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CASE REPORT

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Cystic hepatic hamartoma causing pseudo-prune belly in a female infant in Tamale, Ghana

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Introduction

Prune belly syndrome (PBS) is a congenital abnormality that was initially described in 1901 by Osler and the term PBS was coined by Eagle and Barret in 1950 [1,2]. It consists of a triad of congenital paucity of the abdominal wall musculature, bilateral cryptorchidism in males, and genitourinary malformations [3]. This rare anomaly has an incidence of approximately one in 40,000-50,000 live births with 95-97% of affected newborns being male [3]. The pseudo-prune belly includes all females because of their lack of cryptorchidism, as well as males with unilateral or bilateral testicular descent, and boys with no urinary tract dilation [3]. In this paper, we present a brief case report of a prune belly variant encountered in a female neonate in Ghana, West Africa. The prune-like wrinkled abdomen made for an initial impression of PBS. The appearance was even more pronounced after surgery. This presentation was due to a large hepatic mass causing distension of the abdomen and thinning out of the abdominal wall muscles and she had no genitourinary abnormalities.

Case report

The patient was a 7-day old female delivered via spontaneous vaginal delivery in a district health facility to a 30-year old Gravida 1, Para 1 female. The birth weight was 2.7 Kg. The mother's past medical history along with the pregnancy and delivery history were unremarkable. There was no evidence of antenatal ultrasound scan having been performed. The patient was referred on day 7 of life to the Tamale Teaching Hospital, which is the only tertiary referral center in Northern Ghana, for further evaluation and management of abdominal distension and obvious laxity of the abdominal wall musculature.

Initial physical assessment at our center revealed a term female neonate who was not in any respiratory distress and had been feeding well and voiding and stooling regularly since birth. The abdominal wall was lax and distended (Figure 1) with a palpable mass at the right upper quadrant. No other organs, including both kidneys, were palpable and all the other systems were normal. The serum electrolytes, kidney function tests, and a Complete Blood Count (CBC) performed on admission were within normal ranges.

The child had been tolerating full breast feeds with appropriate stools and urination. An abdominal ultrasound was obtained at 19 days of life which noted hypoplastic abdominal wall musculature along with a large 205 cm3 cystic lesion in the right upper quadrant that was displacing the liver and bowel loops to the left. The liver otherwise showed a normal echographic pattern. The kidneys, spleen, ureters, and urinary bladder were also sonographically normal.

At 23-days of age, she underwent exploratory laparotomy via a right subcostal incision and was found to have a large, simple, cystic lesion of hepatic segments five and six. There was a segment of attenuated liver and a diminutive gallbladder over the cyst and a long, thin cystic duct (Figure 2a and 2b). There was no intraoperative fluoroscopy available. To determine patency and continuity of the gallbladder with the biliary tree, the gallbladder was needle-aspirated and found to have green bile. Dilute betadine (1 mL betadine with 4 mL normal saline) was instilled into the gallbladder with appropriate distension and color change of the gallbladder, however, the cystic duct, common bile duct, and duodenum did not change color, suggesting an atretic cystic duct which did not communicate with the common bile duct. Initially a small opening in the hepatic cyst was made in order to decompress the cyst, suctioning the fluid. Then, using electrocautery, the cyst was excised at its junction with the normal appearing liver maintaining meticulous hemostasis. A small portion of the cystic wall was left attached to the liver. The atretic cystic duct was ligated with an absorbable suture ligature near its junction to the common bile duct. The bowel was examined and there was normal rotation present with a mobile right colon. There was also an omphalomesenteric duct remnant (Meckel's) band to the umbilicus, which was transected with electrocautery. The uterus and adnexal structures were grossly normal. The fascia was closed with simple interrupted absorbable sutures. The skin was closed with a running, subcuticular absorbable suture. A dry gauze and tape were applied. The procedure was 80 minutes.

Postoperatively, the patient did not have a nasogastric tube in place; she was kept NPO overnight and had no emesis. About 18 hours after surgery, on postoperative day 1, she was started with small volume cup feeds but then developed significant abdominal distension. An urgent abdominal ultrasound was obtained that same day which showed a small volume of free fluid in the infrahepatic location with extensive bowel gas shadowing and minimal bowel peristalsis consistent with paralytic ileus. The remaining hepatobiliary system, spleen and pancreas were normal on ultrasound. Her bowel function gradually returned over the next few days, her feeds were advanced without event and she was discharged to home on postoperative day 7. She was doing well during the first 2 visits after discharge but has since not returned for subsequent follow-ups.

The pathology report showed an opened cystic mass measuring 10 x 6.5 x 1 cm with variable thickening of the wall 0.2-0.4 cm. Sections of representative portions of the cystic mass showed proliferating variable sized blood vessels within the fibrous stroma, nodules of benign hepatocytes separated by thin fibrous septae, dilated hepatic sinuses, and proliferating bile ducts with fibrous septae consistent with a hamartoma. No malignancy was seen (Figure 3).

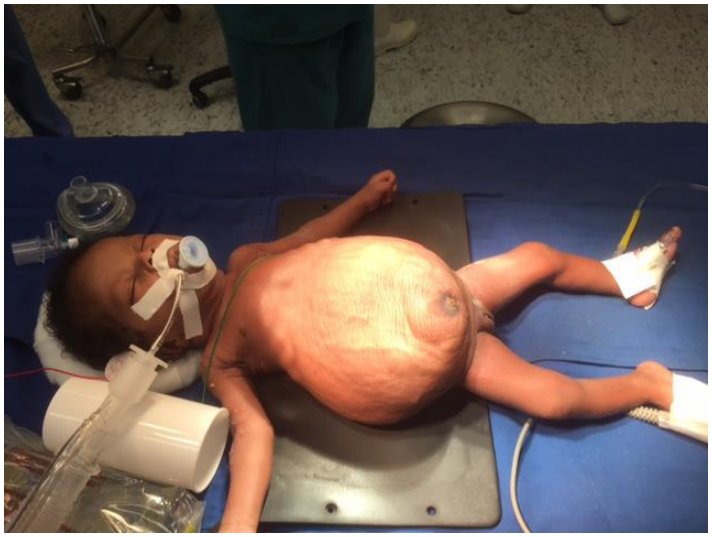

Figure 1. *Preoperative appearance of the abdomen of the patient.*

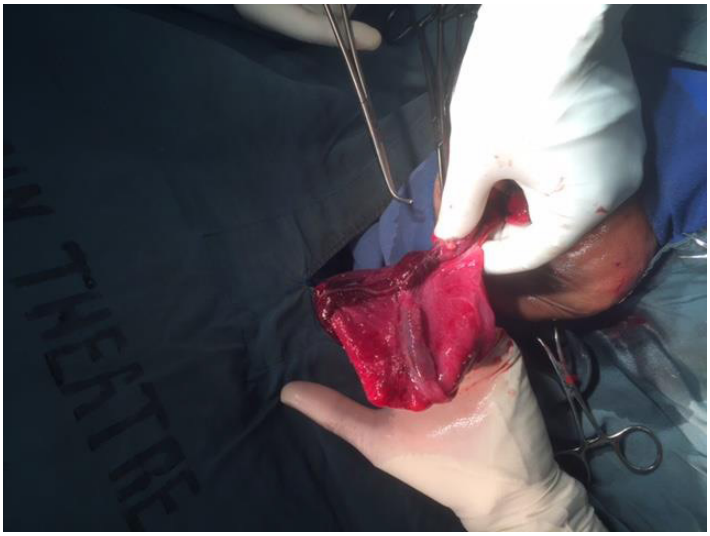

Figure 2b. *Intraoperative appearance of cystic hamartoma, gallbladder is filled with dilute betadine solution.*

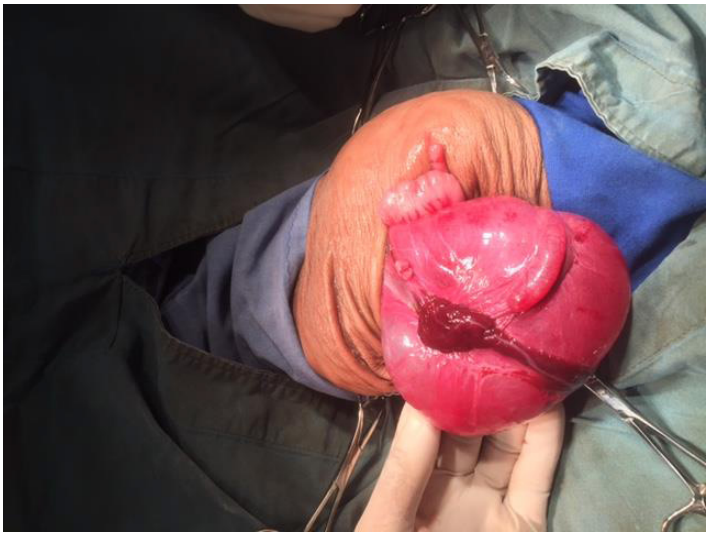

Figure 2a. *Intraoperative appearance of cystic hamartoma.*

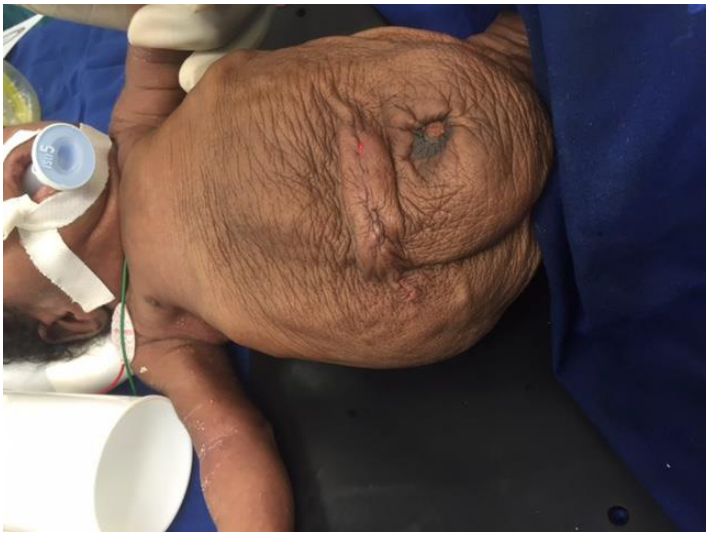

Figure 3. *Post-operative appearance of the abdominal wall of the patient.*

Discussion and conclusion

Prune belly syndrome has generally been a congenital disease that primarily affects males however variations of presentation affecting female neonates have been reported [4,5].

The pathogenesis of PBS remains unknown however there are two leading theories. The first theory describes an abdominal wall mesodermal defect during early embryogenesis that leads to abdominal wall muscular hypoplasia with subsequent severe urinary tract dilatation. The leading theory suggests that over-distension of the abdominal wall occurs from the bladder due to a urinary tract outlet obstruction which leads to defects or absence of the abdominal wall musculature. PBS is also associated with a spectrum of other abnormalities ranging from cardiac, respiratory, musculoskeletal, and gastrointestinal.

Our patient had hypoplasia of the abdominal wall musculature secondary to the large hepatic cystic hamartoma without any genital or urinary abnormalities. Her genito-urinary exam appeared normal on postnatal clinical, ultrasonographic, and surgical examination. Due to limited resources in the region, a voiding cystourethrogram (VCUG) for further evaluation of the urinary tract was unavailable.

There are several published reports of neonates with cystic abdominal liver masses diagnosed in utero [4-7]. These can be treated with fetal hepatic cyst aspiration and weekly sonographic monitoring; nevertheless, some fetuses progressed to fetal hydrops, postnatal respiratory failure, and death despite interventions. Our case demonstrates a variant of PBS in a female neonate secondary to a cystic hepatic hamartoma diagnosed shortly after birth, in a resource poor country. With unavailable intraoperative fluoroscopy, a visiting pediatric surgeon from Kumasi, Ghana, must be commended for the technique of utilizing dilute iodine to delineate the lack of patency of the cystic duct, prior to completion of the cystectomy. We are grateful for his experience and insight.

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