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Giant Multiple Skull Hydatidosis in a Child: A Case Report & Review of the Literatures

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Abstract— Background*: Cranial hydatidosis is a relatively rare disease, accounts for 3% of hydatid disease, most of the times, cyst(s) is (are) confined to the white matter, in the supratentorial regions, esp. the parietal lobe being the most common involved part i.e. in the vascular territory of middle cerebral artery. Overall, extraaxial hydatid cysts occur in a rare manner, esp. skull hydatidosis which is reported very scarcely, makes up 3-4% of bone hydatidosis.* ***Method:*** *In our case, we represent a single case of giant multiple skull hydatid cysts (> 100) occupying most of the left hemisphere and discuss its delayed pattern of presentation, radiological features emphasizing on its erosive nature, operative procedure & its nuances, outcome and some effects of rapid decompression of chronically compressed brain. The literature of both skull & epidural cerebral hydatid cyst were reviewed.* ***Conclusion****: Skull hydatids, are very rare, can be multiple, multilocular, can reach a huge size, & can cause sever bone erosions before the patient seeks medical help. Surgical therapy is the mainstay modality*.

Keywords— *Skull hydatidosis, Echinococcus, radiological features, treatment*.

I. INTRODUCTION

Hydatid cyst (HC) is an endemic zoonotic parasitic disease caused by *Echinococcus* genus, among them, *E. granulosus* is the most common infectious species in humans [1, 2]. Skull involvement is extremely rare, constitutes 3-4% of bone *hydatidosis* [3]. Peds account for 70-80% of the cases affecting CNS. In this paper, we report a case of secondary giant multiple skull *hydatid cysts* manifested by symptoms of raised intracranial pressure, left sided head deformity, & multiple soft cystic scalp bulges. The objective of this paper is to highlight the importance of demography & radiological images in diagnosis and perioperative & intraoperative measures that can reduce recurrence, mortality, & morbidity.

II. CLINICAL PRESENTATION

11 year old Bedouin female, who lives in Jordan south desert, was referred to our clinic with a 4-year history of headache & nausea, that became more severe and associated with a slowly progressive decreased level of consciousness, intermittent vomiting & low grade fever over the last few days before referral, there was a history of contact with dogs & sheep. On physical examination, she had a GCS score 14/15, left sided head deformity, multiple soft cystic bulges can be felt under the scalp, slight conductive hearing on the left side, otherwise, examination revealed nothing. Preoperative audiotympanometry showed left sided conductive hearing loss with air bone gap & *Carhart* notch negative pressure, and right sided mild low frequency sensorineural hearing loss with normal tympanic pressure (Fig. 1). Routine hemogram, electrolytes, and liver function tests all were within normal limits. Serology test, unfortunately was not available at that time. *Computed tomography* (CT) scan (Fig. 2) & *magnetic resonance* (MR) (Fig. 3) imaging of the brain, showed a well-defined honeycomb appearance, giant multiple-multilocular skull cysts, with a density & intensity similar to the cerebrospinal fluid (CSF), hypointense cystic wall on T2-weighted images, intense pericystic enhancement, and scattered cysts wall calcification, the cysts were occupying most of the left fronto-temporo-parietal region with extension to the posterior fossa, resulting in a severe brain shift, calvarial & skull base erosions, & cysts protrusion through the skull to under the scalp, orbital cavity, middle ear, facial canal, proximal external auditory canal, & infratemporal fossa. Chest-abdomen *CT scan* & *echocardiography* disclosed only a single liver *hydatid cyst*. The patient was loaded with *Albendazole*, *Hydrocortisone* & anti-epileptics up to her weight prior to surgery. The patient underwent left fronto-temporo-parietal craniotomy, ICP lowering measures was done, and precautions to avoid trans-calvarial protruded parts was taken into consideration during the opening, which was very challenging (Fig.4), unfortunately, the largest mother cyst puncturing was inevitable because it was protruding through the skull, partially calcified, and adherent to skull & dura, thus immediate aspiration & thorough irrigation using *hypertonic saline* (i.e. 3% saline) & *formalin* (1%) were made alternatively, then the remaining daughter cysts (> 100 cysts) which were whitish & transparent, were taken out intact one by one using *Dowling technique* (Fig. 5) i.e. saline was injected under mild controlled pressure into the cyst-dura mater & cyst-cyst interfaces, though some of the very small cysts were taken out by suction. The calcified, adherent wall of the punctured cyst was dissected as much as possible from the surrounding structures (Fig. 6). The cavity then was explored carefully & irrigated thoroughly by hypertonic *saline* & *formalin* alternatively, then filled with cotton soaked by *hypertonic saline* for 30 minutes, space was explored again then filled by *Gelfoam* sponge & *hypertonic saline*. *Redivac*®

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drain was inserted below the skin flap & kept closed for a few days post surgery.

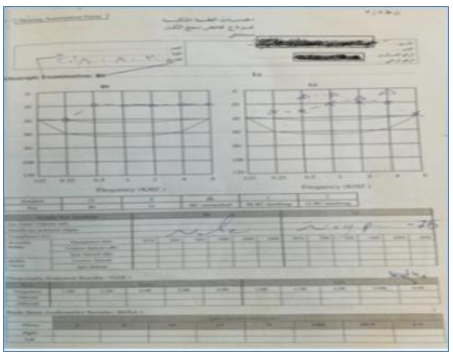

Figure 1 Preoperative audiotympanometry.

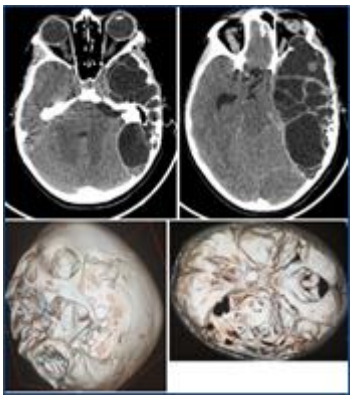

Figure 2: Preoperative CT scan show multilocular skull cysts, with a density similar to CSF & scattered cysts wall calcification. 3D CT scan reconstruction showed the severity & extension of bone erosion.

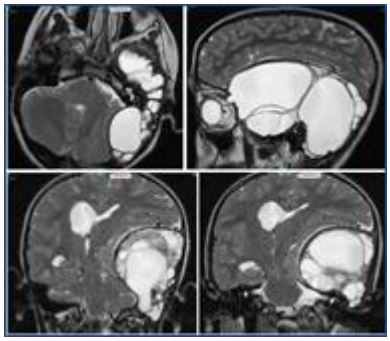

Figure 3: Preoperative MRI showed multilocular epidural cysts that follow CSF-intensity, cysts spread over supratentorial, infratentorial, & extracranial spaces, causing severe brain shift & intense pericystic enhancement.

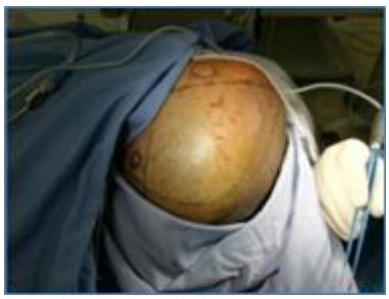

Figure 4: Marking for incision of left craniotomy & the trans-calvarial palpable protruded parts.

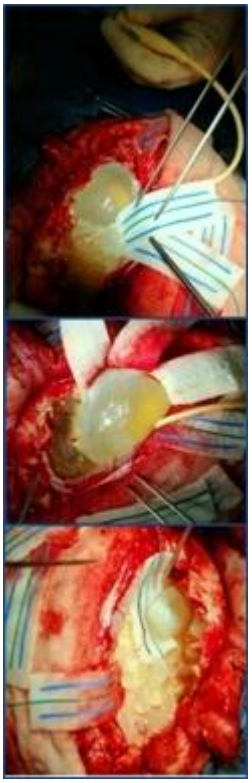

Figure 5: Intraoperative photos shows the enormous number of the cysts & cysts removal using the Dowling technique.

Figure 6: Intraoperative photos show skull base erosions & dissection of calcified adherent cystic wall.

Postoperatively the patient had delayed verbal response, focal right facial fits, few attacks of generalized tonic clonic seizures, & grade 2 left facial paresis (*House-Brackmann* system), all of them resolved totally over one week, except the facial paresis, which exhibits slow partial improvement. Postoperative CT scan serial images showed no remnant cysts but progressive intense ipsilateral hemispheric parenchymal edema that peaked at day 15 post-surgery and subsided totally 2 months after the peak (Fig. 7). Parenchymal edema was successfully treated with *Decadron*® & other ICP lowering measures. Postoperative MRI was done 2 months later showed

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no any remnant cysts but mild ipsilateral parenchymal edema, mildly atrophied brain & enlarged left sided ventricular chambers compared to the contralateral side (Fig. 8). The histopathology of the cysts confirmed the diagnosis of a *hydatid cyst* (Fig. 9). One year later the patient had no remaining symptoms including facial paresis & follow up brain MRI shows no recurrent cysts.

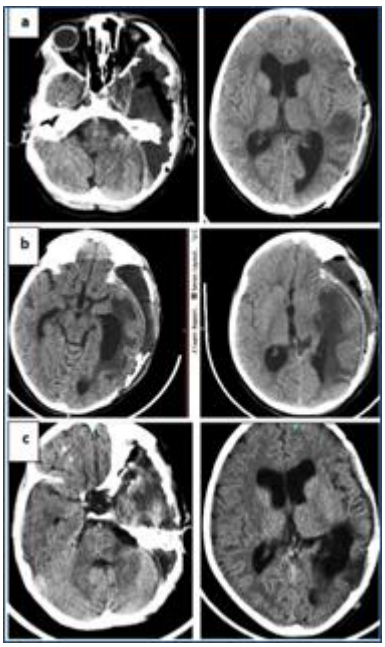

Figure 7: CT scan shows the progressive intensity of post-operative parenchymal edema; (a) POD 1, (b) POD 4 & 15, (c) the intensity decreased significantly 2 months after the peak of the edema.

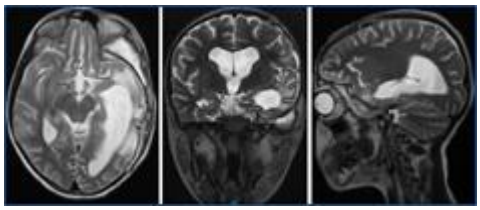

Figure 8: Postoperative MRI at 2 months later shows no any remnant cysts but mild ipsilateral parenchymal edema.

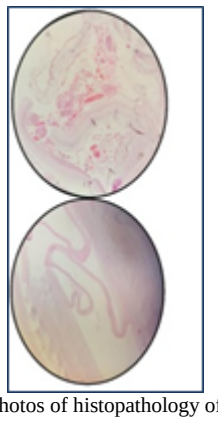

Figure 9: Photos of histopathology of the cysts.

III. DISCUSSION

Hydatid cyst (HC) is an endemic zoonotic parasitic disease caused by *Echinococcus* genus, among them, *E. granulosus* is the most common infectious species in humans [1]. The *larvae* stage is the cause of the disease in humans [2]. The incubation period of the disease may last 5-15 years [4]. Though it is considered common in many regions e.g. Mediterranean region, the Middle East, Africa, the eastern part of Turkey, Australia, and parts of South America [5]; but it may also be encountered in non-endemic countries too, most probably due to the ease of traveling. Humans (the intermediate host) get infected either by eating the contaminated food with ova in the feces of the infected dogs or by direct contact with the infected dogs (the definitive host). HC may occur in any part of the body with the most commonly involved organs are the liver (75%), lungs (15%), brain (2-4%) [6], genitourinary tract (2-3%), and osseous (0.5-2.5%) [7]. Among the space-occupying brain lesions, HC account for 1-5% and 0.2-2% in endemic areas & worldwide, respectively [8]. However, in the Mediterranean region, the incidence may reach 7.4-8.8% [9]. Skull involvement is extremely rare, constitutes 3-4% of bone *hydatidosis* [3]. Peds account for 70-80% of the cases affecting CNS [10, 11], which are more prevalent in boys [12]. Patent *ductus arteriosus* & patent *foramen ovale* have been blamed for the higher prevalence of cerebral hydatid disease among children vs. adults [13]. Cranial HC occurs as one of three forms; intradural, extradural (including intraosseous-skull), or intradural-extradural [14-16]. Central nervous system (CNS) involvement most of the times is confined to the white matter, as either primary cysts which are usually solitary & fertile due to direct invasion of *larva* or as secondary cysts which are more likely to be multiple, infertile & don't have brood capsule, e.g. embolization from a primary source or rupture of a primary cyst in situ [17]. Epidural HC is extremely rare [18] because the blood flow to the brain is mainly through the ICA, so the likelihood of the *larvae* dissemination through the ECA is very low [19, 20]. The involvement of extradural space occurs in either three ways or a combination of them; (1) involvement of bony spongiosa (2) direct embolization via blood vessels (3) extrusion of intracerebral cysts [3]. Epidural HC has a tendency to occur around the midline; this may be related to the pattern of vascularization & presence of rich venous drainage [3, 21]. Calcification of endocysts indicates the non-viability of cysts, in contrary to pericystic calcification, which may be seen at any stage of the cyst [22-24]. Various surgical options were summarized by *Arana-Iniquez*, among them [25], *Dowling-Orlando* technique is the most popular used one, in which the cyst can be delivered by irrigating warm saline simultaneous with lowering the head of the bed. According to *Ameli* & *Turtas*, complete removal of intact cyst(s) is difficult in extraaxial lesions [26-29], owing to the presence of the cysts in the skull trabeculae & their adhesion to the dura, this explaining frequent recurrence. Though surgical therapy is the mainstay of treatment, preoperative & postoperative adjunct medical therapy is also important to prevent recurrences and, in certain circumstances, as the primary mode of treatment [30]. Patients with intraosseous HC should be treated by *Albendazole* for at least three months & followed for up to twelve months before they are considered cured [28]. All measures in case of rupture of the cyst should be prepared

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before incision including 10% *hypertonic saline*, 1% *formalin*, or 0.5% *silver nitrate* solution [31]. In a review of the literature, we found only very rare reported cases of isolated skull HC, but no one reaching a very large size causing severe bone erosion & cysts protrusion through the skull in peds. On our review of literature we failed to find as many cysts being removed from a single patient successfully.

IV. Conclusion

Cranial skull *hydatids* without intraxial involvement are very rare, & can be primary or secondary in nature. CNS involvement has a variety of clinical presentations that can lead to significant morbidity & mortality. Preoperative diagnosis by the correlation between demographic & other patient information, clinical findings, and radiological findings, is very crucial to diagnosis & good surgical planning that definitely will reduce surgical complications including recurrence, widespread dissemination, anaphylactic shock, or even death. Rapid decompression of chronic brain shift may lead to serious complications including severe edema, hyperemia, and cranial nerve paresis/palsy. Inflation of a chronically compressed brain can take a very slow course relatively. Pre- & post-operative *Albendazole* is of high importance in reducing intraoperative complications & postoperative recurrences.

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