Journal of Pediatric Surgery 51 (2016) 1390–1393

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Pediatric Surgical Image

Further evidence of the etiology of prune belly syndrome provided by a transient massive intraabdominal cyst in a female

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ARTICLE INFO

Article history:
Received 12 April 2016
Received in revised form 14 May 2016
Accepted 21 May 2016

Key words:
Prune belly syndrome (PBS)
Etiology
Pathogenesis
Fetal cyst

ABSTRACT

We present a female neonate born with prune belly syndrome (PBS) in whom a large intraabdominal cyst was diagnosed at 12 weeks of gestation. Rapid and exponential growth of the cyst caused pressure effects on the intraabdominal organs and stretching of the anterior abdominal wall by 19 weeks of gestation. This led to drainage of the massive cyst at 20 weeks of gestation to prevent fetal demise. This case provides further clues to the likely etiology of PBS: transient stretching and attenuation of the fetal abdominal wall secondary to gross fetal abdominal distension – from any cause.

The prune belly syndrome (PBS) affects up to 1/50,000 newborn babies [1]. It is defined by a triad of abdominal muscle hypoplasia, bilateral cryptorchidism in males and urinary tract abnormalities. Its etiology has been subject to vigorous debate over the years. Females with PBS are sometimes described as having pseudoprune belly syndrome, prune belly variant or prune belly-like appearance (because of the absence of cryptorchidism) but it is likely that they all are a manifestation of the same process as seen in classical PBS.

1. Case presentation

A 12 week gestation routine ultrasound examination of a 38 year old type 1 diabetic and hypertensive primiparous woman revealed a 19 mm × 13 mm × 13 mm cystic structure within the fetal abdomen. The cyst, of uncertain origin but separate from the bladder (Fig. 1), became massive over the next few weeks (Table 1). By the 19 week scan it had caused the fetal abdominal wall to be thinned (Fig. 2). It was elected to drain the massive cyst at 21 weeks gestation because it was compressing internal structures and stretching the anterior abdominal wall: approximately 140 ml of straw colored fluid was aspirated. The fluid contained bile acids (5 mmol/L) and cholesterol (2.8 mmol/L) suggesting a possible origin from the liver. Further scanning post aspiration at 23 weeks gestation showed the multi-septated residual cyst (measuring 32 mm × 32 mm × 29 mm) to be within the liver parenchyma (Fig. 3). At 26 weeks the cyst had resolved, and all that remained was an echogenic focus in the liver (Fig. 4). Delivery was expedited at 31 weeks because of pre-eclamptic toxemia and a prune belly-like appearance of the abdomen was noted (Fig. 5). Post-natal ultrasound scan of the abdomen showed a cyst with an irregular border, within the inferior aspect of the liver measuring 4 mm × 13 mm × 20 mm (Fig. 6). The ultrasound further showed a duplex right kidney with no dilatation and normal parenchyma; the left kidney and other intraabdominal organs were normal. However a malrotated midgut was suspected from the plain film leading to an upper GI contrast study which confirmed this finding. During the Ladd's procedure, attenuation of the rectus abdominal muscles was noted. Histology of the abdominal wall confirmed the hypoplastic muscle fibers and an increase in the fibrous connective tissue (Fig. 7). This was consistent with prune belly sequence. Microarray and karyotype showed no significant chromosomal imbalance, with a female result.

2. Discussion

Prune belly syndrome is a term first used by William Osler in 1901 to describe the appearance of the wrinkled thinned abdominal wall because of apparent deficiency of the abdominal wall musculature [2]. PBS affects males almost exclusively with only 3–5% of cases reported in females [2]. The other components of this syndrome, in males include varying degrees of urinary tract dilatation and cryptorchidism.

Only male patients fully satisfy the classical triad of anomalies. The cryptorchidism is usually because of inability of the intraabdominal testes to gain access to the internal inguinal ring because of an enlarged bladder [1]. Theories to explain the etiology of PBS include failure of primary mesodermal differentiation, transient retention of the yolk sac within the abdominal cavity and obstruction or dysfunction of the

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http://dx.doi.org/10.1016/j.jpedsurg.2016.05.017
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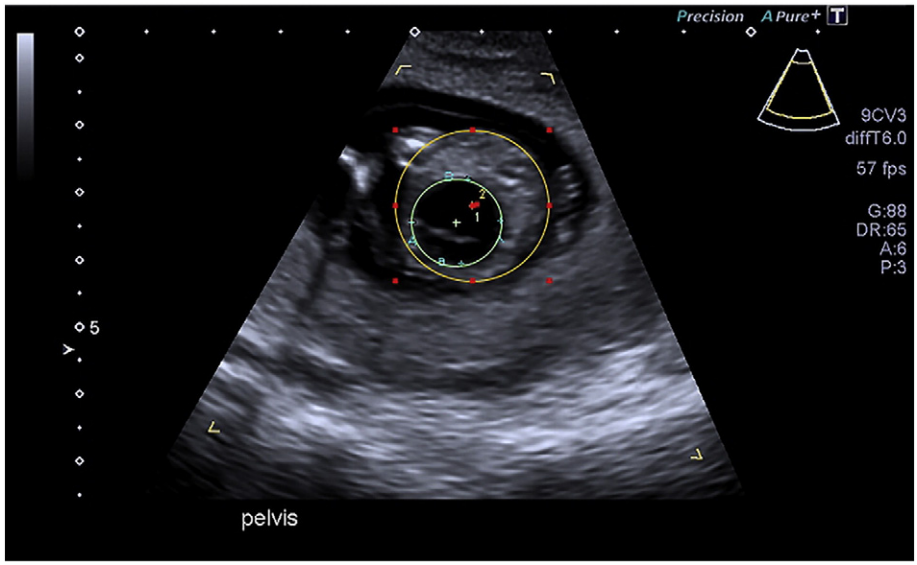

Fig. 1. The cyst (smaller circle) in relation to the fetus (larger circle).

Table 1
Growth of the cyst with gestation on serial ultrasonography.

Gestation	Cyst dimensions on ultrasound scan
12 weeks	19 mm × 13 mm × 13 mm
13 weeks	29 mm × 25 mm
16 weeks	48 mm × 63 mm × 67 mm
19 weeks	68 mm × 61 mm × 72 mm

urinary tract causing dilatation and distension of the fetal abdominal wall [3].

The etiology of our case is consistent with transient but gross fetal abdominal distension early in gestation markedly stretching and attenuating the anterior abdominal wall.

The actual cause for the gross abdominal distension varies: Beasley et al. concluded that the abdominal wall appearance can be a secondary manifestation of gross distension of the bladder and ureters from a transient obstruction at the junction of the penile and glanular urethra [3]. Yamamoto et al. reported PBS in a boy with a giant liver cyst diagnosed at 27 weeks gestation and a normal urinary tract. In their case the cyst was not drained *in utero* unlike our case where the cyst was drained at 20 weeks gestation. They concluded that the prune belly appearance resulted from the splitting of the abdominal wall as a consequence of early severe fetal abdominal distension [4]. In our case the intraabdominal cyst appears to have originated from the liver based on the fluid analysis and image findings post-aspiration, even though the large size of the cyst initially obscured its origin. The images have shown a multi-septated cyst consistent with a mesenchymal hamartoma, or any other hepatic developmental cyst.

Hirose et al. reported a girl with PBS with hydrops fetalis and transient fetal ascites. In addition, there are three cases of females with prune belly syndrome because of hydrops fetalis, even though they have sometimes been called pseudoprune belly syndrome because of the absence of cryptorchidism [5]. A literature review by Hassett et al. suggested that midgut malrotation (as occurred in our case) was the most common gastrointestinal condition associated with prune belly syndrome. Gastroschisis, omphalocele, and cloacal anomalies were

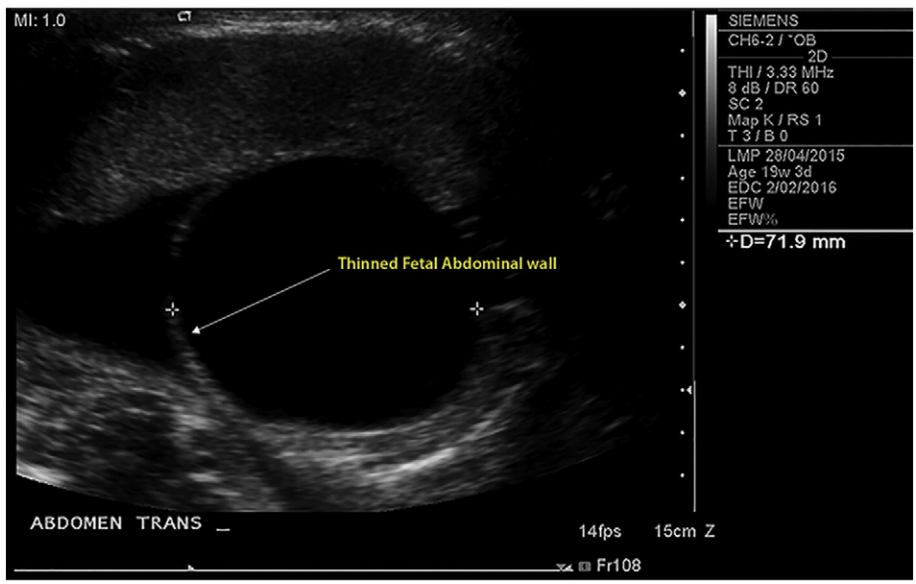

Fig. 2. Subsequent scan at 19 weeks showing the rapid growth of the cyst to 68 mm × 61 mm × 72 mm. The attenuated abdominal wall is clearly seen on the left.