

Clarissa U. Prazeres da Costa
Helga Gräfin von Einsiedel
Rüdiger Disko
Achim Berthele

Multicystic tumor in the fourth ventricle

Consider neurocysticercosis

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Sirs: Neurocysticercosis (NCC) is the most common parasitic infestation of the CNS worldwide. In Latin America, Asia and Africa, where the prevalence of cysticercosis is high, NCC is the most frequent cause of seizures and hydrocephalus in adults and is thus easily recognized as a cause of acute neurological deterioration [1, 2]. By contrast, prevalence in Eastern Europe and the Iberian Peninsula is moderate, and the disease is assumed to be eradicated in Northern Europe. However, 'imported cases' can still be encountered [3]. Thus, outside endemic regions diagnosis of NCC might be delayed as in our patient:

A German female in her sixties developed hydrocephalus due to CSF obstruction. Symptoms started in 2002 when she noticed slightly disturbed motor coordination—she was not able to improve her golf handicap! Ataxia worsened quickly, accompanied by constant headache, and finally progressive cognitive deficits evolved. Cranial CT followed by MRI revealed obstructive hydrocephalus due to a tumorous mass within the fourth ventricle (Fig. 1A–C). After placement of a ventriculoperitoneal shunt, the patient recovered quickly and the neurological deficits resolved. Concerning the tumorous mass, neoplasia was suspected. Due to the Gd-enhancement an ependymoma was assumed to be most likely, and she was advised to have the tumor excised. Eventually, the patient presented her MRI scans to one of the authors (HE). Knowing that the patient had lived for more than 15 years in Mexico and Brazil (between 1972 and 1990), NCC was suspected for the first time, with the Gd-enhancement disclosing probable viable cysts and ependymitis. Serum ELISA and Western Blot detected *Taenia solium*-specific antibodies. Since clinical and radiological signs of systemic cysticercosis were lacking, we refrained from a confirmatory CSF analysis. Antihelminthic chemotherapy was administered with Albendazole at 15 mg/kg daily for 30 days, accompanied by a taper of dexamethasone. In follow-up MRI regression of cysts could be noticed as early as 14 days after the beginning of Albendazole. Two, three and four months later, regression of cysts was still ongoing and the decision was made not to give a second course of chemotherapy.

Definite diagnosis of intraventricular NCC, which accounts for

approx. 10–20% of NCC cases [4, 5], can be difficult for various reasons. First, MRI signs [5] are less pathognomonic, since it may comprise only unilocular cysts, especially in the fourth ventricle. Second, serological diagnostics of NCC have several obstacles [2, 7, 8]. Antibody titers can be low especially in the case of circumscribed NCC, or even absent when immune tolerance towards the parasite develops. False-negative results in serum ELISAs may occur in about 30% of cases, and, at least in endemic areas, false-positive results are frequent owing to cross-reactivity with other helminths [9].

Moreover, the treatment of intraventricular NCC is still a matter of debate [2]. For years, surgical excision has been advocated as first line therapy [10]. However, it has recently been proposed that in the case of concomitant ependymitis shunt surgery might be superior [4] and several cases of successful treatment with antihelminthics have been reported. Proano et al. have published a small prospective study on the treatment of fourth ventricle cysts with Albendazole: in 8 out of 10 patients, the cysts completely disappeared [11].

In our patient Albendazole treatment was well tolerated. 1½ years after therapy, CT disclosed a calcified remnant in the fourth ventricle next to moderate signs of persisting local ependymitis on MRI, and the ventricles were of normal size (Fig 1D). Thus, we refrained from any further therapy, and she is still doing well.

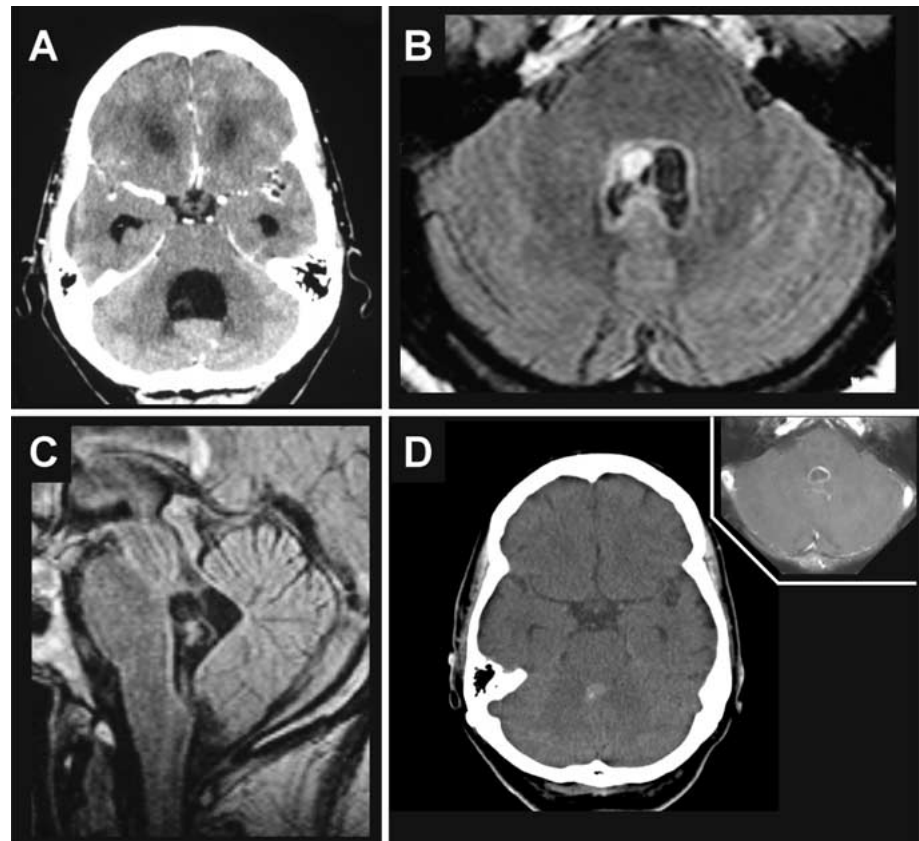
To conclude, although NCC in its classical clinical picture of a disseminated parasitosis is easy to recognize, it might as well present with solitary lesions

C. U. Prazeres da Costa · R. Disko
Inst. of Medical Microbiology
Immunology and Hygiene
Technical University
Munich, Germany

H. Gräfin von Einsiedel
Dept. of Neuroradiology
Technical University
Munich, Germany

A. Berthele (✉)
Neurologische Klinik der TU Muenchen
Moehlstr. 28
81675 Muenchen, Germany
Tel.: +49-89/4140-4673
Fax: +49-89/4140-4867
E-Mail: achim.berthele@neuro.med.
tu-muenchen.de

Fig. 1 CT and MRI scans of neurocysticercosis of the fourth ventricle. The initial CT scan (A) revealed a hydrocephalus occlusus due to a tumorous mass in the fourth brain ventricle. MRI (FLAIR) disclosed at least three cysticercal vesicles with one of them in the colloidal stage of involution rendering a high intensity signal (B and C). 1½ years after therapy, the cysts have calcified (plain CT in D), but signs of ependymitis still remain (insert; T1+Gd MRI)



mimicking tumors of non-infectious origin. Thus, even in Northern Europe, NCC should always be considered in patients with a cystic tumor and prompt physicians to carefully search the patient's history for a putative exposure to *T. solium* larvae. This is most obvious in the case of immigrants, but must not be forgotten in travellers or European expatriates returning from endemic regions.

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