

Case Report

A case of imported neurocysticercosis in Portugal

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Abstract

Neurocysticercosis (NCC) is the most common cause of acquired epilepsy in resource-poor countries. We report the case of a 24-year-old woman born and residing in Guinea-Bissau, who was transferred to Portugal two months after the onset of a possible meningitis (fever, headache, seizures, and coma) that did not respond to antibiotic treatment. The diagnosis of NCC was confirmed by MR imaging, which showed multiple lesions compatible with cysticercus, and by polymerase chain reaction (PCR) of the cerebrospinal fluid. After 28 days on albendazole and dexamethasone without improvement, she was started on praziquantel, which she completed in six weeks with progressive recovery.

Key words: neurocysticercosis; *Taenia solium*; Portugal; Guinea-Bissau.

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Introduction

Neurocysticercosis (NCC) is a chronic inflammatory infection of the central nervous system (CNS) caused by the larval form of the tapeworm *Taenia solium*. The disease is endemic in many low- and middle-income countries of the world where sanitation is poor, pork consumption occurs, and pigs are managed under free-range conditions and are exposed to human feces. Infections with cysticercus occur after humans consume the ova either from exogenous sources or through self-infection via the fecal-oral route. Ova release oncospheres, which penetrate the intestinal wall, reach the bloodstream, and develop into cysticerci in any organ; most are located in the CNS, eyes, muscles, or subcutaneous tissues. In general, symptomatic infections occur in the eyes and in the CNS.

The importance of NCC was not truly recognized in the past, as most cases occur in resource-poor countries. However, it is now becoming more clear that greater priority should be given to this disease, which is now classified by the World Health Organization (WHO) as one of the 17 neglected

tropical diseases [1]. The disease has an enormous health and economic impact, and according to the Food and Agriculture Organization for the United Nations, cysticercosis is one of the few potentially eradicable diseases [2].

Approximately 90% of the world's 50 million people with epilepsy live in resource-poor countries [3], and a linkage between epilepsy and NCC in countries endemic for *T. solium* is now accepted. It is estimated that people with cysticercosis have a 3.4 to 3.8-fold increased risk for developing epilepsy [4].

However, the global burden of the disease is mostly unknown, as diagnosis requires specific serology and brain imaging, a technology commonly unavailable in resource-poor areas where the disease is most common, and clinical diagnosis is unreliable, as clinical manifestations are diverse and unspecific [5]. Recognition of NCC as a serious public health problem in sub-Saharan African countries is increasing [6]. According to autopsy studies in Africa, the prevalence of NCC in pig-raising areas varies from 0.45% to 7% [7]. In epileptic patients in South Africa,

approximately one-third had evidence of NCC on computerized tomography (CT) scans [8].

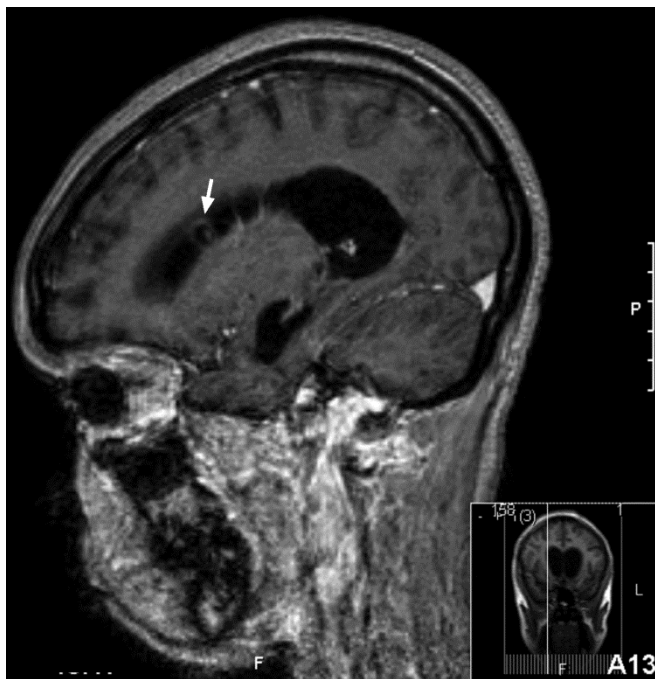
In European countries that have strong historical and economic links with former African colonies, such as Portugal, most NCC cases occur in immigrants from endemic regions or, less frequently, in travelers to those regions. However, since NCC is not a notifiable disease, the total number of cases in Europe is mostly unknown. According to one publication, between 1970 and 2013, 176 cases of NCC were diagnosed in Europe [9]. However, even in Europe, NCC is surely underreported; in a single hospital in Lisbon (Santa Maria Hospital), over a period of 10 years (2003 to 2013), 53 cases were diagnosed, mostly imported from Portuguese-speaking African countries.

Case Report

One of those cases is reported here, occurring in a previously healthy 24-year-old woman, born and residing in Guinea-Bissau. In May 2013, she was admitted to the Simão Mendes Hospital in Bissau because of fever and a severe headache, which was followed by seizures and coma. Most details on diagnosis and treatment for a period of two months were unknown; the diagnosis of bacterial meningitis was considered, and the patient completed two weeks of ceftriaxone without clinical improvement. A brain CT scan, done in Senegal in early July 2013, apparently did not reveal any abnormalities. In July,

she was transferred to Lisbon, Portugal, where she was admitted to the Infectious Diseases Department of Santa Maria Hospital. On admission, she was in a coma (Glasgow coma scale [GCS] 6), she was unable to focus her vision, and her eyes revealed a deviation to the right side, with occasional horizontal slow movements. She had left-sided hemiparesis and revealed neck stiffness. The magnetic resonance imaging (MRI) showed hydrocephalus, edematous frontal and temporal areas, enhancement of the leptomeninges, and multiple lesions compatible with viable cysticercus, one being intraventricular (Figure 1). She had an external ventricular drainage that was later changed to a ventriculo-peritoneal shunt, which improved her state of consciousness to GCS 9-10. An electroencephalogram (EEG) revealed diffuse slow rhythm and a right-sided predominance, without evidence of epileptic activity. In-house PCR for cysticercus in the cerebrospinal fluid (CSF) was positive (National Health Institute, Doutor Ricardo Jorge, Lisbon). She was started on albendazole (15 mg/kg/day) and dexamethasone (40 mg/day). After 28 days without clinical or radiological improvement, she was started on praziquantel (50 mg/kg/day). After six weeks, she had a slow but progressive clinical recovery, which was confirmed by concomitant radiological improvement. She was discharged in November 2013, having re-gained speech as well as the ability to swallow. Her left-sided hemiparesis improved slightly, and though she gained mobility in her extremities, she continued to be dependent at the time of hospital discharge.

Figure 1. Brain MRI scan (T1-weighted image) of a 24-year-old woman showing an intraventricular cyst (arrow)



Discussion

This case illustrates the difficulty in the diagnosis of NCC in countries where the disease is more prevalent. Diagnosis requires a reliable medical infrastructure, knowledgeable medical personnel, brain imaging technology, serological assays, as well as access to medication. In many endemic countries, such as Guinea-Bissau, healthcare facilities are fragile and none or few of these conditions or technologies are available. Healthy life expectancy at birth is 42 years, and there are fewer than five physicians per 100,000 persons in the country [10]. In the framework of health protocols established between Portugal and the former African colonies, some severely ill patients can be transferred to Portuguese hospitals. Between 2004 and 2010, approximately 3,250 patients were transferred from Guinea-Bissau to Portugal [11], almost all to Santa Maria Hospital in Lisbon.

The diagnosis of NCC is suspected in a person from an endemic area or who traveled to those areas and presents with seizures and headache. More than 70% of symptomatic patients develop seizures [12], but clinical presentation is related to the cyst localization. When cysts are ventricular, such as in this case, meningitis, meningoencephalitis, and hydrocephalus due to the blockage of the subarachnoid space are common [12].

There are absolute criteria for NCC diagnosis that permit an unequivocal diagnosis even if considered alone. Besides the histological demonstration of the parasite from a biopsy of a brain or spinal cord lesion, and the direct visualization of subretinal parasites by fundoscopic examination, the presence of cystic lesions showing the scolex on CT or MRI also enables the diagnosis. The presence of cystic lesions demonstrating the scolex is considered pathognomonic of NCC [13]. The scolex can be visualized as a bright nodule within the cyst, producing the so-called hole-with-dot image [14].

The mainstay for treatment of NCC is a combination of albendazole and dexamethasone along with symptomatic medication, including anticonvulsants or surgery. The optimal duration of the regimen continues to be debated, but mainly depends on the number and localization of the lesions. It is accepted that patients with multiple lesions should receive a longer treatment regimen. Treatment for patients with subarachnoidal lesions is even more controversial. Studies attempting to clarify the role of longer regimens (> 28 days) or regimens with or without praziquantel [15] are generally based on small samples, but combined or sequentially treatment regimens emerge as alternatives [16]. The case presented here underlines a combined approach as valid and beneficial, despite the known interactions between corticosteroids and praziquantel.

Autochthonous cases still exist in Portugal as well as in other European countries [17] but are rare, contrary to what was stated in a publication from 2013 suggesting that Portugal is an endemic country for cysticercosis [9]. In fact, the data presented in that work refers to publications from the 1990s and from a study in a geographically restricted area in the north of Portugal [18]. However, clinicians must be aware that NCC can be diagnosed in persons born in non-endemic areas, and in such cases it is crucial to search for a household contact with a tapeworm infection.

In Western European countries, the reduction in the number of cases of human disease was feasible through improved animal husbandry and regular meat

inspections, measures that can eradicate porcine cysticercosis. Nonetheless, these strategies are not possible in many regions of Africa, where subsistence farming is the standard. Infected pigs can be diagnosed by the presence of cysts in the tongue, but in many cases, poor farmers cannot afford the economic losses of condemnation of these animals and they will sell that meat through informal channels. In endemic regions, mass administration of praziquantel has been attempted as a means to eradicate human tapeworm carriage [19], but this is still not an easy option for most African countries. A recent paper has called upon the tropical diseases community to support programs aimed at elimination of the infection [20].

References

- World Health Organization (2014) The 17 neglected tropical diseases. Available: http://www.who.int/neglected_diseases/diseases/en/. Accessed 6 March 2014.
- Maguire JH (2004) Tapeworms and seizures-treatment and prevention. *N Engl J Med* 350: 215-217.
- Weka RP, Ikeh EI, Kamani J (2013) Seroprevalence of antibodies (IgG) to *Taenia solium* among pig rearers and associated risk factors in Jos metropolis, Nigeria. *J Infect Dev Ctries* 7: 67-72. doi:10.3855/jidc.2309
- Quet F, Guerchet M, Pion SDS, Ngougou EB, Nicoletti A, Preux PM (2010) Meta-analysis of the association between cysticercosis and epilepsy in Africa. *Epilepsia* 51: 830-837.
- Coyle CM, Mahanty S, Zunt JR, Wallin MT, Cantey PT, White AC Jr, O'Neal SE, Serpa JA, Southern PM, Wilkins P, McCarthy AE, Higgs ES, Nash TE (2012) Neurocysticercosis: neglected but not forgotten. *PLoS Negl Trop Dis* 6: e1500.
- Qekwana ND (2013) Report on one health approach: Risk Management for Neurocysticercosis workshop- 28th June 2013, Pretoria, South Africa. *S Afr Med J* 103: 881.
- Geerts S (1995) Cysticercosis in Africa. *Parasitol Today* 11: 389-390.
- Danseu RD, Hay M, Cowie RL (1992) Seizures and neurocysticercosis in black men. *S Afr Med J* 81: 424-425.
- Fabiani S, Bruschi F (2013) Neurocysticercosis in Europe: Still a public health concern not only for imported cases. *Acta Trop* 128: 18-26.
- World Health Organization (2010) Global Health Indicators-Part II. Available: http://www.who.int/whosis/whostat/EN_WHS10_Part2.pdf. Accessed 6 March 2014.
- Henriques MA (2012) A imigração PALOP em Portugal. O caso dos doentes evacuados. *Forum Sociol* 22: 52-61.
- Carabin H, Ndimubanzi PC, Budke CM, Nguyen H, Qian Y, Cowan LD, Stoner JA, Rainwater E, Dickey M (2011) Clinical manifestations associated with neurocysticercosis: a systematic review. *PLoS Negl Trop Dis* 5: e1152.
- Dumas JL, Visy JM, Belin C, Gaston A, Goldlust D, Dumas M (1997) Parenchymal neurocysticercosis: follow-up and staging by MRI. *Neuroradiology* 39: 12-18.

14. Suss RA, Maravilla KR, Thompson J (1986) MR imaging of intracranial cysticercosis: comparison with CT and anatomopathologic features. *Am J Neuroradiol* 7: 235-242.
15. Kaur S, Singhi P, Singhi S, Khandelwal N (2009) Combination therapy with albendazole and praziquantel versus albendazole alone in children with seizures and single lesion neurocysticercosis: a randomized, placebo-controlled double blind trial. *Pediatr Infect Dis J* 28: 403-406.
16. Garcia HH (2008) Antiparasitic drugs in neurocysticercosis: albendazole or praziquantel? *Expert Rev Anti Infect Ther* 6: 295-298.
17. Luessi F, Sollors J, Frauenknecht K, Schwandt E, Mueller HD, Stoeter P, Blum J, Thoenke F (2009) Neurocysticercosis with a single brain lesion in Germany: a case report. *Cases J* 2: 8692.
18. Monteiro L, Coelho T, Stocker A (1992) Neurocysticercosis-a review of 231 cases. *Infection* 20: 61-65.
19. Cruz M, Davis A, Dixon H, Pawlowski ZS, Proano J (1989) Operational studies on the control of *Taenia solium* taeniasis/cysticercosis in Ecuador. *Bull World Health Organ* 67: 401-407.
20. Nash TE, Mahanty S, Garcia HH (2013) Neurocysticercosis-more than a neglected disease. *PLoS Negl Trop Dis* 7: e1964.

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