the right parasagittal parietal white matter, with a rounded area of approximately 7 mm of adjacent maximum diameter. She was hospitalized for diagnostic clarification. Electroencephalogram (EEG) disclosed interhemispheric asymmetry, evidencing right dysfunction, predominantly in the centroparietal regions. On day 4, Brain Magnetic Resonance Imaging (MRI) was performed revealing a focal, parietal cortico-

subcortical lesion and signal enhancement after contrast administration, highly suggestive of NCC in the vesicular-colloidal phase. Another temporal lesion was identified, possibly having the same etiology (**Fig. 2 and 3**).

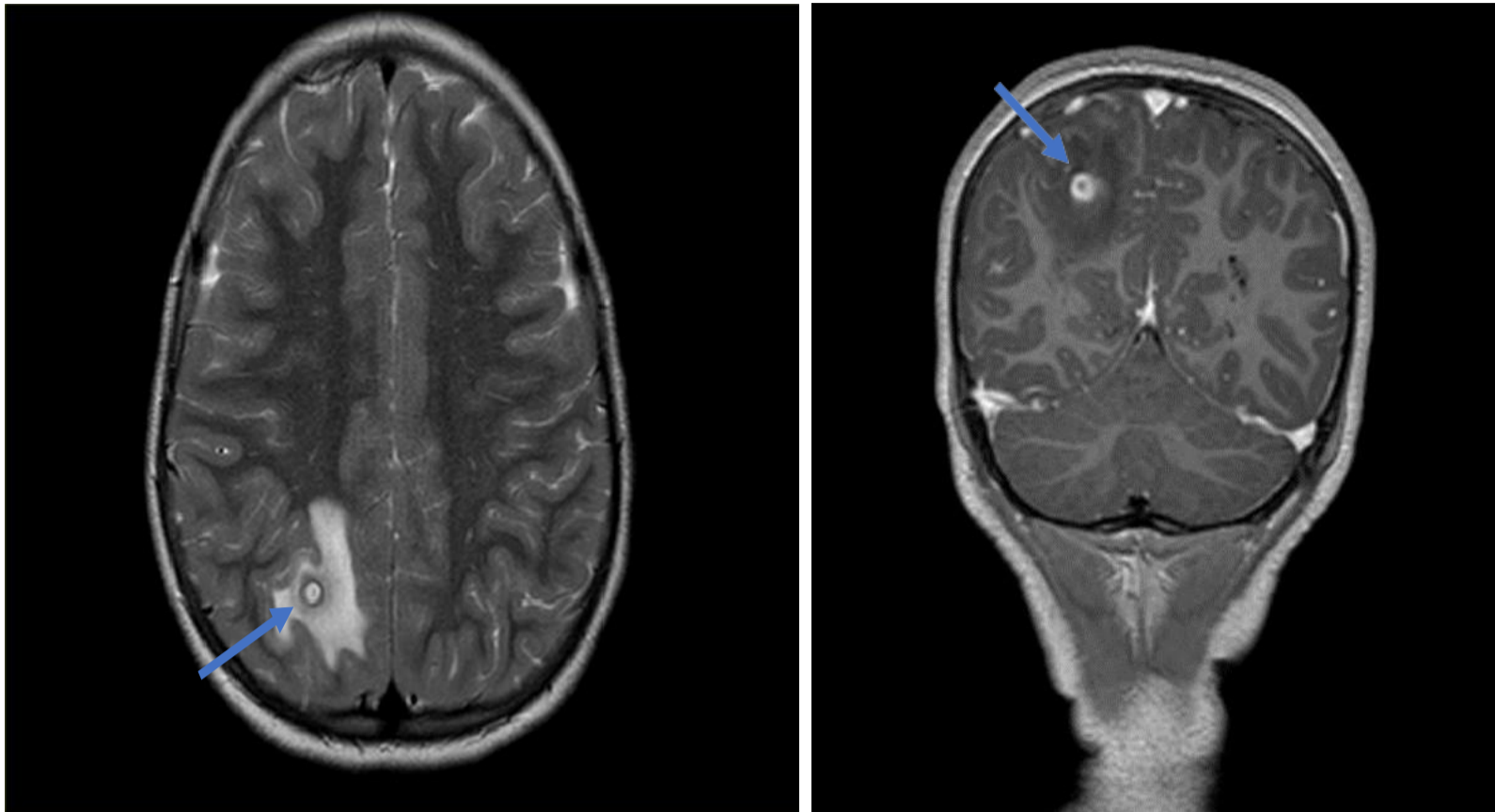


Fig 2 and 3: Brain MRI with a focal, high right parietal cortico-subcortical lesion, rounded, pericentimetric, with T2 hyposignal wall, T2 hypersignal center, and signal enhancement after contrast administration, bordered by an extensive area of edema.

The case was discussed with Infectious Diseases of a level III hospital, and further investigation was initiated aiming to discard other conditions that can mimic single or multiple ring or nodular enhancing lesions such as tuberculoma or cystic lesions of the brain including cystic echinococcosis. Laboratory tests for tuberculosis were negative, as were parasitic (*Taenia solium*; *Echinococcus*; *Cryptococcus*; *Toxoplasma gondii*), and retroviral serologies. Ophthalmology review through fundoscopy and additional chest X-ray, abdominal ultrasound, and echocardiogram were normal.

Anti-seizure medication (ASM) was started from day two with oral levetiracetam 20 mg/kg/day. It was then decided to start treatment with

albendazole, praziquantel, and prednisolone, complying with antiparasitic therapy for 14 days, subsequently weaning from corticosteroids, and maintaining ASM. There were no complications, nor seizures recurrence. In an imaging reassessment (MRI) at 12 weeks, practically total resolution of the lesion was observed, with a clear volumetric reduction of the contrast-enhancing lesion at the right parietal level, with very slight marginal edema, thus fulfilling confirmatory criteria for NCC (Del Brutto OH, et al *NeuroSci* 2017). EEG at 6 months of evolution maintaining right parietal epileptic activity and control Brain MRI at 9 months revealed continued imaging resolution of the lesions, with only focal sequelae (**Fig. 4 and 5**).

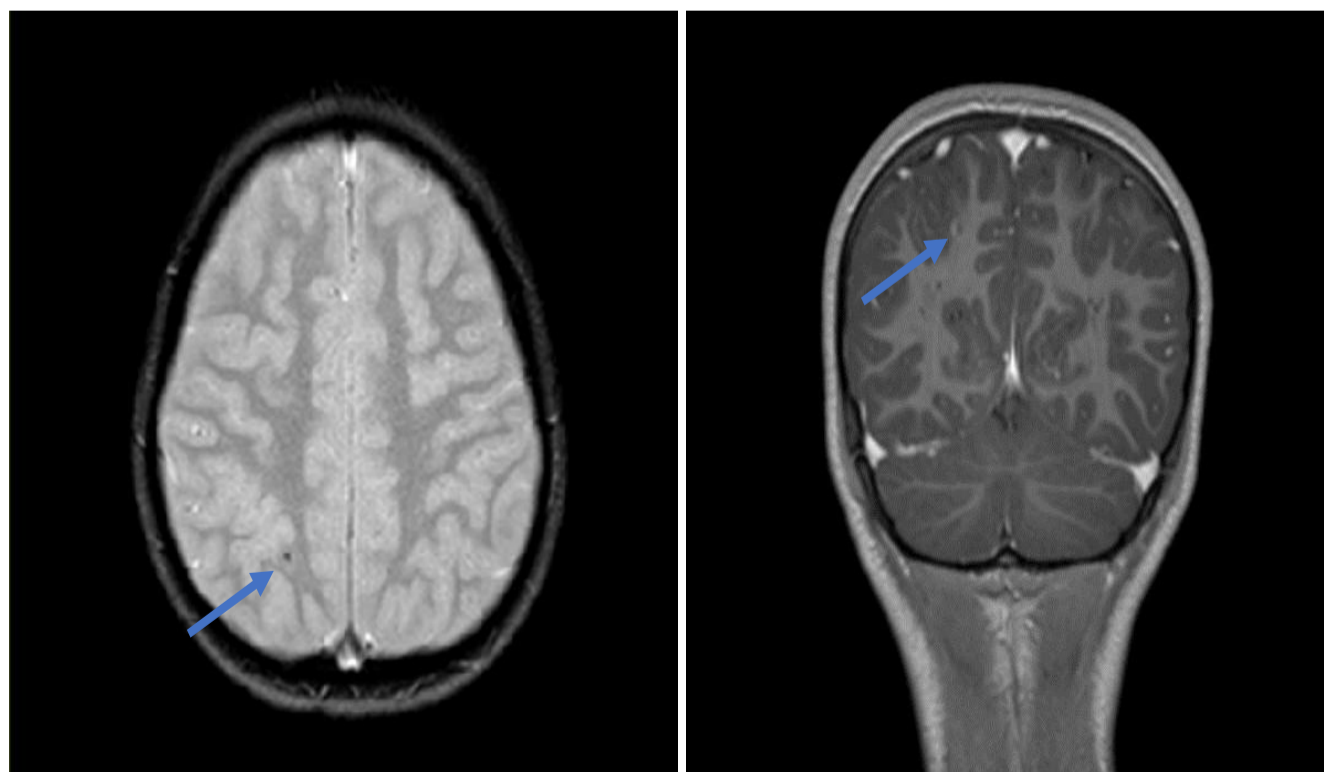


Fig 4 and 5: Brain MRI subsequent study at 9 months, with continued imaging resolution of the lesions, with only focal sequelae.

Clinical evaluation at 12 months disclosed no impact on learning skills. Levetiracetam monotherapy was maintained.

Discussion /Conclusions

NCC is the most frequent cause of neuro parasitosis in the world, being a major cause of epilepsy, mainly in developed countries, where is endemic.[1]

It is caused by encysted larvae of *Taenia Solium*, that has a two-host life cycle, human and pig, being the human the definitive host for the adult form of parasite, while both can be intermediate hosts, carrying the larval form (cysticercus). Cysticercosis is transmitted by the ingestion of *Taenia solium* eggs passed in the feces of a human being carrying the parasite.[5]

Humans become carriers of the parasite (intestinal infection) by eating raw or undercooked pork that contains cysticerci, being at risk of self-inoculation of eggs via the fecal-oral route, and subsequent development of symptomatic cysticercosis.[6]

In developing countries, where sanitary regulations are often neglected, pigs acquire the infection through ingesting food or water contaminated by human feces from carriers of intestinal infection, becoming the intermediated host. In the pig's intestinal tract, eggs embryonate into oncospheres, crossing the intestinal wall and, through the bloodstream, migrating to the target tissues. The cycle is interrupted if individuals accidentally ingest eggs through food or water contaminated by feces. In this case, the parasite perceives the human as an intermediate host and disseminates to a variety of organs, including the central nervous system, where cysticerci develops. [6,7] The cysts pass through a sequence of four morphological stages: vesicular, colloidal, granular nodular, and nodular calcified stage.[8] Because of its decreasing incidence in our local epidemiology, diagnosis may be more difficult if clinical suspicion is low and if risk factors are not addressed during anamnesis. Our patient had some risk factors, including recent residence in an endemic area and the absence of a clear social history of non-exposure to contaminated food or water.

The clinical manifestations of NCC are variable, depending on the number, stage, and location of the lesions as well as the host response. A large part of individuals holding the parasite in the central nervous system are asymptomatic and the symptoms may develop many years after the infection.[7] Most cysts are located in the brain parenchyma and therefore may provoke seizures, which was the initial symptom in our patient.[8]

The diagnosis of NCC is based on epidemiology/exposure, clinical manifestations, neuroimaging, and immunological testing. Neuroimaging with either a CT scan or MRI is considered the gold standard for diagnosis of neurocysticercosis. [3,9]

The most frequent manifestations in cases of parenchymal location are seizures; learning disability, behavior changes, and difficulty with balance, depending on the affected area. Extra-parenchymal forms of NCC cause strong inflammatory reactions with signs of increased intracranial pressure (headache, vomiting, and papilledema), often accompanied by fever and eosinophilia. Parenchymal forms of NCC have a better prognosis than extraparenchymal forms. [3,9] Other rarer presentations (~ 1% of cases) of extra parenchymal NCC are spinal cord NCC and ocular NCC.[5] Since our patient presented no other signs or symptoms before the initial seizure, differential diagnosis at admission encompassed causes of afebrile seizures. Ionic alterations and hypoglycemia were excluded through lab work-up. The

absence of fever and prodromous symptoms made it less likely a central nervous system infection like meningitis. Even though she was in Angola in the past year and her past medical history included malaria, the suspicion of a CNS infection was low. CNS involvement by *Plasmodium* can provoke focal seizures, but it usually presents with fever and headache, often leading to encephalopathy, which was not the case, nor did the lab work show frequent accompanying changes like thrombocytopenia or hepatic cytolysis. Post-malaria neurological syndromes usually present 2 months after recovery and would be, therefore, even more unlikely. Additionally, the full recovery after the seizure was also not in line with other diagnoses such as stroke and drug/toxin overdose. Accounting for the focal characteristics of the seizure, concern about lesions in the CNS arose, therefore leading to a decision to perform a head CT which showed parenchymal lesions compatible with parasitosis.[10]

When assessing potential parasites involving the CNS, particular attention should be added to NCC and *Plasmodium*, as they are the two most frequent causes of seizures as the first presentation. However, all human parasites can potentially infect the CNS, including toxoplasmosis, echinococcosis, or schistosomiasis, and less frequently paragonimiasis, toxocariasis, onchocerciasis, trypanosomiasis and angiostrongyliasis. Some of these infections are less likely to present with multinodular lesions in the cerebral parenchyma, including toxoplasmosis' predisposition to affect basal ganglia, schistosomiasis with granulomas and infarctions, trypanosomiasis with edema and arachnoiditis or paragonimiasis in which exudative inflammation and cerebral hemorrhage/infarction prevail. Echinococcosis can present with cysts mimicking NCC. When CNS parasitosis is suspected, particularly when toxoplasmosis is in the differentials, screening for potential causes of immune compromising should be performed, namely HIV serologies, which were negative.[10]

Serological and Cerebrospinal (CSF) fluid tests have been developed to detect anti-cysticercosis antibodies and cysticercosis antigens. The best serological test is the enzyme-linked immunoelectrotransfer blot (EITB), which uses antigens (glycoprotein fractions) to detect antibodies against *T. solium* in serum. The sensitivity of the EITB is 98% for patients with two or more viable cysts in the nervous system.[5]

A lumbar puncture to examine the CSF is usually not necessary but may be useful to exclude other diagnoses or extra parenchymal forms of NCC with aseptic meningitis. CSF changes are more frequent in patients with active inflammation and multiple lesions or in cases of ventricular or subarachnoid NCC. They include mononuclear pleocytosis and a slight increase in protein concentration; low CSF glucose concentrations have been associated with a worse prognosis.[5]

Direct visualization of the parasite on fundoscopy is pathognomonic of NCC, however, our patient had no remarkable findings in the ophthalmological review. Despite ocular NCC being infrequent, even in endemic areas, and many individuals are asymptomatic, the inflammation associated with the cyst in the degenerative phase can threaten vision.[11]

For the treatment of NCC, a multidisciplinary approach is recommended, and an individualized approach must be taken in to account due to the clinical

manifestations of the disease, as well as the number, location, and stage of injuries. Due to the different clinical manifestations and the limitations of the literature, many of the recommendations are based on observational studies, informal data, or expert opinion and not based on randomized clinical trials or controlled. The initial therapeutic approach includes symptomatic therapy, and it may be necessary surgery in some cases of extra-parenchymal NCC. Antiparasitic therapy, combined with anti-inflammatory agents, is important and recommended, but never urgent, and must be preceded by therapeutic symptomatic. NCC-associated epilepsy with viable cysts usually responds well to first-line ASM, which is recommended for all individuals with seizures, for a minimum of two years after the last convulsion, with progressive weaning. [5,12]

In patients without intracranial hypertension, it is recommended the administration of antiparasitics, albendazole [15mg/kg/day] in monotherapy for 10 to 14 days in patients with one or two viable cysts. In patients with more than two viable cysts, dual therapy with albendazole [15 mg/kg/day] and praziquantel [50 mg/kg/day] for 10 to 14 days is recommended. In both cases, patients should undergo adjuvant corticosteroid therapy prior to treatment with antiparasitic drugs, as these can worsen the symptoms of NCC by induction of inflammatory response. Since our patient had 2 lesions, the latter therapeutic was chosen. [9,12,13]

Patients should be followed clinically for seizure recurrence and optimization of ASM. The first clinical follow-up after the initial diagnosis should be performed at 2–4 weeks to determine if the patient has developed any recurrent seizures or new or worsening symptoms/signs. New or worsening symptoms should advise prompt reimaging. Neuroimages show degeneration and/or resolution of active parenchymal lesions after 3–6 months of antiparasitic treatment and should be performed every 3–6 months until resolution of the cystic lesion since it is suggested retreatment in those with persistent lesions. At the 3-month follow-up radiology screen, our patient had a substantial improvement in findings and therefore was decided to rescreen only after another 6 months. A CT scan should be performed prior to consideration of stopping ASM to determine if calcifications have developed. [12,13]

Although actually not frequent, at least in the pediatric population, the NCC incidence has been increasing in Portugal as in other developed countries as a result of the growing number of immigrant populations from endemic areas. This case highlights the need to consider a parasitic infection of the central nervous system, especially NCC, in a child presenting with a new onset of focal epilepsy, especially if he had a long-term stay in endemic regions or has emigrated from an endemic area. In cases of relevant suspicion of NCC, neuroimaging examinations are decisive and usually highly indicative, and immunological testing may be necessary in selected cases. [2]

conflict of interests: The authors declare that there is no conflict of interest.

Statements and contributions of authors

The authors declare that they:

- Agree to the author proposed to be the corresponding author.

- Agree to the number of authors and their order of presentation in the article.
- Have contributed significantly to the work identified hereinabove, namely to the conception and design, or acquisition of data, analysis or interpretation of data; and to drafting the article or revising it critically for important intellectual content; and to the final approval of the version to be published; and agreeing to be accountable for all aspects of the work.

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