Atypical radiological findings in cerebral hydatid disease

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ABSTRACT

يعد مرض الكيس العداري الدماغي نادراً جداً، ويمثل 2% فقط من مجموع الأمراض الدماغية. ويستند تشخيص المرض عادة على التصوير المقطعي، ونادراً ما تكون الصورة شاذة في تشخيص الكثير من الآفات مثل الآفات الدماغية التعفنية، أو الأوعية الدموية، أو الأورام. نستعرض في هذا التقرير حالتين غير نمطية (شاذة) من الكيس العداري الدماغي والذي تم تشخيصه عند سيدتين عمرهما 21 و24 عاماً. أشارت نتائج الأشعة المقطعية إلى احتمال وجود ورم الدبقيات القليلة التغصن في الحالة الأولى، وظهور الخراج الدماغي في الحالة الثانية. وقد كان التصوير بالرنين المغناطيسي مفيداً في تشخيص الحالتين. لقد خضعت كل مريضة لعملية جراحية ناجحة مع تحقيق نتائج جيدة. وتم تأكيد الطبيعة العدارية للكيس من خلال تحليل الأنسجة في كلتي الحالتين.

Cerebral hydatid disease is very rare, representing only 2% of all cerebral space occupying lesions. The diagnosis is usually based on a pathognomonic CT pattern. Exceptionally, the image is atypical raising suspicion of many differential diagnoses such as intracerebral infectious, vascular lesions, or tumors. We report 2 atypical cases of cerebral hydatid cysts diagnosed in a 21, and a 24-year-old woman. The CT scan results suggest oligodendroglioma in the first case and brain abscess in the second. An MRI was helpful in the diagnosis of the 2 cases. Both patients underwent successful surgery with a good outcome. The hydatid nature of the cyst was confirmed by histology in both cases.

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Hydatid disease is a parasitic infection caused by a cestode called *Echinococcus granulosus*. It is still an endemic disease in Mediterranean countries. The lung and the liver are the most frequently involved organs. Brain involvement occurs in only 2% of human hydatid infestations.[1,2] The CT scan is the tool of choice for establishing an accurate diagnosis preoperatively and for planning surgery. Classically, it demonstrates a large well-defined spherical cyst, smooth, thin-walled, with an inner density similar to CSF, and no perilesional edema or contrast enhancement. Exceptionally, the image is atypical raising the problem of differential diagnoses with intracerebral infectious, vascular, or tumoral processes. Herein, we report 2 cases of atypical radiologic findings of cerebral hydatid disease, where the diagnosis of hydatid nature was made during surgery in one case and by definitive histology in the other case. Our objective in presenting these particular cases is to highlight the atypical radiological features of cerebral hydatid disease.

Case Report. *Patient 1.* A 21-year-old woman, with no significant medical history, was hospitalized in our department as an emergency case of grand mal epilepsy. Anamnesis revealed that she suffered from an epileptic crisis when she was 10 years old. The crisis was well controlled by antiepileptic treatment (carbamazepine 800 mg daily). She had never undergone neuroradiological assessment before, however, at admission, her neurological examination was normal. The EEG disclosed focal slowing in the right frontal region with 6-3 Hz waves and epileptiform discharges. Cerebral CT scan demonstrated a right frontal calcified lesion, in contact with the frontal horn of the lateral ventricle, with no contrast enhancement, or perilesional edema (**Figure 1**). An MRI revealed a spherical heterogeneous mass in both T1 and T2 weighted sequences, with no contrast enhancement after gadolinium. In the center of the lesion, we noticed collapsed membranes with scalloped outlines (**Figure 2**). She was operated on through a right frontal approach. At surgery, we discovered a yellow cerebral mass flushing with the cerebral cortex (**Figure 3**). The content of the lesion was pulpy and easy to remove. Total excision of the lesion was achieved and confirmed by postoperative CT scan. Histological study revealed calcified cerebral hydatid cyst. The postoperative course was uneventful, and she did not receive any antiparasitic drugs. Two years

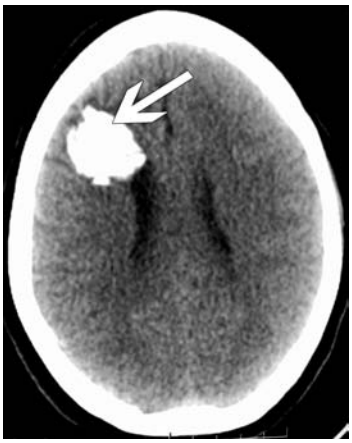

Figure 1 - Cerebral CT scan showing a right frontal calcified lesion (arrow) near the frontal horn of the lateral ventricle, with no contrast enhancement or perilesional edema.

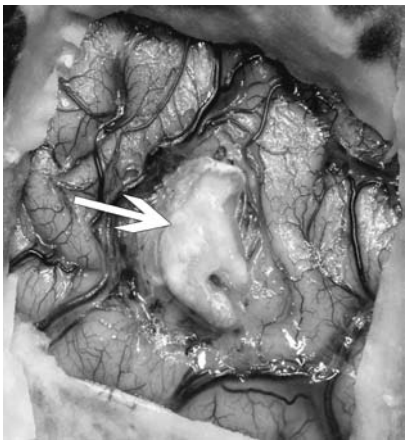

Figure 3 - Intraoperative view demonstrating a cerebral mass flushing with the cerebral cortex (arrow).

after the procedure, she remains asymptomatic, always under a single antiepileptic drug (carbamazepine at the rate of 400 mg per day), and since the intervention she reported no episode of convulsive crisis.

Patient 2. A 24-year-old woman, recently treated for chronic sinusitis, was admitted to the neurosurgical emergency department with a history of severe headache and vomiting of 2 weeks duration. On examination, she was conscious, with a normal neurological examination and pupils were bilaterally equal and reacting to light. The ophthalmologic examination revealed bilateral papilledema. A CT scan demonstrated a right frontal well-shaped spherical cyst, 5x4 cm in diameter, causing a mass effect on the midline structures. There was a contrast enhancement of the cyst wall after injection (**Figure** 4). Laboratory findings included an elevated erythrocyte sedimentation rate (80 mm/h), and a slight leukocytosis with a white blood cell (WBC) count of 14000/mm^3 (normal range 4000-12000 mm^3). The diagnosis of cerebral abscess was considered, and she underwent a right frontal trepanopunction as an emergency where 40 ml of xanthochromic-looking liquid was evacuated. Cytological analysis showed the presence of inflammatory cells with mononuclear pleocytosis of 80 WBC/mm^3. Bacteriologic culture was negative. She received intravenous ceftriaxone, gentamicin, and metronidazole, in addition to oral anticonvulsive treatment. A systematic cerebral CT scan was carried out by the end of the first week of treatment. It showed the persistence of the right frontal collection causing slight mass pressure on the homolateral lateral ventricle. Therefore, she was subjected to a second trepanopunction. However, and despite the 2-week treatment with systemic antibiotics, her ophthalmologic status worsened; the visual acuity was reduced to finger counting in the right eye and to 20/200 in the left eye. A CT scan on follow-up revealed

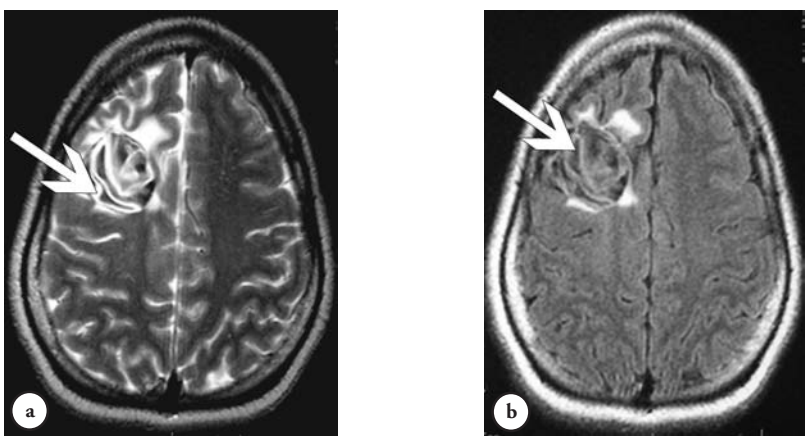

Figure 2 - Cerebral MRI in **a**) T2, and **b**) FLAIR sequences demonstrating a spherical heterogeneous mass, composed by collapsed membranes (arrows) with scalloped outlines.

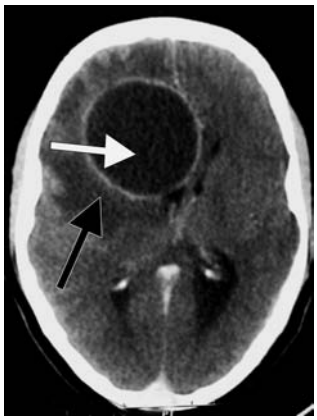

Figure 4 - Cerebral CT scan demonstrating a right frontal enhancing well-defined spherical cyst (white arrow) with important peripheral edema (black arrow) and a notable mass effect on the midline structures.

the same frontal accumulation with perilesional brain edema and persistent mass effect. A cerebral MRI demonstrated a smooth thin spherical cystic lesion, with complete rim enhancement surrounded by brain edema. The lesion was hypointense in T1 and hyperintense in T2 sequences, whereas the diffusion sequences suggested the possibility of cystic tumour (**Figure 5**). Subsequently, a direct approach of the lesion was chosen. At surgery, we discovered an encapsulated mass containing interiorly a translucent fine whitish membrane whose macroscopic appearance pointed out, with no doubt, a hydatid cyst. The mass was easily dissected from the surrounding brain parenchyma; 5 mm thick, greyish pink in color and tough in consistency. The histological study revealed a cerebral hydatid cyst widely reshaped by an inflammatory reaction, and surrounded by a granulomatous reaction composed of epithelioid and multinucleated giant cells. Postoperatively, she received chemotherapy with oral Albendazole 400 mg twice daily for 6 months. She has been on follow-up for the last 2 years with no further clinical or radiological evidence of recurrence.

Discussion. Hydatidosis is a parasitic disease caused by the metacestode of the larval stage of the genus *Echinococcus granulosis*. Embryos or scolices of the parasite reach the brain after successful passage through the lymphomesenteric, hepatic, pulmonary, and coronary capillary filtering systems, and grow with minimal resistance into hydatid cysts. The usual reaction of the brain tissue is to form a protective, and supportive layer composed of glial cells around the cysts. Brain tissue is not infiltrated, but is pushed aside by the cyst.[1] Generally, cerebral hydatid disease is most commonly seen in children and young adults,[1] and there is no significant difference in the rate of occurrence between the sexes. The clinical symptoms are various and not specific of the disease.[1] The lesions of cerebral hydatid disease are usually seen in the middle cerebral artery territory, particularly in the parietal lobe. Most cysts are found in the cerebrum and quite rarely in the posterior cranial fossa or ventricles.[3,4] A CT scan is an accurate method for establishing the diagnosis preoperatively.[2] It usually demonstrates a spherical and well-defined, smooth, thin walled, homogeneous cystic lesion containing a fluid with a density similar to the CSF. The cyst is classically non-enhanced after contrast injection and not surrounded by perifocal brain edema. Compression of the midline structures and ventricles is seen in most of the cases. The presence of daughter cysts is considered pathognomonic, but has been rarely reported.[3] These classical features make it easy to distinguish cerebral hydatid cysts from cystic tumors, porencephalic cysts, brain abscess, or arachnoid cysts.[3] However, unusual aspects may cause difficulties in CT

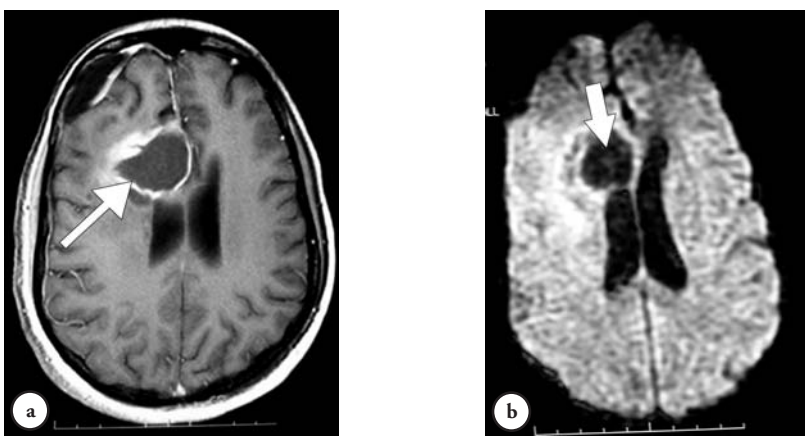

Figure 5 - Axial cerebral MRI in **a)** contrast T1, and **b)** diffusion-weighted sequences demonstrating a smooth thin spherical cystic lesion surrounded by brain edema with complete rim enhancement (arrow).

scan interpretation, and may lead to confusion with other space occupying lesions such as brain abscess or tumors.[5,6] In such situations, MRI including spectroscopy and diffusion sequences appear to be the tool of choice to confirm diagnosis of atypical or complicated hydatid cysts and to plan surgery.[2-4] The classic MRI appearance of cerebral cystic echinococcosis is a spherical or oval and well-defined, smooth, thin walled, homogeneous CSF-isointense cystic lesion, which may reach up to 15 cm in size.[7] The cyst wall usually shows a rim of low signal intensity on both T1- and T2-weighted images. Cerebral edema around the cyst is not a common finding, but may occur. A thin rim of contrast enhancement limited to the capsule region may be detected.[7]

Indeed, the presence of rim enhancement and peripheral edema was reported in complicated/infected cases.[2,3,8] In fact, infection of a preexisting intracranial hydatid cyst is a rare phenomenon.[8] The mechanism of this suppuration is still unknown and the source of infection is usually a coexisting bacterial infection with secondary hematogenous spread or contiguous contamination.[8] In our case (patient 2), the primary focus of infection could have originated from pansinusitis. This pansinusitis was controlled by antibiotics before hospitalization, and this could explain the macroscopic aspect of the liquid at the first trepanopunction and the absence of germs on bacteriological analysis of the liquid.

El-Shamam et al[4] classified hydatid cysts based on the presence or absence of perifocal edema and contrast enhancement. They sorted them as simple (non-complicated) cysts while they do not exhibit perilesional edema or contrast enhancement, and as infected (complicated) whenever cysts present perifocal edema and contrast enhancement. Their series of 16 patients included 4 cases of proven infected cerebral hydatid cyst, and 6 recurrent cysts, which occurred after rupture of the primary cyst at surgery.

In cerebral hydatid disease, calcifications of the wall are rare, being less than 1%. Two types of calcifications were described, intra or extramural. Calcification of the hydatid cyst represents a sign of inactivity of the parasite.[9] Pathophysiological mechanisms contributing to calcifications are still unknown; they might be explained by re-absorption of the liquid of the cyst, or by an intense gliosis around the cyst. Either way, these mechanisms cut the food supply to the cyst and consequently cause the death of the parasite. In calcified cerebral hydatid cysts, differential diagnoses may include several lesions, particularly tuberculoma, oligodendroglioma, cavernoma, and calcified arterio-venous malformation.[6] In these cases, MRI has an essential role, not only in specifying the anatomical relationship of the lesion and by eliminating other differential diagnoses, but also by demonstrating the typical image of unstuck membranes with scalloped outlines.[9]

Complete surgical removal of the cyst is the treatment of choice. However, the applicability of the Arana-Iniguez method[1] (Dowling's water dissection technique) to remove the cyst as a whole and to preserve integrity is difficult to achieve in complicated cases. In our opinion, medical treatment is indicated in the postoperative stage to reduce the risk of recurrence.[10] Nevertheless, controversies persist concerning the dose and duration of this treatment. Albendazole, mebendazole, and praziquantel have been shown to be effective in the medical treatment of hydatid cysts of the liver and abdomen. However, limited data have been published on the medical treatment of intracranial hydatidosis, especially drug penetration across both the blood-brain barrier and the membrane of hydatid cyst.

In conclusion, hydatid disease is a worldwide zoonosis produced by ingesting the larval stage of the echinococcus tapeworm. It remains a major health, and economic problem in developing countries due to its widespread endemic nature. Diagnosis is usually made preoperatively basing on anamnesis and pathognomonic CT scan pattern. The possibility of intracranial hydatid cysts should always be kept in mind whenever an unusual CT finding is encountered, especially in endemic areas.

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