CONGENITAL HEPATIC CYST WITH SEVERE MALNUTRITION IN AN INFANT: A CASE REPORT

Anthony O. Atimati and Samuel S. Anikoh

Department of Child Health, University of Benin Teaching Hospital, Benin City, Edo State

Correspondence Address: Dr Anthony O. Atimati, Department of Child Health, University of Benin Teaching Hospital, Benin City, Edo State. *E-mail:* tonyatimati@yaho.com, *Phone number:* +2348023417855

ABSTRACT

Congenital hepatic cysts are usually asymptomatic, very rare conditions in infancy and are most often discovered incidentally during radiologic imaging. They are more common in females with no racial differences. Most simple hepatic cysts are small and disappear spontaneously but in about 15% of cases, the cysts may be large and compress on adjacent structures leading to different clinical manifestations.

We present a 10month old girl who presented with abdominal swelling, fever, pallor and difficulty with breathing. An abdominal CT scan confirmed congenital hepatic cyst which was managed by percutaneous needle aspiration and sclerotherapy alongside nutritional intervention for severe malnutrition.

Keywords: Congenital hepatic cyst, Severe malnutrition, Needle aspiration, Sclerotherapy

INTRODUCTION

Congenital hepatic cyst is a usually asymptomatic and very rare condition in infants and children. Most cases are small in size and are usually discovered incidentally during radiologic imaging. Congenital hepatic cysts result from obstruction of the peri-biliary glands which normally arise from the ductal plate at the hepatic hilum around the 7th week of gestation. Persistence and dilatation of small remnants of the ductal plate malformation that separate out of the biliary tree gives rise to these cysts.[1]

The prenatal incidence is lower than the postnatal incidence which ranges from 2.5 – 4.25%.[2,3] It is more common in females with no racial differences. The cyst resolves without intervention in most cases but a few may enlarge and present with complications such as haemorrhage, abdominal pain or discomfort, intraperitoneal rupture, respiratory distress, torsion of a mobile cyst, early satiety, jaundice, infection necessitating surgical intervention.[3,4] In view of the rarity of this condition a case of a large congenital hepatic cyst complicated by severe malnutrition is hereby presented.

CASE SUMMARY

A 10month old girl presented with abdominal swelling of 7months duration, reduced urinary volume of 2months, paleness of the palms and feet of 2 weeks, fever of a week and difficulty with breathing of 2 days. She was delivered in a maternity home to a 33 year old lady at term. An abdominal scan done at 35 weeks gestation revealed a unilateral anechoic fluid filled kidney in the fetus. Abdominal swelling was noticed at 3 months of age and she presented in a hospital where an abdominal scan was done. Abdominal scan revealed a huge cystic retroperitoneal lesion and a diagnosis of? Dermoid cyst to rule out a lymphangioma was made. The parents declined admission on account of financial constraint and

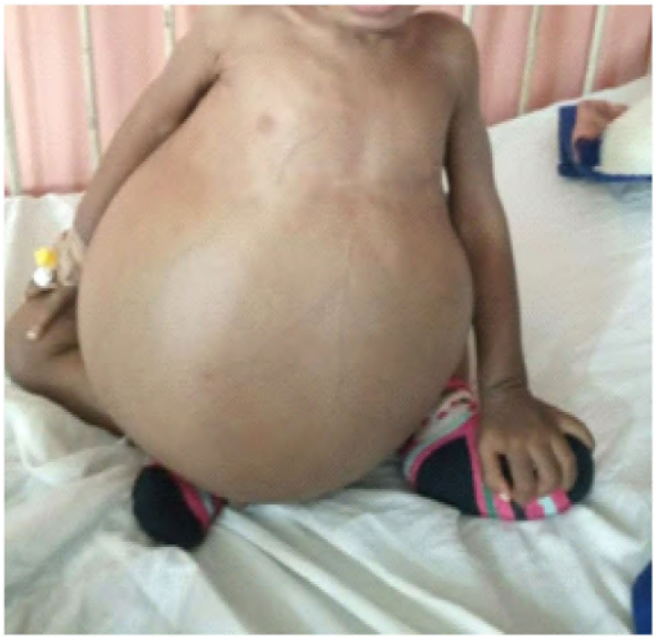

Fig. 1: Abdominal distension from huge hepatic cyst

presented 7months later with above symptoms. She was exclusively breastfed and commenced on corn gruel mixed with infant formula which she takes scantily due to early satiety. She was fully immunized for age and had normal developmental milestones. She was chronically ill-looking, markedly pale with bony prominences and loose hanging skin. Her weight was 5.5kg, length 65cm, weight for height z-score was<-3 SD, mid-upper arm (MUAC) was 10cm indicating severe acute malnutrition. She was markedly dyspnoeic and tachypnoeic with a fast bounding pulse and reduced oxygen saturation. The abdomen was markedly distended with a girth of 70cm (see figure 1). The full blood count showed a packed cell volume of 13%, normal white cell count and differentials and normal electrolyte and urea. The liver function test was slightly deranged; alkaline phosphatase 39 IU/L (9 – 35), Aspartate aminotransferase 116 IU/L (5 – 35), Alanine aminotransferase 55 IU/L (0 – 55), total bilirubin 0.5mg/dl (0.5 – 1.2), conjugated bilirubin 0.2mg/dl (0.0 – 0.5). Abdominal CT scan revealed an ill-defined cystic mass having thick internal septations in the left lobe of the liver (see figure 2). The kidneys, spleen and bowels were displaced while the pancreas was poorly visualized. The conclusion was that of a huge congenital hepatic cyst with features of secondary infection.

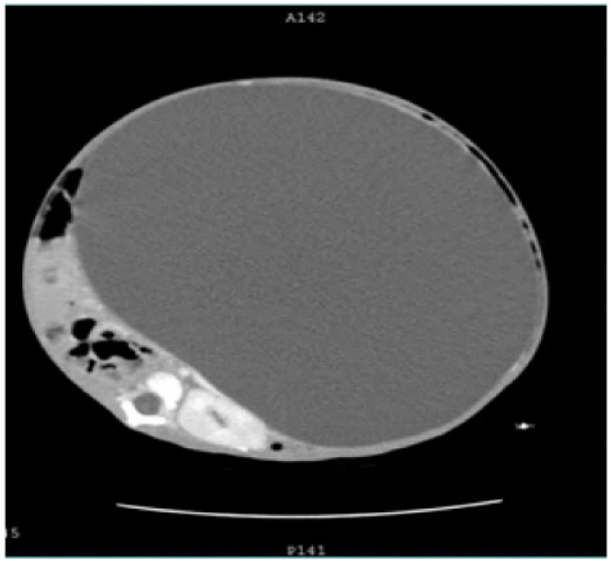

Fig. 2a: CT scan showing massive hepatic cyst

She was transfused with sedimented blood in 3 aliquots, commenced on antibiotics (cefuroxime) and nutritional rehabilitation. About 1 litre of straw-coloured fluid was drained from the intra-abdominal cyst under ultrasound guidance using a nephrostomy set. A pig-tail catheter was secured insitu and connected to a bag which drained 3650 mls over a 48-hour period. Alcohol was instilled into the sac to induce sclerosis. Patient was discharged on account of financial

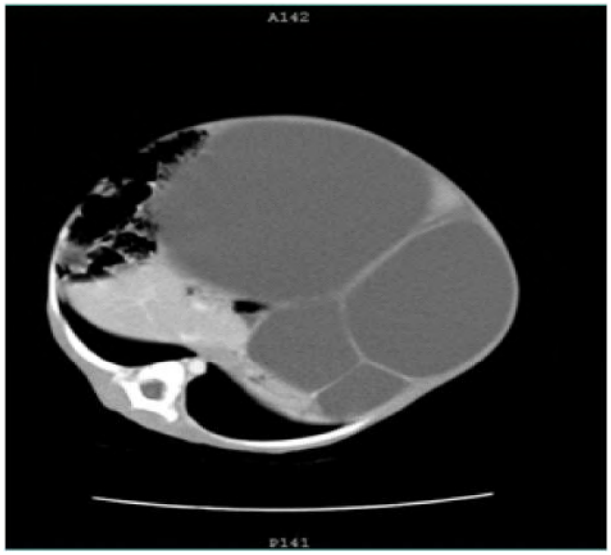

Fig. 2b: CT scan showing massive hepatic cyst with septations

constraint. The cytology of the aspirate was negative for malignancy and hydatid cyst. A repeat abdominal ultrasonography two weeks later showed re-accumulation of the cyst which was drained under ultrasound guidance with a drain in-situ. A review a week later revealed minimal drainage of about 30mls daily and an empty sac on ultrasound scan. Tetracycline capsules mixed with normal saline was infused into the cavity. Patient's weight had appreciated to 7.0kg (weight for length z score was 0) with no sign of re-accumulation of the cyst a week later but defaulted from subsequent clinic appointment.

DISCUSSION

Congenital cysts of the liver are mainly solitary non-parasitic cysts. They are usually referred to as simple cysts which differentiate them from other types of liver cysts – polycystic liver disease, parasitic or hydatid cyst, cystic tumours. They are rare with an incidence ranging from 2.5 – 4.25% in the literature.[3] Most hepatic cysts are small and usually disappear spontaneously. A 20 year review of prenatal ultrasound scan in 54,500 fetuses between 13th and 17thweek of gestation detected 7 hepatic cysts, six of which disappeared between the 18th – 24th week while one disappeared post-natally.[5] Females are more affected with a F:M ratio of 1.5:1in asymptomatic cysts and 9:1 in those with complicated or symptomatic cysts.[6] This patient had a complicated cyst which is in consonant with previous report of complicated cysts being prevalent in females.[6] Most simple hepatic cysts are asymptomatic and are usually picked incidentally during radiological imaging. A few may increase in size and cause symptoms such as pain and other manifestations resulting from compression on adjacent structures. About 15% of persons with simple cysts become symptomatic.[7] A cyst measuring greater than 4cm in its largest diameter is classified as a large cyst.[3] The onset of symptoms depends on the rapidity of growth of the cyst. Bhosale *et al* reported a neonate who presented at 3 days of life with marked respiratory distress following a huge hepatic cyst.[8] The first presentation of this patient was at 3 months and later at 10 months with failure to thrive and marked respiratory distress due to splinting of the diaphragm. The easy satiety predisposed to the severe malnutrition and severe anaemia observed in this patient.

Diagnosis can be made with an abdominal ultrasound which is cheap, non-invasive, available and less technical. The specificity is between 90 – 95%[6] but the diagnosis with ultrasound was missed twice – prenatally and at 3months – in this patient. Computerized tomography (CT) scan is more accurate and detailed in diagnosing hepatic cyst which was the diagnostic tool in this patient. Magnetic resonance imaging is also very accurate, though more expensive, requiring expertize and is not readily available. The liver function test was slightly deranged which was more marked in the aspartate aminotransferase which is similar to the finding of Bryce *et al*[9]. The serum bilirubin is normal except in large cysts causing obstruction to biliary flow which usually manifest with clinical jaundice.

Modalities for treatment of simple hepatic cysts include percutaneous aspiration and sclerotherapy using alcohol, minocycline/tetracycline hydrochloride; de-roofing of the cyst either by open or laparascopic technique; and liver resection. Re-accumulation of cyst is a disadvantage of the aspiration and sclerotherapy treatment modality. Simple liver cysts are lined by cuboidal or columnar epithelium which is similar to that of the bile duct epithelium.[7] The epithelium continues to secrete plasma-like fluid which often re-accumulates after drainage. In a report of 10 cases of symptomatic hepatic cysts managed by laparascopic and open deroofing, two of which had previously been treated by aspiration and sclerotherapy, eight were

cured while two had asymptomatic recurrence. Sewing of the omentum to the margin of the cyst (omentoplasty) after surgical deroofing is advocated to reduce recurrence. Our patient had aspiration and sclerotherapy, first with alcohol and later with tetracycline to achieve a resolution before being lost to follow up.

Prenatal abdominal ultrasound scan is advocated to pick early any hepatic cyst which should be repeated after delivery to ascertain the status of the cyst. Recinos *et al* reported disappearance of a congenital hepatic cyst, diagnosed in utero, after 7 months of follow-up postnatally.[2]

CONCLUSION

Simple hepatic cysts are rare and usually asymptomatic congenital cysts but occasionally manifests with symptoms in large varieties often necessitating intervention. Abdominal ultrasonography is a useful diagnostic tool but in uncertain situations a CT scan should be requested to facilitate early diagnosis and treatment in order to avoid complications such as severe malnutrition as seen in this patient.

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