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GIANT SOLITARY CEREBRAL HYDATID CYST IN A CHILD, COMPLICATED WITH POST-OPERATIVE SUBDURAL HAEMATOMA: CASE REPORT AND REVIEW OF LITERATURE

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M. A. MAGOHA, N. K. MOHAN, C.K. MUSAU and M.P. OKEMWA

SUMMARY

Hydatid disease is a parasitic infestation caused by taenia *Echinococcus granulosus* that rarely affects the brain. It usually involves all organ systems of the body but is more common in the liver, lungs. We present an eight year old male child presented with a six months history of headache and left sided weakness. After routine investigations including CT scan, craniotomy and cruciate durotomy was done and the hydatid cyst was extirpated whole using Dowling's technique. Post-operative period was uneventful and the child was discharged home to continue on anti-helminthic treatment. He developed bilateral subdural haematomas one month later, which was treated with bilateral burr hole drainage with complete success, and no neurological deficits.

INTRODUCTION

Hydatid disease caused by the larval stage of *Ecchinococcus granulosus*, is endemic to many countries including sub-Saharan Africa and Kenya, where in the Turkana region it is reported to have the highest world incidence of 198 surgical cases per 100,000 people per annum(1). The disease is reported to cause a worldwide annual loss of US\$193,529,740 showing great socio-economic burden.

Cerebral hydatidosis occurs in 2-5% of all forms of hydatid disease in humans. Most commonly seen in children and young adults (50-75%).

We present a child from the endemic region who presented with a giant solitary hydatid cyst and developed bilateral symptomatic subdural haematomas post-operatively as complication.

CASE REPORT

An eight year old male child presented to our neurosurgery clinic with a six month history of left sided weakness and headache. For the past two months he needed assistance in walking and had

episodes of vomiting a week prior. He resides in a rural area in Kenya and the parents are pastoralists and keep cattle, sheep and dogs.

On examination he had normal vital signs. His Glasgow coma scale was 15/15, had a left upper motor neuron facial palsy and left sided hemiparesis worse in upper limbs than lower limbs of power grade three with increased tone and reflexes. Fundoscopy revealed papilloedema. The cardio-vascular, respiratory and abdominal examinations were normal.

The haemogram results were normal revealing no eosinophilia. Erythrocyte sedimentation rate of 20. A computed tomography (CT) scan of the head showed a large, well defined cyst in the right frontal-parietal temporal region which was hypodense and the margins did not take up contrast. There was no surrounding oedema but the right lateral ventricle was effaced with significant midline shift (Figure 1). The impression made from the clinical findings and images were of a cerebral hydatid cyst and the patient prepared for craniotomy.

Figure 1a,b
Pre-operative non-contrast and contrast CT shows well defined cyst non-enhancing cyst that has density like CSF

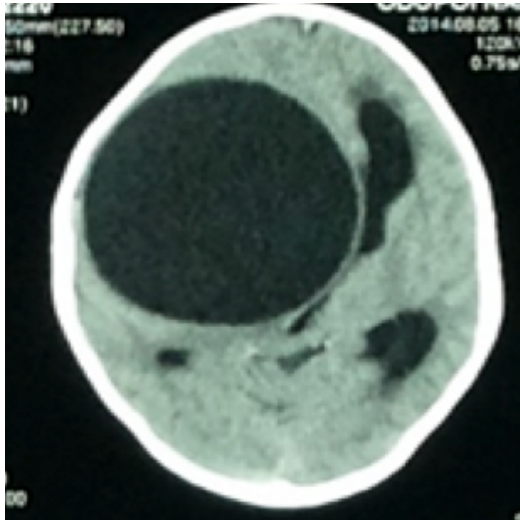
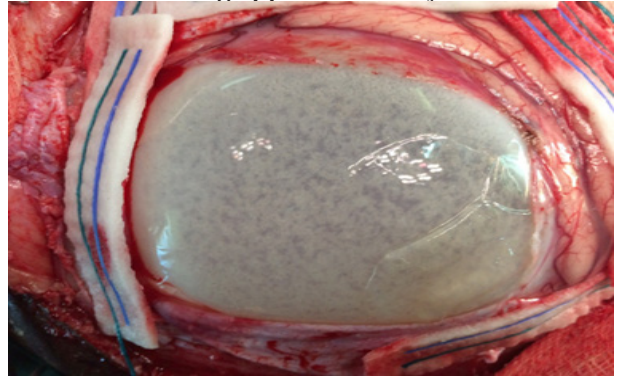


Figure 2
Image shows cyst bulging out soon after dura opened. Large hydatid cyst after dura opening, good plane between cyst and arachnoid seen. Glistening appearance of cyst wall



In supine position and under general anaesthesia, a large right frontal- temporal parietal flap was made and a large bone-flap raised. Cruciate durotomy made and a pearly white cyst wall seen which could be seen bulging just beneath the dura.

The arachnoid plane could be seen between the cyst and cortex (Figure 2). Total extirpation of the cyst was achieved using dowling technique.

Figure 3

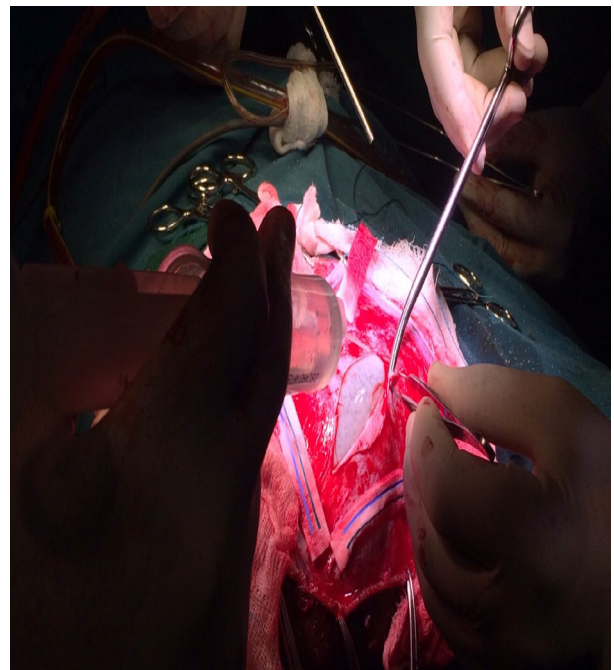
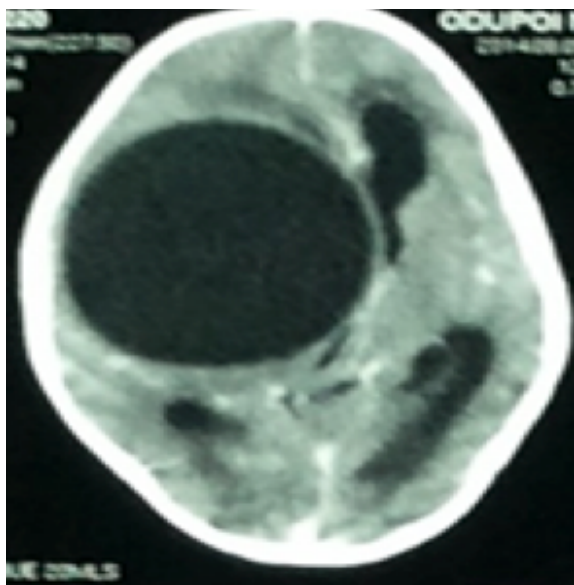
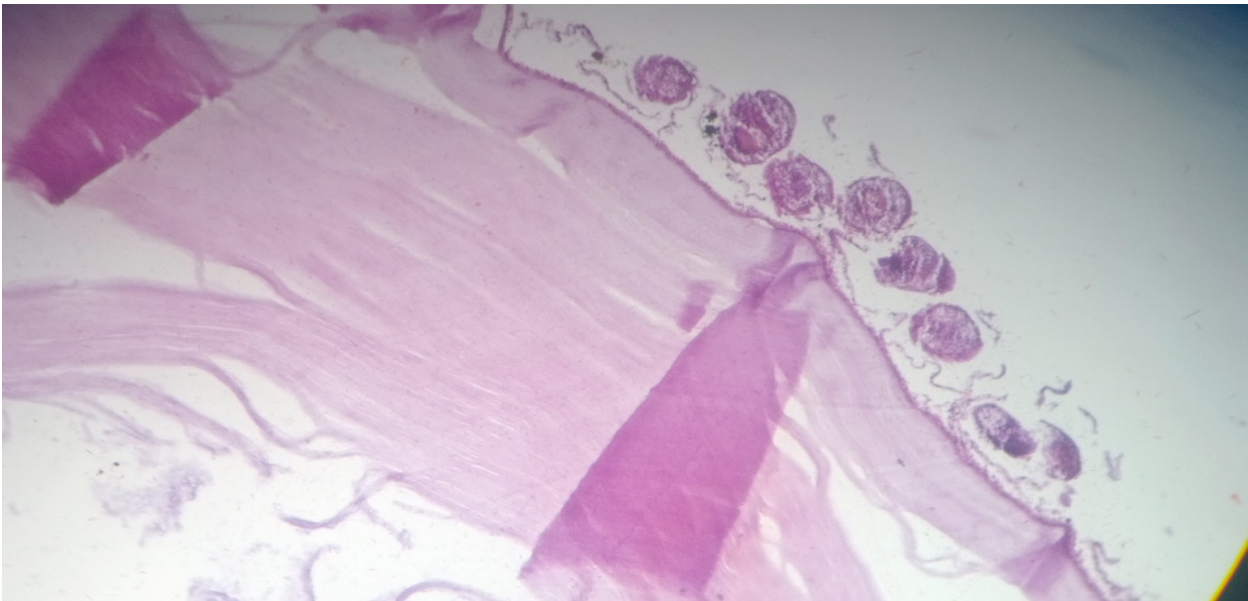


Figure 4
Protoscolices attached to the inner germinal layer and outer fibrous wall (exocyst) shown (magnification X 100)



Immediate post-operative course was uneventful with a repeat CT scan revealing total cyst excision (Figure 5a) but a large volume of negative intra-cranial space occupied by normal saline with neurology intact. The child was allowed home to continue on antihelminthic treatment.

On follow up, one month after, the child was noted to be somnolent with a Glasgow coma scale of 14. Repeat CT scan revealed massive bilateral subdural haematomas (Figure 5b) and bilateral burrhole drainage done. He did well with no neurologic deficits. He continues with albendazole therapy to complete a six month course.

Figure 5a immediate postop axial CT scan showing total excision of hydatid cyst

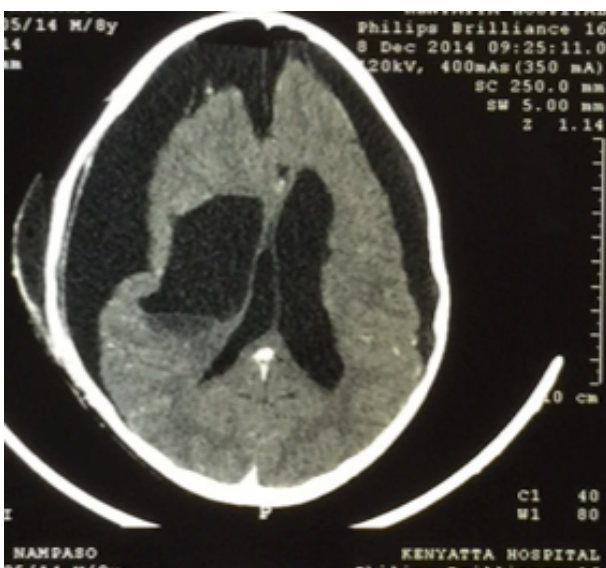
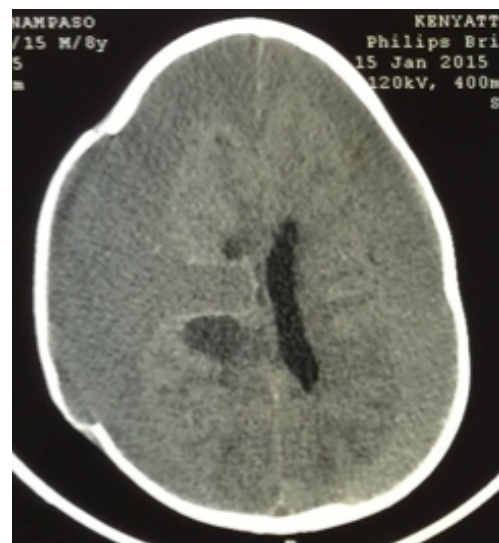


Figure 5b: Axial CT scan showing bilateral subacute subdural haematomas with mass effect causing previous bone flap bulging



DISCUSSION

In 1807, Chaussier reported the first case of cerebral hydatid cyst and Chaussier of vertebral hydatid.

Echinococcosis caused by infestation due to the tapeworm *Echinococcus*. Four *Echinococcus* species have been identified that infect humans; *E. granulosus* and *E. multilocularis* are the most common, causing cystic echinococcosis and alveolar echinococcosis, respectively. The two other species include *E. vogeli* and *E. oligarthrus* are very rare and present only in Latin America. We shall discuss the *E. granulosus* infestation (cystic hydatidosis). Cystic hydatidosis is a significant public health problem in South and Central America, the Middle East, some sub-Saharan African countries, China, and the former Soviet Union. In endemic rural areas prevalence rates of two to six percent was reported.

The definitive host which harbour the adult tapeworms are canines, usually dogs. The intermediate host include sheep, goats, cattle and humans who are incidental hosts and do not play a role in the transmission cycle and get infected by faecal-oral route. Human to human infection does not occur. Transmission frequently occurs in endemic areas where dogs ingest offals of slaughtered sheep, cattle. Humans get infected by environmental contamination of water and cultivated vegetables by eggs excreted in faeces of dogs, or contact between infected domestic dogs and humans especially in children. The small intestine of the definitive host inhabit the adult worms which release eggs (containing oncospheres) in the faeces, each worm producing thousands per day. Eggs can survive in harsh environments for many months in temperatures ranging from -30°C to 38°C. These eggs are then ingested by the intermediate or incidental host (humans) and oncospheres hatch from the eggs. The oncospheres then penetrate the intestinal mucosa, enter the blood and migrate to the liver or other visceral organs. A hydatid cyst then begins to develop and multiple layers form and protoscolices develop within the hydatid cyst. In definitive hosts who ingest intermediate host visceral organs containing hydatid cysts composed of protoscolices, the protoscolices vaginate, attach to the intestinal mucosa and develop into adult worms. The life cycle takes four to seven weeks.

The liver is affected in about 65% of cases, the lungs in about 15–20% and the brain in about 2–5%. Cerebral hydatid cysts are more common in children and young adults with a ratio of 7:1. Cerebral hydatidosis may be more common due to patent ductus arteriosus. They commonly occur in the distribution of the middle cerebral artery, in the parietal lobe and in white matter. Other rare unusual reported sites include infratentorial in the pons, cavernous sinus (11) and diploe of the calvarium (12). Rare case of infected form of hydatid cyst reported (13).

Hydatid cysts maybe single or primary type which is more common or may present as multiple cysts or secondary type cystic lesions can be single (77%) or multiple (23%). Multiple cysts is due to traumatic, spontaneous or iatrogenic rupture of the primary cerebral hydatid cyst or rupture of cyst in the left side of the heart (15). Rarely do multiple hydatid cysts present as primary type. Largest reported cyst measured 12.5cm in diameter. The growth rate been reported from 1.5cm to 10cm per year.

The cyst wall is made up of three layers: inner germinal layer (endocyst) that has attached brood capsules (hydatid sand) containing protoscolices, intermediate layer (exocyst) and the outer fibrous layer (pericyst). The outer layer in cerebral hydatid is very thin unlike in extracranial cases due to little inflammatory reaction. Secondary cysts do not contain brood capsules and are infertile (18). The contents are usually clear or whitish fluid. The cyst wall is well delineated from cerebral normal tissue, does not infiltrate the dura or calvarium. It however causes asymmetry and bulging of the calvarium especially in children due to unfused sutures as is seen in our case. Ten percent of patients with cerebral cysts may have hydatid cysts in other organs especially in the liver and lungs.

The signs and symptoms depend on the location and size of the cyst. Headache, vomiting are the most common symptoms. Papilloedema usually present and indicates increased intra-cranial pressure. Hemiparesis, abducens and facial nerve palsies, visual deficits, seizures and gait disorders occur in supratentorial lesions.

Tetrad of Schroeder described by Arana-Iniguez which was present in 86% of patients but not specific to cerebral hydatidosis. The tetrad included: (1) a patient who is a country dweller (2) in good general condition; (3) signs and symptoms of raised intra-cranial pressure and (4) without marked focal findings.

Haematological investigations show a raised eosinophil count. Immunologic tests useful are Casoni's intradermal skin test, immunoelectrophoresis, indirect immunofluorescence and Weinberg's complement fixation tests. Skull radiography may show erosion, localized bulging and signs of increased intra-cranial. The CT scan reveals an intraparenchymal hypo-dense lesion with a clearly defined margin. The cyst fluid has the same density as of CSF. Calcifications rarely seen. There is significant mass effect with midline and ventricular shift. Perilesional oedema usually absent but few cases been reported. Margins of the cyst may enhance if infected or in extradural cysts. Hydatid cysts in MRI are hyperintense on T2-weighted images and hypointense on T1-weighted images, and are slightly hyperintense when compared with CSF. No perilesional oedema present in uncomplicated cases.

MRI is useful than CT in showing the pericyst layer, which appears as a halo, and showing perilesional oedema. MRS shows peaks in lactate, acetate and succinate. Imaging of the chest, abdomen and heart are necessary to rule out systemic infestations.

Surgery is still the mainstay for treating intra-cranial hydatid cysts and the aim is to excise the cysts entirely without rupture where possible. This is as cysts may drain into the ventricles or rupture into the subarachnoid space which may be catastrophic leading to Anaphylactic shock, Meningitis and local recurrence.

Medical management as the primary option for single and solitary small cysts or for inoperable cysts has been attempted in India. Albendazole in concert with Praziquantel were used in the circumstances precluded to earlier as the primary mode of treatment. Albendazole results in the disappearance of up to 48% of cysts and a substantial reduction in size of the cysts in another 28%(1). The duration of the treatment is four weeks or more, and recently many authors have favoured a prolonged therapy. The change in levels of cyst markers such as alanine, succinate, acetate and lactate, measured before and during treatment on Proton Magnetic Resonance Spectroscopy (MRS), correlate well with shrinkage and resolution of cyst findings on conventional MRI and help in evaluating the efficacy of chemotherapy.

The current accepted standard approach is surgical resection combined with Pre and Post-operative Albendazole and praziquantel. The Dowling-Orlando technique as described in standard texts remains the most recommended method whereby the cyst is delivered by lowering the head of the operating table and instilling warm hypertonic saline between the cyst and the brain. This delivers the cyst whole, ensuring minimal spillage.

COMPLICATIONS

As the patient had a large thin walled cyst, with peri-ventricular location, complications are expected notably cyst rupture, and complications of the cavity which include subdural haematoma's in Giant cysts as well as cardio-respiratory failure, cortical collapse and brain oedema. However cardio-pulmonary failure is reported in multiple cysts.

CONCLUSION

In conclusion, hydatid cyst disease in childhood remains a serious health problem especially in rural areas of north eastern Kenya and Turkana. In children, cysts can grow to enormous sizes as in the case reported. However unlike ours, others have been reported to remain neurologically intact. For this reason, severe headaches in childhood should be carefully investigated in countries where hydatid

disease is pandemic, in order to prevent or minimize morbidity associated with late diagnosis.

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