



## Primary hydatid disease of the occipital bone presenting as space occupying cystic mass of the posterior fossa

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### ABSTRACT

Brain involvement of hydatid disease accounts for 1 to 4% of all cases. Extra-Dural, intra-calvarial location is even less. We report, a 24 years old woman with primary intra-calvarial hydatid cyst that presented as an extradural cystic mass lesion of the posterior fossa. In the case presented, removal of the pathology without rupture could not be achieved. As same with the other locations, hydatid cysts of calvaria must be tried to be removed without rupture when possible. However, this exceptional host, the calvaria, seems to ease the pre-mature rupture of the cyst due to the neighboring sharp edges of the bone. In such situations, excessive irrigation with hyper-tonic solutions, %3 NaCl for example, is known to be effective for preventing dissemination. Pre-operative diagnosis is very important for surgical planning. Extra-dural locations of cystic lesions must remind the surgeon hydatid cysts especially in the countries where the disease is more common. *J. Exp. Clin. Med.*, 2012; 29:77-79

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### 1. Introduction

Hydatid disease is a worldwide zoonosis. *Echinococcus granulosus* and *Echinococcus multilocularis* causes the two common types of hydatid disease, cystic and alveolar echinococcosis respectively (McManus et al., 2003; Beskonakli et al., 2005). Unlike *E. granulosus* which produces cystic lesions, *E. multilocularis* produces invasive solid lesions, full of connective tissue and a jelly-like material (Oktar et al., 1999). Although hydatid cyst of *E. granulosus* may indwell in any organ of the human body, the most affected ones are liver and lung (McManus et al., 2003; Beskonakli et al., 2005).

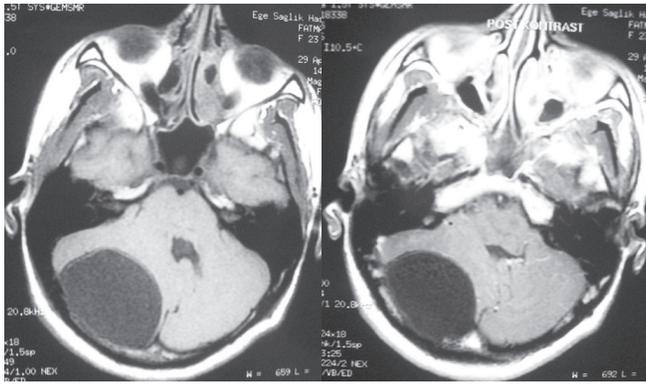
Cerebral involvement of hydatid disease is reported only up to 4% of cases (Ersahin et al., 1993; Beskonakli et al., 1996; Erman et al., 2001; Beskonakli et al., 2005). It accounts for 0.02% to 0.22% of all intracranial space-occupying lesions (Furtado et al., 2009). Beskonakli et al. (2005) classifies the intracranial hydatid disease in three forms which are consisted of intracerebral, extra-cerebral and combined. In this

paper we report, a 24 years old woman with primary intra-calvarial hydatid cyst which fulfills the pure extra-cerebral form. In the case presented, removal of the pathology without rupture could not be achieved.

### 2. Case

Twenty four-year-old woman was admitted to a physician with the complaints of insisting headache and newly onset of dizziness. When the cranial Magnetic Resonance Imaging (MRI) taken on the order of the first physician revealed a cystic mass of posterior fossa, the patient was immediately referred to our department. On the admission her neurological examination revealed deteriorated cerebellar tests with a mild truncal ataxia. Papillary stasis was seen on fundoscopic examination bilaterally. Besides these pathological findings, patient was found to be fully conscious with a score of 15 in Glasgow Coma Scale (GCS). Her headache was prominent especially in the mornings with a new onset of dizziness.

Cranial pre-and post-contrast MRI suggested an extra-axial location of a large cystic mass with fine contours (Fig. 1a, 1b).



**Fig. 1.** T1 weighted pre- (a) and post- contrast (b) axial MR images depicting fine contoured extra-axial cystic mass lesion in the posterior fossa on right cerebellar cortex. Cystic walls are faintly enhanced after gadolinium administration (b).

An operation was carried out on a routine scheduled day for total resection of the cystic mass. Head was put on a three-pin holder and the patient was positioned prone. A right paramedian sub-occipital approach was planned. Soon after the skin incision the exposed occipital bone depicted an area of erosion about 3x3 cm with a membrane shining under tension through the bone deficit. Despite the extreme attention for keeping the cyst un-ruptured, during the excision of the bone flap, membrane was punctured. Content of the cyst with its membrane defined the pathology as hydatid cyst. All the mass was located at extra-dural space that originating from the occipital bone. Immediately after diagnosing the disease, excessive irrigation with hypertonic saline was performed. Meanwhile albendazole was made ready in the operation room and finally it was used for extra-dural irrigation. Rest of the operation was performed in regular fashion with bleeding control and closure of the layers properly. Patient was discharged after three more days of hospitalization with an uneventful recovery. She was started albendazole 400 mg/q 12 h per oral before discharge and her further medication and follow-up was planned with collaboration of the infectious disease department.

### 3. Discussion

Infestation with the tapeworm *E. granulosus* and less frequently, *E. multilocularis* causes echinococcosis (hydatidosis) (Ciurea et al., 2006). The disease is reported to be endemic in South America, Australia, India, northwest China, Mediterranean and the Middle East (Ersahin et al., 1993; McManus et al., 2003; Ciurea et al., 2006). But it is possible that it may be seen more often in the western countries in near future because of increasing rates of migration and travelling. The incidence of the disease ranges from 3 to 50 cases per 100,000 inhabitants in endemic areas (Krajewski and Stelmasiak, 1991; Ciurea et al., 2006). Highest rates are among the communities involved in sheep farming where the sheep strain of *E. granulosus* in transmission to people (McManus et al., 2003). Lungs and liver are the most common sites for the hydatid cysts and cranial manifestation is rare and accounts less than 4% of all cases (Ersahin et al., 1993; McManus et al., 2003; Ciurea et al., 2006; Furtado et al., 2009; Is et al.,

2009). When cysts occur intracranially, they are usually unilocular. They prefer intracerebral and supratentorial localization in the distribution of the terminal branches of the middle cerebral artery, usually temporal and parietal lobes (Ersahin et al., 1993; Erman et al., 2001; Ciurea et al., 2006; Kayaoglu, 2008; Is et al., 2009). Hydatid cysts are scarcely located both in the calvaria and the posterior fossa (Erman et al., 2001; Polat et al., 2003; Kayaoglu, 2008; Is et al., 2009). Only a few extradural posterior fossa cysts are reported in the English literature (Beskonakli et al., 1996; Erman et al., 2001).

The initial phase of the primary infection is always asymptomatic. Small cysts may remain asymptomatic for many years. The incubation period of cystic echinococcosis may vary from many months to years and is not known clearly. The infection may be manifest by rupture of the cysts or the mass effect (McMnaus et al., 2003). But cerebral location of the disease probably causes an earlier onset as 50 to 75% of cerebral hydatid cysts occur in childhood (Ersahin et al., 1993; Is et al., 2009).

Although radiological investigations are very helpful in identifying hydatid cysts preoperatively the certain diagnosis remains to be a pathological one. Computerized Tomography (CT) and MRI are the current diagnostic modalities of choice. MRI appears to be superior to CT in the detection of rim enhancement and surrounding edema in the brain parenchyma (Ciurea et al., 2006). The role of diffusion weighted MRI in the diagnosis of cerebral hydatid disease is currently under investigation (Kitis et al., 2004; Ciurea et al., 2006). According to Kitis et al. (2004) cerebral *E. granulosus* lesions, unless complicated, reveal CSF-like MR signal intensity on diffusion weighted intensity. CT is advantageous in rare cases of hydatid cysts located at the calvaria. It is expected to depict the classical findings of intraosseous solitary cystic lesion with regular contours (Erman et al., 2001). Lack of pre-operative CT scan probably diminished the chance of pre-diagnose in our case. But its existence naturally do not guarantee the exact diagnose prior to the operation as eosinophilic granuloma, cystic fibrous dysplasia and calvarial epidermoid tumour must be considered in the differential diagnosis of calvarial hydatid cyst (Erman et al., 2001).

Certain treatment of hydatid cyst is total resection avoiding rupture (Lunardi et al., 1991; Ersahin et al., 1993; Erman et al., 2001). Dowling has defined and named the technique himself, as giving birth to the intracerebrally located cysts by continuous irrigation with hypertonic saline between the cyst and the brain tissue (Carrea et al., 1975). But in rare localization of calvaria, no definite techniques are defined. Principally a large craniectomy surrounding the eroded center of the calvarial bone sounds to be effective whilst in practice all the cases are reported to be early ruptured before the excision. Early rupture may be a resultant of sharp bony edges surrounding the defect eroded by the cyst and its adherence to the dura and osseous trabeculae. In case of early rupture intraoperatively, extensive irrigation of the site with a hypertonic solution is strongly recommended (Erman et al., 2001).

Obstructive hydrocephalus and subdural effusions are well known post-operative complications (Beskonakli et al., 2005; Ciurea et al., 2006). In one of the recent and largest series, Ciurea et al., (2006) report a rate of 36.8% for post-operative subdural effusion. Seizures, pencephalic cysts and non-bacterial meningitis are among other post-operative

complications reported in the literature (Lunardi et al., 1991; Beskonakli et al., 2005; Ciurea et al., 2006). Rupture of the cyst during surgery may cause anaphylaxis and mortality in the per-operative period (Ersahin et al., 1993; McManus et al., 2003). Our case was discharged after an uneventful recovery and early post-operative CT-scan revealed no pathologies secondary to the operation.

We conclude that rare phenomenon of intra calvarial and

extra-dural hydatid cysts must be kept among the differential diagnosis of cystic masses of this location. Although un-ruptured excision of these cysts is primary goal, this unique location surrounded by sharp bony edges seems to be obstructing this aim. In case of pre-mature rupture, excessive irrigation with hypertonic saline and albendazole is recommended. Anti-echinococcal medical therapy must be continued post-operatively.

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