Multiple giant hydatid cysts of the brain

Case report

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A rare case of multiple primary hydatid cysts of the brain is reported in a 9-year-old girl. There were five cysts, occupying most of the right supratentorial region. The biggest cyst measured 9 cm across, while the smallest one was 4.5 cm in diameter. The diagnosis was based on computerized tomography findings. The patient did not have any evidence of hydatid disease elsewhere in the body. The delivery of all the cysts resulted in the dramatic neurological recovery of this patient.

KEY WORDS • computerized tomography • hydatid cyst • multiple cysts • brain cyst • Echinococcus

Multiple hydatid cysts of the brain are rare; to date, only nine such cases have been encountered. In all of the reported cases, multiple cysts have resulted from rupture of a primary (mother) cyst. As far as we know, there is no previously documented case of multiple primary giant hydatid cysts of the brain. We have recently treated such a case in a 9-year-old girl.

Case Report

This 9-year-old girl presented with a history of intermittent headache for 2 years, focal seizures involving the fingers of the left hand for 10 months, and abnormal posturing of the left upper limb for 9 months. During the 7 months before admission she developed progressive left hemiparesis.

Examination. The patient was a well nourished girl. The right parietal region of the skull was bulging, and tympanic note could be elicited on percussion. Neurological examination revealed bilateral early papilledema, left central facial nerve palsy, and clawing of the left hand and foot. There was a spastic left hemiparesis with Grade 3 motor power and brisk deep tendon reflexes. She exhibited dystonic movements involving the left upper limb. Laboratory studies were within normal limits.

Skull films demonstrated thinning of the skull vault on the right, evidence of raised intracranial pressure (ICP), and localized bulging of the right parietal region. Computerized tomography (CT) showed multiple large cystic lesions with clearly defined borders occupying almost the entire right supratentorial region (Fig. 1). There was marked compression of the right cerebral hemisphere and displacement of the ventricular system to the opposite side. The lesions contained a fluid with a Hounsfield unit value similar to that of cerebrospinal fluid (CSF). There was no enhancement after contrast injection.

Operation. A preoperative diagnosis of multiple hydatid cysts of the right supratentorial region of the brain was made. A large bone flap was turned, the

Fig. 1. Computerized tomography scans showing multiple hydatid cysts occupying the right supratentorial region, causing a shift of the ventricular system to the opposite side. There was no enhancement with injection of contrast medium.
The largest cyst measured 9 cm across, while the smallest was 4.5 cm in diameter; one measured 7 cm, and two 5 cm. The cysts were aspirated and removed using carefully controlled suction, without any spilling of their contents. The remaining cyst was finally delivered by saline irrigation, assisted by changing the position of the patient's head. In total, five giant cysts were delivered intact. After the removal of the two superficial cysts, three deeply placed cysts were exposed. As the deeper cysts were tightly surrounded, dissection using saline irrigation could not be carried out. Other methods described for the delivery of cysts were not considered because it was feared that the cysts might rupture during delivery. Two cysts were aspirated and removed using carefully controlled suction, without any spilling of their contents. The remaining cyst was finally delivered by saline irrigation, assisted by changing the position of the patient's head. In total, five giant cysts were removed. The largest cyst measured 9 cm across, while the smallest was 4.5 cm in diameter; one measured 7 cm, and two 5 cm.

Pathological Examination. The cysts were pearly white. Their outer surface was smooth and the inner lining was finely granular. Microscopic examination showed a characteristic laminated ectocyst which was lined on the inner surface by an endocyst composed of flattened germinal epithelial cells, brood capsules, and scolices. The lumen showed several brood capsules with scolices, confirming that all the cysts were primary hydatid cysts of the brain.

Postoperative Course. The postoperative course was uneventful, and the patient showed marked recovery in her neurological status. Motor power improved to a nearly normal level. Detailed investigations failed to reveal any evidence of hydatid disease elsewhere in the body.

Discussion

Hydatid cysts of the brain are unusual: the brain is involved in only 2% of all cases of hydatid disease. These cases are seven times more common in children than in adults. Multiple hydatid cysts of the brain are rare, and as far as we know multiple primary hydatid cysts of the brain have not been described before. In a review of 112 children with cerebral hydatid cyst, Carrea, et al., found only four cases of multiple cysts in the brain. Only two of the 29 patients with hydatid cysts of the brain reported by Carrea, et al., had more than one intracranial cyst. One of these two patients had one cerebral and one cerebellar acephaloceles (without scolices), and the second had a calcified parieto-occipital cyst (a dead cyst) with multiple secondary acephaloceles. The cysts are always solitary when the primary localization is in the brain. Multiple cysts have been reported in the brain when a cyst that was attached to the wall of the left ventricle of the heart ruptured, and following a head injury with rupture of a viable primary cyst of the brain. The presence of multiple primary hydatid cysts in the present case, with no history of trauma to the head or rupture of cysts elsewhere in the circulation, is most unusual.

In most of the reported cases, an associated cyst could be demonstrated elsewhere in the body. According to Dew, 80% of cerebral hydatid cysts are associated with cysts in the liver. The location of all the hydatid cysts in the right supratentorial region in the present case may be explained by all the hydatid larvae taking the same path. The multiple hydatid cysts resulting from the rupture of a primary (mother) cyst are acephaloceles and are termed “secondary cysts.” They lack a brood capsule and are infertile. All the cysts in the present case were primary cysts because they contained brood capsules and scolices.

Cerebral hydatid cysts are often very large, especially in children. The slow compression of surrounding tissue without invasion explains why these space-occupying lesions are tolerated for a long period. The lack of early symptoms is also due to the remarkable tolerance of the brain and the expanding properties of the skull in children. The majority of cases present with raised ICP, with or without visual symptoms. Patients with this disease rarely exhibit focal neurological deficit initially.

Although our patient was in good general condition, she had raised ICP and focal left hemiparesis. Roentgenograms of the skull showed signs of raised ICP and thinning of overlying bone. Neither skull films nor angiography can be relied on in the differential diagnosis of cases of multiple hydatid cysts, but CT scanning has been found to be valuable. The CT scans showed multiple large intraparenchymal cystic lesions with clearly defined borders containing a fluid with a Hounsfield unit value similar to that of CSF. There is no ring enhancement. Since there is no previous report in the literature of the CT finding of...
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Multiple giant hydatid cysts of the brain, we presume these features are characteristic. These cysts can be differentiated from brain abscesses by their lack of significant ring enhancement and absence of perifocal edema. Arachnoidal cysts can be differentiated by their shape.

The treatment of hydatid cysts is surgery, with two basic aims: the cyst must be removed en masse, and dissemination of the hydatid fluid should be avoided. Although the superficial cysts can be removed by the “hydatid birth” technique via an episiotomy of the cortex and using the Dowling technique, the intact delivery of the deeper multiple cysts is a difficult problem, as seen in our case.

References


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