Case Report

Prune belly syndrome with pouch colon and absent dermatome

Aejaz A. Baba, Syed A. Hussain, Altaf H. Shera, Rekha Patnaik

ABSTRACT

Prune belly syndrome (PBS) is a rare congenital constellation of defects in pediatric surgical practice. Although anorectal anomalies have been reported in association with PBS, only few case of pouch colon with PBS has been reported.[1] In addition, our patient had deficient abdominal wall with absent dermatome in left upper quadrant, which has never been reported in the English literature. This association with abdominal wall deficiency and absent dermatome not only strengthens the theory of mesodermal arrest in the etiology of PBS but also points towards a defect in the ectodermal development.

Key words: Prune belly, pouch colon, anorectal malformation

DOI: 10.4103/0189-6725.59356

INTRODUCTION

Prune belly syndrome (PBS) represents a spectrum of anomalies with variable degrees of severity predominantly affecting males with male:female ratio being 2.0:1 [2] The incidence of PBS has been reported to be 1 in 29,000 to 1 in 40,000 live births.[3] The three major findings are a *deficiency of the abdominal musculature*, *bilateral intraabdominal testes*, and an *anomalous urinary tract*.[4] The urinary tract may have variable degrees of hydronephrosis, renal dysplasia, dilated tortuous ureters, an enlarged bladder, and a dilated prostatic urethra. Additional associated anomalies exist involving the respiratory tract, gastrointestinal tract, cardiac system, and musculoskeletal system.[5]

PCS is a combination of absence of varied lengths of colon with replacement of the distal portion of the colon with a dilated pouch. Presence of a urinary fistula and abnormal vasculature are part of the PCS.[6] We hereby report a case with PBS with pouch colon and abdominal wall deficiency with absent dermatome which has never been reported in the literature.

Department of Pediatric Surgery, SKIMS Srinagar, Kashmir, India

Address for correspondence:
Aejaz A. Baba,
Senior Resident, Department of Pediatric Surgery,
SKIMS Srinagar, Kashmir, India.
E-mail: dr_aejaz@yahoo.co.in

CASE REPORT

One day old neonate was referred to our department after been resuscitated for neonatal respiratory distress with an APGAR score of 5 at birth and 7 at 5 minutes after birth. On examination, the baby was found to have lax anterior abdominal wall, protruding most prominently in the midline. There was a 5 x 4 cm^2 area in the left hypochondrium with deficient muscle, subcutaneous tissue and skin. Perineal examination showed bilateral cryptorchism, absent anal opening although natal cleft was well developed. There was also meconium staining at the urethral meatus suggesting presence of gastrointestinal-urinary fistula. Urethra could not be catheterized because of microurethera.

Investigations showed hyponatremia with Na of 123.1, urea of 81, and creatinine of 1.68. Ultrasound imaging showed mild dilatation pelvicalyceal system of right kidney grossly dilated both ureters, left kidney at ectopic position, distended urinary bladder containing debris, and distended bowel loops. Cross-table transtrochanteric x-ray was suggestive of high anorectal malformation.

Patient was planned for a colostomy and taken to the operating room with following intraoperative findings; i) type IV pouch colon [Figure 1] with colo-vesical fistula; ii) ectopic left kidney with hilum facing left; iii) tortuous and dilated ureters.

The pouch was excised and fistula closed, and colon brought out as end colostomy. In addition, suprapubic cystostomy was done for decompression as well as for monitoring urine out put [Figure 2]. Postoperative micturating cystogram revealed bilateral vesico ureteric reflux [Figure 3]. At present patient is on follow up for over two months with serum creatinine of 1.0.



Figure 1: Operative photograph showing pouch colon



Figure 2: Photograph showing lax abdominal wall, abdominal wall deficiency with end colostomy

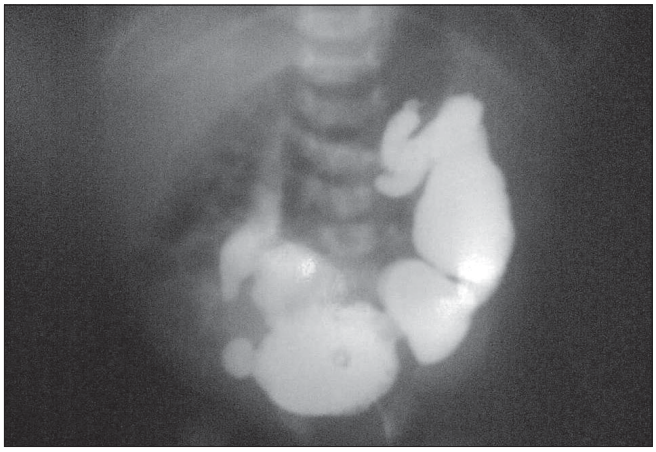


Figure 3: Contrast study showing bilateral severe vesico ureteric reflux

DISCUSSION

PBS is also known as triad syndrome, Eagle Barrett syndrome or abdominal musculation syndrome.[7] Parker *et al.* first recognized the three components of this syndrome.[4] The exact embryogenesis of PBS is not known. However, there are three theories that have been put forward to explain various anomalies associated with this syndrome. These are:[8]

a. Early in utero urethral obstruction leading to urinary tract dilatation and fetal ascitis
b. Mesodermal arrest theory
c. Defect in yolk sac

The mesodermal arrest theory explains most of the anomalies associated with PBS. This is further supported by the histological findings of abdominal wall, urinary, and genital tracts. The association of examphalos and gastroschisis with PBS also strengthens this theory.[9,10] The presence of a abdominal wall deficiency with absent dermatome points towards a defect in both mesodermal and ectoderm development.

Gastro-intestinal anomalies are seen in over 30% of the patients.[11] Although anorectal agenesis is quite common with PBS, congenital pouch colon has been reported in only few cases.

The anterior urethra in cases of PBS is usually normal; however, atresia or megalourethra has been reported. Our patient had microutrethra which could not be catheterized. Other systems with anomalies include cardiac, pulmonary, musculo skeletal, etc.

They may need a supravesical diversion, cutaneous vesicostomy, anterior urethral reconstruction, reduction cystoplasty, ureteral reconstruction, and reconstruction of anterior abdominal wall. Despite optimal treatment 30% of the patients develop renal failure and require renal transplantation. These patients need multidisciplinary management of a neonatologist, nephrologists, and pediatric urologist for an optimal outcome.

REFERENCES

1 Bangroo AK, Tiwari S, Khetri R, Sahni M. Congenital pouch colon with prune belly syndrome and megalourethra. Pediatr Surg Int 2005;21:474-7.
2 Rabinowitz R, Schillinger JF. Prune Prune belly syndrome in the female subject. J Urol 1977;115:454-6.
3 Greskovich FJ 3rd, Nyberg LM Jr. The prune belly syndrome: a review of its etiology, defects, treatment and prognosis. J Urol 1988;140:707-12.
4 Parker R W. Case of an infant in whom some of the abdominal muscles were absent. Trans Clin Soc Lond 1895;28:201-3.
5 Salihu HM, Tchuinguem G, Aliyu MH, Kouam L. Prune belly syndrome and associated malformations. A 13-year experience from a developing country. West Indian Med 2003;52281-4.
6 Rao KL, Menon P. Congenital pouch colon associated with anorectal agenesis (pouch colon syndrome). Pediatr Surg Int 2005;21:125-6.

7 NUNN IN, STEPHENS FD. The triad syndrome: a composite anomaly of the abdominal wall, urinary system and testes. J Urol 1961;86:782-94.
8 Stephens FD, Gupta D. Pathogenesis of the prune belly syndrome. J Urol 1994;152:2328-31.
9 Short KL, Groff DB, Cook L. The concomitant presence of gastroschisis and prune belly syndrome in a twin. J Pediatr Surg 1985;20:186-7.
10 Walker J, Prokurat AI, Irving IM. Prune belly syndrome associated with exomphalos and anorectal agenesis. J Pediatr 1987;22:215-17.
11 Wright JR Jr, Barth RF, Neff JC, Poe ET, Sucheston ME, Stempel LE. Gastrointestinal malformations associated with prune belly syndrome: three cases and a review of the literature. Pediatr Pathol 1986;5:421-8.

Source of Support: Nil, **Conflict of Interest:** None.