Case Report

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An Extremely Rare Association of Prune belly Syndrome with Congenital Pouch colon : Case Report and Review of Literature

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Abstract

Prune belly syndrome (PBS) is a triad of an abdominal wall muscular deficiency, cryptorchidism, and dilated urinary tract, and is often associated with other anomalies. Association of PBS with congenital pouch colon is extremely rare with only few handful of such reports in literature. We report here management of this rare associationin a male newborn with multiple genitourinary abnormalities.

Keywords: Congenital pouch colon; Prune belly syndrome; High Anorectal malformation; Megaureter.

INTRODUCTION

Prune belly syndrome (PBS) is a triad of an abdominal wall deficiency in muscular tissue, cryptorchidism, and dilated urinary tract with reported incidence is 1 in 29,000 to 1 in 40,000 live birth.[1] Congenital pouch colon (CPC) is a rare and an unusual high anorectal malformation associated with pouch-like dilatation of a varying degree of shortened colon. CPC is itself an uncommon entity and association with prune belly syndrome is extremely rare with only five such case reports in literature. CPC has a regional preponderancefor India with a review showing that 92.2% of reported cases were from India.[2] We report here management of a male neonate with prune belly syndrome associated with congenital pouch colon and genitourinary abnormalities including severe hydronephrosis, massively dilated tortuous ureters, and congenital megalourethra.

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CASE REPORT

A full-term male neonate (birth weight 2.4kg), with ahistory of antenatally diagnosed bilateral hydroureteronephrosis born by spontaneous vaginal delivery presented with absence of anal opening and progressive abdominal distension. Baby was sick and dehydrated at presentation. Capillary refilling time was >3 seconds. Abdomen was flabby and distended and with visible loops of

intestines. Overlying skin was loose and wrinkled. [Fig.1]

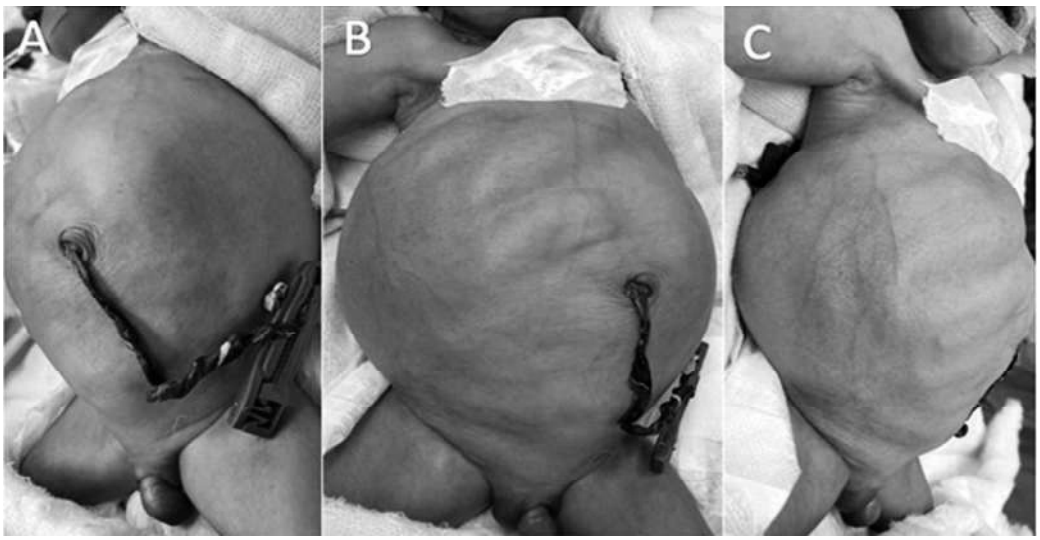

Fig. 1: Showing flabby and distended abdomen with visible loops of intestines

Urine output was low and urine was trickling in drops. Catheterisation was attempted but couldn't be catheterised. On pressing the bladder urine slowly getting filled up in dilated urethra. Anterior urethra was scaphoid megalourethra with absent corporal tissue. Scrotal sac was very small and rugosities were absent. Both testis were absent and impalpable nowhere externally. Anal opening was absent with poorly developed gluteal musculature. Baby was resuscitated with intravenous fluids and nasogastric decompression. Routine investigations were sent. Cross table prone lateral and erect anteroposterior radiograph were done. Radiograph shows high anorectal anomaly and features of pouch colon. [Fig.2] Creatinine

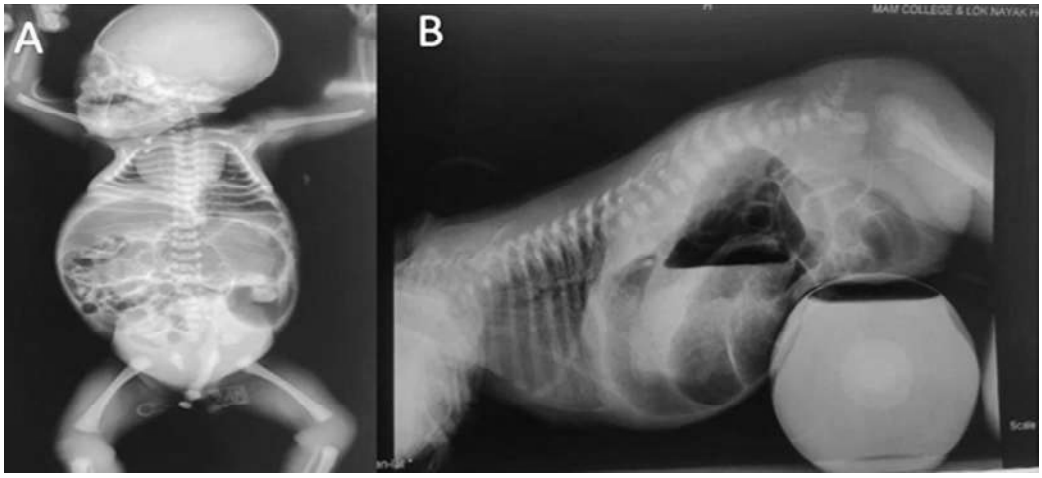

Fig. 2: Showing(A) plain radiograph, Anteroposterior view,(B)Cross table prone lateral view.

was raised (1.8mg/dl) and blood gas analysis showed metabolic acidosis. Antenatal Ultrasound during third trimester showed bilateral hydrourete ronephrosis and oligohydramnios. Post-natal Ultrasound of abdomen showed severe bilateral hydroureteronephrosiswith pelvic diameter on right side was 2.8 cms and on left side was 3 cms along with massively dilated ureters on both sides. On Exploratory laparotomy deficient and loose abdominal wall musculature was evident. Catherization was tried under general anaesthesia but urethra was not catherizable. Type IV pouch colon lacking haustrations, appendices epiploicae, taenia coli with an abrupt transition from the

normal proximal bowel to the distal dilated pouch was identified. [Fig.3] The bladder was also dilated and contained turbid urine. The colovesical fistula was ligated, the pouch colon was excised,

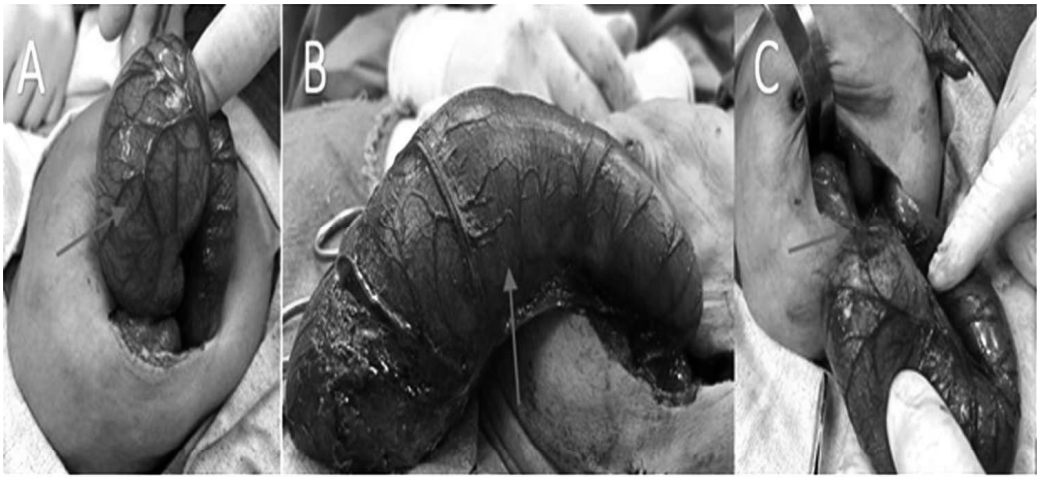

Fig. 3: Showing (A, B & C)Type IV pouch colon lacking haustrations, appendices epiploicae, taenia coli with an abrupt transition from the normal proximal bowel to the distal dilated pouch was identified

and an end descending colostomy was done. In view of massively dilated and tortuous ureters, ureterostomy was done on left side andsuprapubic cystostomy was done. [Fig.4]

In postoperative period baby showed recovery and

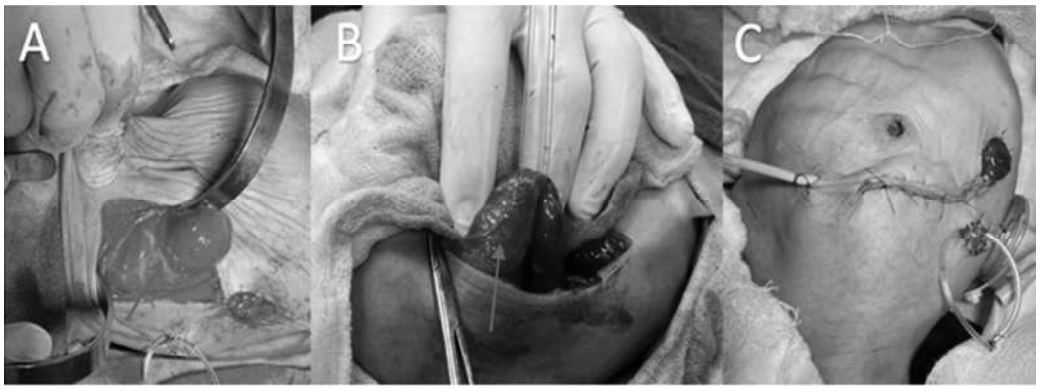

Fig. 4: Showing (A)Enlarged and distended bladder with diverticulum, (B) Massively dilated ureter, (C) End descending colostomy, ureterostomy and suprapubic cystostomy.

creatine came down to 0.9 mg/dl on serial kidney function test reports with adequate urine output. Stoma got functional on third post-operative day. Initially broad spectrum antibiotics were started. Later, post-operative blood culture grew Acinetobacter and urine showed budding yeast cells. Antibiotics were adjusted in accordance with sensitivity pattern along with anti-fungal drugs (Meropenem, Vancomycin and Amphotericin B). Baby developed full blown sepsis and succumbed on eighteenth post-operative day.

DISCUSSION

Prune belly syndrome (PBS) is also known as Eagle-Barrett syndrome and characterized by the classical triad of urinary tract anomalies, deficient abdominal musculature, and bilateral cryptorchidism. PBS has been associated with variety of congenital anomaliesincluding cardiopulmonary, gastroin testinal, musculoskeletal and respiratory abnormalities.[3,4] Gastrointestinal malformations such as mesenteric malrotation, atresia, stenosis, volvulus, anorectal malformation, splenic torsion, hirschsprung disease and gastroschisis are knownand seen in 30% of these patients.[4,5] Genitourinary anomalies areseen in 24% of patients thatinclude renal dysplasia hypoplasia, bilateral hydronephrosis, bilateral hydroureter, vesico ureteric reflux, megacystis, megalourethra, hypospadias, undescended testes and VACTERL anomalies are known.[4] Cardiopulmonary,

gastrointestinal andmusculoskeletal anomalies constitute 75% of associated anomalies.

Congenital pouch colon is arare gastrointestinal association with PBS and has been reported in only few instances before. CPC is also known as congenital short colon,a rare variant of ARM, and it is distinctly different from other ARMs in which a varying length of colon is replaced by a dilated pouch thatinvariably has a wide fistulous communication with the genitourinary tract. The classification proposed by Narasimharao et al is based on the length of normal colon proximal to the colonic pouch.[7] In type 1, the ileum opens directly into a pouch; in type 2, the ileum opens into a short segment of the cecum, which then opens into a pouch; and in type 3, at least 10 to 15 cm of normal colon is present between the ileum and the pouch, in type 4, only the terminal portion of the colon (sigmoid or rectum) is converted into a pouch. CPC cases are exclusively found in Southeast Asia, particularly in India.

PBS association with CPC is extremely rare and only five reports have been documented in literature. [Table I]

Table I: Review of literature showing PBS with associated anomalies.

Studies	PBS with Type of Pouch Colon	Associated Anomalies
Bangroo et al. (8)	PBS + Type I CPC	Anterior Urethral Diverticulum
Baba et al. (9)	PBS + Type IV CPC	Microurethra + absent dermatome
Raghavan et al (5)	PBS + Type I CPC	Penoscrotal Hypospadias
Garge et al. (10)	PBS + Type IV CPC	Scaphoid Megalourethra + Cardiac defects (Coarctation of aorta, patent ductus arteriosus, atrial septal defects)
Annigeri et al. (11)	PBS + Type IV CPC	Bilateral Hydronephrosis with massively dilated & tortuous ureters
This Study	PBS + Type IV CPC	Scaphoid Megalourethra + bilateral Hydronephrosis with massively dilated & tortuous ureters

Antenatally PBS is associated with prostatic hypoplasia, some patients have a urachal diverticulum or megalourethra; a large, thick-walled bladder; tortuous and dilated ureters. Varying amounts of hydronephrosis and varying degrees of renal dysplasia are seen. Perinatal mortality rates are high, ranging from 10 to 25%, with early death primarily attributed to the degree of prematurity, Oligohydramnios and pulmonary hypoplasia.[12] When PBS is associated with CPC it may further jeopardise the prognosis and aggravate the severity, as the pouch in the abdomen can cause physical obstruction that adds to functional obstruction in PBS.

In view of massively dilated and tortuous ureters child may need a supravesical diversion (ureterostomies), cutaneous vesicostomy and later reduction cystoplasty, ureteral reconstruction, and reconstruction of anterior abdominal wall.Anterior urethral reconstruction may also be required in the setting of megalourethra.

CONCLUSION

We can conclude that PBS is an uncommon entity and its association with CPC is extremely rare. This rare association also has poor prognosis. Multifaceted approach is required due to the association with multiple anomalies. Proper pre and postnatal assessment, manoeuvring of surgical procedures, counselling and postoperative care are important for the management of this rare association.

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