Cerebral hydatid cyst in children : A case report

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ABSTRACT

Cerebral hydatid cyst is rare (2%), and mainly affects children. We report the case of a three-year-old patient. The clinical symptomatology was progressive, preceding the hospitalization of one month, dominated by headache and vomiting. The patient presented a tonicoclonic hemicorporeal seizure at the pediatric emergency. The brain CT scan and magnetic resonance imaging evoked a left parietal non-pyogenic abscess. The diagnosis was evoked intraoperatively and confirmed by anapath examination. Extracerebral localizations were sought. The patient also had pulmonary and hepatic hydatid cysts. Hydatid serology was positive. The treatment was surgical (puncture-aspiration technique) with a simple surgical suite.

Key words : cerebral hydatid cyst, child.

INTRODUCTION

Cerebral hydatid disease is a very rare parasitosis but it remains frequent in the Mediterranean basin [1]. In Tunisia, hydatidosis is a public health problem [2]. The most affected organ is the liver, the cerebral localization is rare and does not exceed 2%, it essentially affects children [1,2]. Through this pediatric observation of cerebral hydatid cyst, we report the clinical, radiological and evolving aspects of this disease.

CASE PRESENTATION

This is a three years old boy. He lived in a rural environment and had dogs and sheep in his entourage. The clinical symptomatology was progressive preceding the hospitalization of one month and was dominated by a headache associated with vomiting. The patient presented a right hemicorporeal tonicoclonic seizure on the day of admission. The eye fundus was normal. Brain CT scan showed a lesion with a majority fluid component measuring 45 mm in long axis, peripherally enhanced, surrounded by perilesional edema and transcallous infiltration with some calcifications and mass effect on the surrounding structures. Magnetic resonance imaging (MRI) evoked a left parietal non-pyogenic abscess. The diagnosis was evoked intraoperatively and confirmed by anapath examination. Extracerebral localizations were investigated by thoracoabdominal CT scan, the patient also had two pulmonary hydatid cysts (the largest measured 11mm in long axis) and three hepatic hydatid cysts (the largest measured 10mm in long axis). Hydatid serology was positive. The treatment was surgical (by puncture-aspiration technique) for the cerebral hydatid cyst with a simple postoperative course. The patient has been put on oral medication (albendazole).

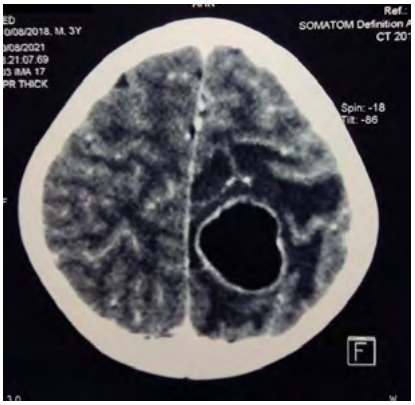

Figure 1 : Axial section of a cranioencephalic CT scan with contrast injection revealing the lesion with significant perilesional edema.

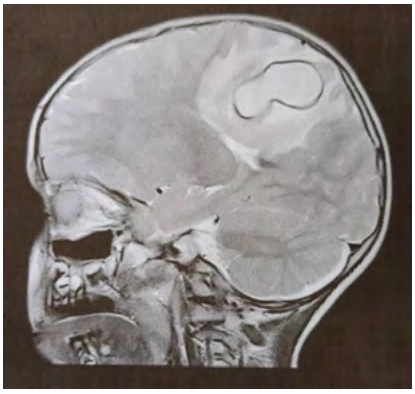

Figure 2 : Left paramedian slice of a brain MRI in sagittal T2 sequence revealing an expansive process (hydatid cyst) in frank T2 hypersignal, well limited by a capsule in hyposignal with important perilesional extended subcortical edema.

DISCUSSION

Hydatidosis is a public health problem in Tunisia. It is a reportable disease. The Mediterranean basin is currently the largest outbreak of hydatidosis; its infestation index in developing countries varies between 5 and 18 cases/100 000 inhabitants per year [1]. It is known that the distribution of hydatidosis in the world varies according to the intensity of sheep farming, our patient was of rural origin and had dogs and sheep in their environment. Hydatidosis is secondary to infestation of the body by the hexacanth embryo of Echinococcus granulosus [2]. The most common sites of hydatidosis are the liver (48%) and the lungs (36%), whereas the brain is rare and occurs in less than 2% of cases. It is more frequent in children and young adults (50 to 70% of cases) and is most often unique [1]. The rarity of cerebral localization of echinococcosis may be explained by the passage of the parasite through two filters (hepatic then pulmonary) before reaching the general circulation [3]. Cerebral localizations are usually hemispheric supratentorial and subcortical, particularly in the territory of the middle cerebral artery in the parietal lobe [1,2]. Rare cases of intraventricular and posterior fossa localizations have been reported [1,2]. In our case, the hydatid cyst was located in the parietal lobe. The cerebral hydatid cyst is most often located on the left side (this is the case of our patient), in fact, the direct origin of the left common carotid artery of the aorta makes it easier for the embolus to migrate directly to the brain. The cerebral hydatid cyst is often unique, multiple localizations are rare and are generally the consequence of a spontaneous or intraoperative rupture and sometimes of massive embolization from a ruptured cyst in the left ventricle. In 10% of cases, the cerebral localization is associated with other visceral localizations, notably pulmonary and hepatic [1,2]. These locations must be systematically sought by chest radiography and abdominal ultrasound. The delay between the onset of the symptomatology and hospitalization was two weeks for our patient. At diagnosis, the cyst was 45 mm in size. Indeed, the cerebral hydatid cyst develops slowly in such a way that the collateral circulation will be able to palliate the risk of ischemia [4]. This slow development explains the fact that cerebral hydatidosis can reach a considerable volume before giving clinical manifestations [5,6]. The clinical progression of the disease in adults is more rapid than in children, which is explained by the inextensibility of the cranium in adults [7]. Clinical symptomatology is polymorphic and dominated by intracranial hypertension syndrome and neurological deficits. Seizures and visual disturbances are less frequent [8]. Physical examination may show an increase in head circumference in infants, motor deficits, and involvement of the cranial pairs. The eye fundus often shows papilledema and optic atrophy exceptionally [1,2]. Cerebral CT scan remains the exam of choice and of first intention in the diagnosis of cerebral hydatid cysts; it allows to specify the location, number, volume and content of the cyst as well as its relationship with the adjacent structures, thus helping in the choice of the surgical approach [1,3]. In the majority of cases, the scannographic aspect is typical in the form of a single, spherical or oval formation, located in the middle of the brain parenchyma, large in size, with a thin and regular wall and having the same density as the cerebrospinal fluid (CSF). It exerts a mass effect on the medial structures and lateral ventricles without contrast and without perilesional edema [1,2]. The quasi constant absence of peripheral enhancement by the contrast product is explained by the thinness of the pericyst in the brain and of the hydatid membrane adhering to this pericyst [3]. It is not possible to distinguish on Tomography between pericyst and cystic membranes unless, exceptionally, fragments of the proligeral membrane become floating when the cyst cracks [2]. Visualization of a floating membrane is pathognomonic and calcifications are extremely rare, less than 1% [1,2,5]. In view of these typical aspects, it is generally easy to eliminate other pathologies. On the other hand, the passage of hydatid fluid into the brain tissue will cause an inflammatory reaction and thickening of the pericyst. Thus the existence of edema and contrast indicate the lack of sealing linked to the fissuring of the cyst [2]. In our patient, the radiological diagnosis was difficult in view of the existence of enhancement in the periphery of the cyst surrounded by perilesional oedema, which was secondary to the fissuring of the hydatid cyst. The differential diagnosis can be made with certain cystic lesions, in particular arachnoid, leptomeningeal, epidermoid cyst, porencephalic cavity, cystic astrocytoma, craniopharyngioma and brain abscess, but in endemic regions, this diagnosis is evoked immediately [3]. MRI currently offers not only additional diagnostic information of cerebral hydatid disease but also allows for more appropriate treatment planning. It reveals a spherical fluid formation, with fine contours, containing a fluid with the same imaging characteristics as CSF [1], hypointense in T1 sequence and hyperintense in T2 sequence with a very thin wall (pericyst) in relative T1 hypersignal and characteristic T2 hyposignal [2]. The absence of the signal on Flair sequences and the hyposignal in diffusion also characterize the hydatid cyst. The relative hypersignal of certain cystic contents in T1 would be related to the existence of hydatid sand.

The absence of perilesional edema and contrast enhancement of uncomplicated cysts is even more evident on MRI [3]. CT scan is superior in the detection of calcifications [1]. Cerebral localization is poorly immunizing. In practice, hydatidosis serologies are less performed because of their low reliability [1]. They are recommended in case of diagnostic doubt. Thus, hydatid serology is negative in more than 80% of confirmed cerebral hydatidosis, and its negativity does not eliminate the diagnosis, which is essentially based on imaging and especially CT. In case of a positive hydatid serology, it is necessary to look for an associated visceral location [9]. For our patient, the treatment of the cerebral hydatid cyst is surgical. Its aim is to excise the whole cyst without causing its rupture in order to avoid dissemination of the scolex, the risk of recurrence and anaphylactic reactions which would be responsible for circulatory collapse and cardiac arrest, thus guaranteeing a definitive cure [2]. The most widely used surgical technique is that described by Arana Iniguez and consists of delivery of the cyst by hydro-dissection using hypertonic saline [2]. This technique is the most widely used and is performed through a flap centered on the cystic lesion followed by the introduction of two Nelaton probes between the cyst and the brain parenchyma. Injection of physiological saline under high pressure allows the cyst to be detached, and the residual cavity is then washed with hypertonic saline [1]. The second possible technique is puncture-aspiration (this is the case of our patient), it is less used and reserved for cysts with a high risk of rupture such as cysts of the fourth ventricle, cysts of the brain trunk and thalamus [9]. The main problems of surgical treatment are the site of the cyst, the number and the risk of intraoperative rupture which occurs in 16 to 25% of cases according to different studies [1], this rupture may be complicated by anaphylactic shock and secondary dissemination. Treatment with benzimidazoles (albendazole and mebendazole) has been used by some teams in cases of recurrent, disseminated hydatidosis, judged inoperable or ruptured intraoperatively. Albendazole is characterized by better digestive absorption, higher blood and intracystic levels, and conversion to an active metabolite that also has a good intracystic concentration [10]. Albendazole administration is rarely associated with major side effects [10]. The results of drug treatment of hydatid cysts remain variable according to the series, with a response rate ranging from 43.5 to 80%. This variability is probably due to the different composition of the series, the different treatment regimens applied and the choice of evaluation criteria [10]. The world health organization has recommended a minimum of 12 months for an objective evaluation [1]. The prognosis is variable, it is conditioned by the localization and number of cysts and also depends on the occurrence of postoperative complications such as: meningitis, subdural hematoma, intracerebral hemorrhage, infection of the residual cavity, epilepsy, visual disturbances and recurrence. The recurrent hydatid cyst is usually multiple with a thicker membrane and faster growth [1,2].

CONCLUSION

Cerebral hydatidosis is a rare condition that mainly affects children. The diagnosis of cerebral hydatid cyst must be evoked in endemic countries in front of a symptomatology of intracranial hypertension. In the majority of cases, CT scan allows a definite preoperative diagnosis. However, MRI offers a better topographic delimitation, especially in multiple locations. The prognosis is generally good if the diagnosis is made rapidly leading to early treatment, thus avoiding neurological sequelae. The key word in management is prevention.

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