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Case Report

Surgical management of congenital pouch colon in Lebanon: A case report and review of the literature ☆

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ABSTRACT

Congenital pouch colon (CPC) is a rare congenital abnormality, in which a pouch-like dilatation partially or completely replaces the colon, creating a fistula with the urogenital. Congenital colonic pouch is an extremely rare congenital disease mainly reported in India, and to date there are no reported cases in Lebanon. In this paper, we present a case of full-term male neonate diagnosed with a congenital colon pouch in Lebanon. A full-term neonate presented with imperforate anus, abdominal distention, and vomiting. Diagnostic assessments revealed a well-encapsulated mass compressing the intestines and ureter. Surgical intervention identified a type I CPC with a meconium-filled pouch directly connected to the small intestine, and an absent ileocecal valve, prompting an ileostomy. Following the surgery, no postoperative complications were noted. Early imaging can help address the diagnosis to start the appropriate management, plan for surgery to prevent the development of a megacolon and therefore, perforation. The objective is to be aware and consider congenital colonic pouch diagnosis after encountering newborns with such clinical presentation in order to direct clinical investigations toward the diagnosis and treatment of the case early, thus reducing the risk of complications and improving the patient's quality of life.

Introduction

Congenital pouch colon (CPC) is a rare congenital abnormality, in which a pouch-like dilatation partially or completely replaces the colon, creating a fistula with the urogenital tract. Depending on the length of the normal colon proximal to the colonic pouch, it is divided into four types (I-IV) [1,2]. In males, the pouch usually terminates in a colovesical fistula just proximal to the bladder neck. While in females, the terminal fistula

Abbreviations: CPC, Congenital pouch colon; AMR, Anorectal malformations; NVD, normal vaginal delivery; NPO, Nothing by mouth; NICU, neonatal intensive care unit; OG, orogastric; GBS, Guillane Barre Syndrome.

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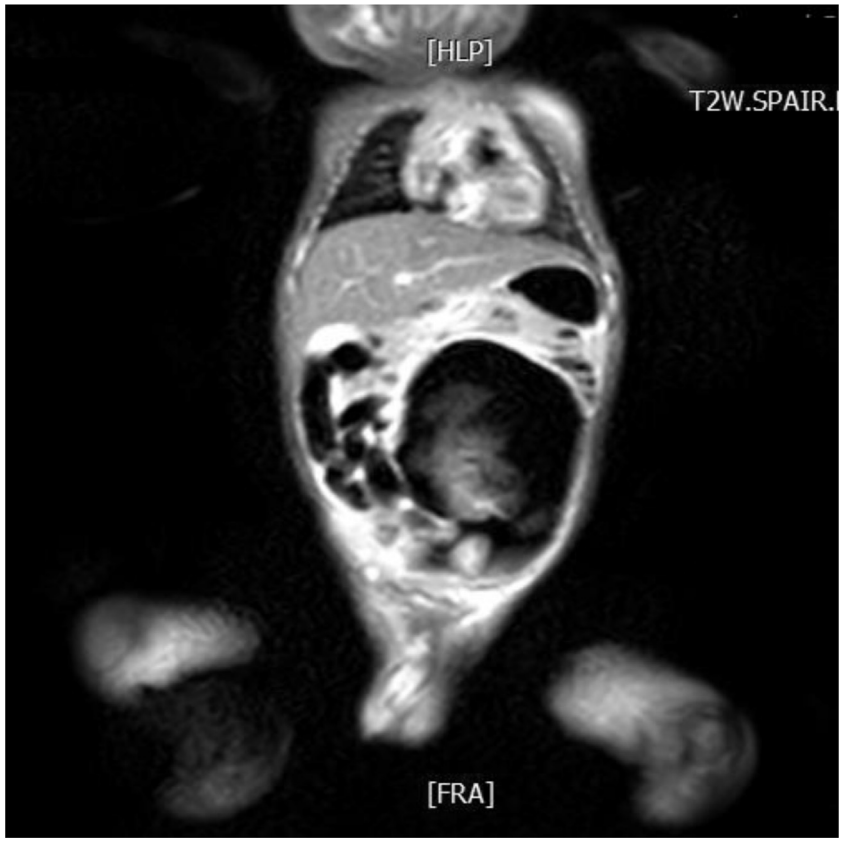

Fig. 1 – MRI anterior view showing a well encapsulated mass containing large amount of gas, compressing and displacing the small intestines forward.

opens either into the urethra or in the vestibule, close to the urethral opening. Females usually have 2 vaginas with a wide inter-vaginal bridge, a monocornuate uterus on each side, and urinary incontinence due to a widely open bladder neck. Males tend to be 2 to 7 times more affected than females [1,2]. CPC is listed as a rare variant in the Krickenbeck classification of anorectal malformations (ARM) [1]. CPC is much more common in India than in other countries as a review study showed that 92.2% of the reported cases were from India [1]. Congenital colonic pouch is an extremely rare congenital disease that has no reported cases in Lebanon. In this paper, we present a case of full-term male neonate diagnosed with a congenital colon pouch in Lebanon.

This manuscript was prepared following the CARE guidelines (https://www.care-statement.org).

Case report

Patient presentation

A full-term male neonate born to a 41-year-old Lebanese G7P4A3 non-smoker mother, via normal vaginal delivery (NVD) with vigorous apgar score 8 and 9 at 1 and 5 minutes respectively and birth weight 3770 grams, was found to have imperforate anus upon physical examination. Sixteen hours after birth, the patient developed abdominal distention and started vomiting. The patient was immediately admitted to the neonatal intensive care unit (NICU) on room air, was kept NPO and on orogastric (OG) tube. The neonate did not develop respiratory distress. An abdominal ultrasound was done showing an intra-abdominal retroperitoneal mass with macro-calcification, with mass effect on proximal portion of the left ureter. X-ray and MRI abdomen was done and showed a left sided 55 × 75 × 85 mm well encapsulated mass containing large amount of gas, compressing and displacing the small intestines forward (Figs. 1-3). Minimal ascites was also found. Moreover, the patient had decreased urine output.

Prenatal history

Prenatally, the mother had a no history of genitourinary or respiratory tract infection. Guillane Barre Syndrome (GBS) was negative. Abdominal and pelvic ultrasound was done at 18 weeks of gestation, and showed calcified intra-abdominal mass. For further investigations a morphological scan was done at 19 + 2 weeks showing calcified mass inside the transverse colon causing mild obstruction, and mild bilateral pyelectasis.

Management

Exploratory laparotomy was done and showed a pouch-like colon that later on was treated surgically.

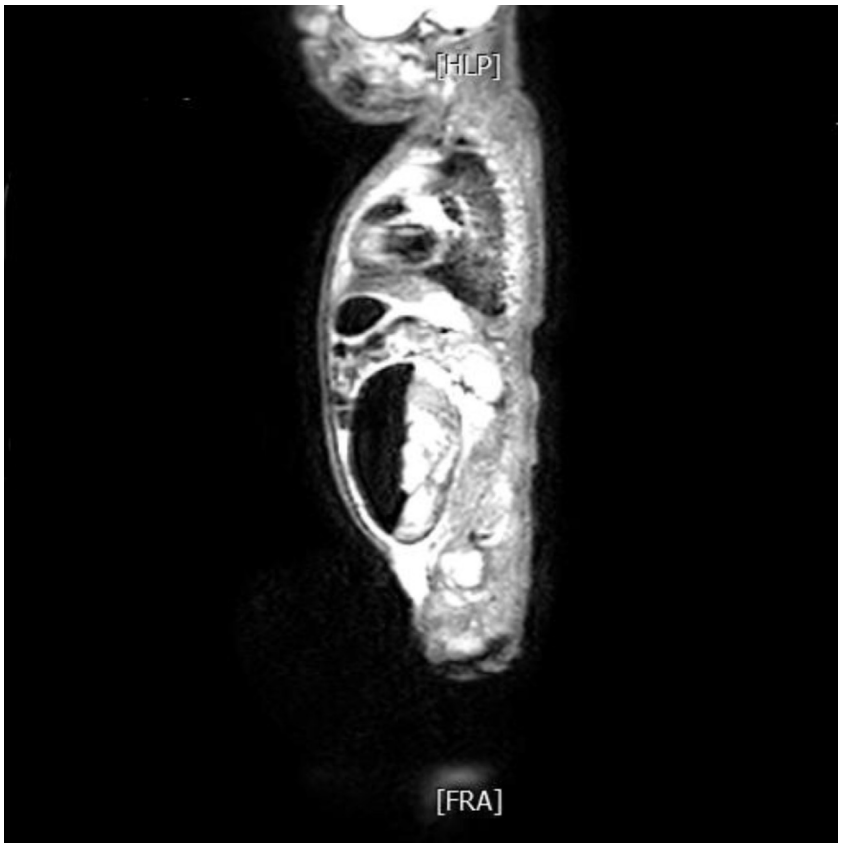

Fig. 2 – MRI lateral view showing a well encapsulated mass containing large amount of gas, compressing and displacing the small intestines forward.

Table 1 – Anatomical morphologic classification of CPC.

Type 1	Normal colon is absent, ileum opens into pouch
Type 2	Ileum opens into a normal cecum that opens into pouch colon
Type 3	Normal ascending colon and transverse colon open into pouch colon
Type 4	Normal colon with recto sigmoid pouch
Type 5	Double pouch colon with short normal interopositioned colon segment

On day 3 of life, a colostomy was planned to relieve the obstruction (imperforate anus) and to be followed by a subsequent surgery to address the colonic mass. During the surgery, after incision, a colon pouch filled with meconium was seen, so the pouch was opened and meconium was evacuated. Upon investigation, no ileocecal valve nor a large intestine were detected. The small intestine was directly connected to the colon pouch, type 1 CPC (Table 1). The rectum was directly connected to the colon pouch. For this reason, ileostomy was done instead of colostomy. The patient had Meckel's diverticulum which was resected alongside a part from the small intestines and was sent to pathology (Fig. 4). Histopathological findings were reported to be normal. Following the surgery, no postoperative complications were noted, and urinary output improved gradually.

Discussion

Early identification of CPC and differentiation from colon dilatation due to anorectal malformation is essential for the patient's welfare.

Colon dilatation differentials includes but are not limited to: pneumatosis intestinalis, defined as presence of gas in the intestinal extraluminal space; Colonic intussusception, which is an invagination of a bowel segment into the adjacent segment; and Hirschsprung which is a rare congenital intestinal disease and characterized by the absence of ganglion cells of the intestines [3–5].

CPC is more common in males, usually noticed in the neonatal period with abdominal distention, absence of anus, and intestinal obstruction. CPC is managed surgically depending on its type [1,6].

Congenital colonic pouch is an extremely rare congenital disease that has no reported cases in Lebanon. It presents with imperforate anus and abdominal distention which might be associated with obstipation and vomiting as in the case of this patient. It is a serious condition as it might lead to mega colon, bowel perforation, and early mortality [6]. What makes this study unique is its prevalence in Lebanon, in which this anomaly is exclusively seen in Northern India, hence all reported series are from this part of world with only a few cases reported from elsewhere. However our patient's clinical pre-

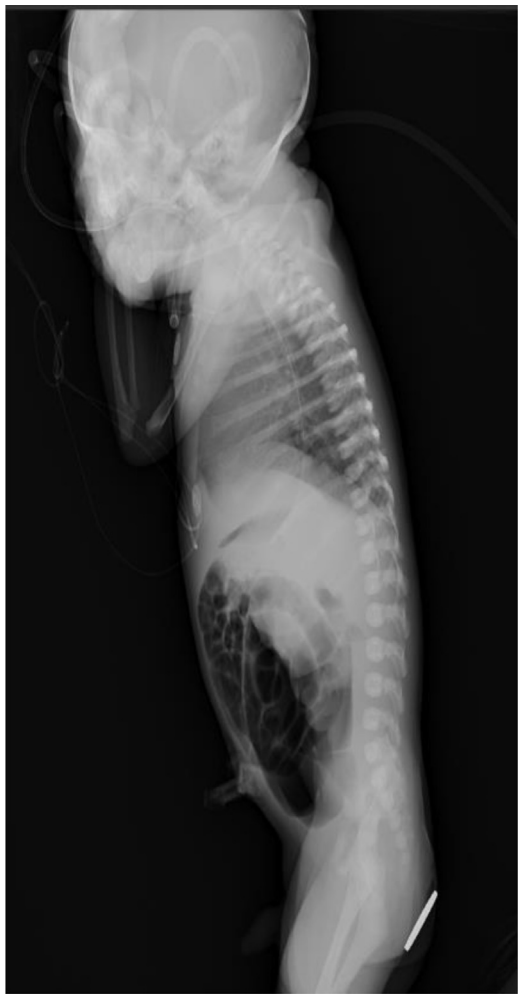

Fig. 3 – X-ray showing a dilated and pouch-like structure.

sentation was similar to other cases reported in several studies. Similarly, the majority of patients usually present within the first 72 hours of life with anorectal malformations, seen predominantly in males [6–8].

Early imaging can help address the diagnosis to start the appropriate management. The primary procedure that should be done for type I CPC, is colorrhaphy and diversion [6]. What was surprising that in our case, no fistula was detected intra operatively. Moreover, histopathological findings in our case were reported to be normal, contrary to a study previously published reported histological findings of mucosal, submucosal and serosal congestion, edema and hemorrhage as well as disrupted muscle layers [9].

The objective of this case is to be aware and consider congenital colonic pouch diagnosis after encountering newborns with such clinical presentation in order to direct clinical investigations toward the diagnosis and treatment of the case early, thus reducing the risk of complications and improving the patient's quality of life.

Strength and limitations of the study

The strength of this case report is the rarity of this congenital abnormality CPC, which is not commonly encountered in medical practice, and more specifically in Lebanon. Moreover, this case provides comprehensive details about the patient's prenatal history, presentation, multidisciplinary approach, diagnostic workup, surgical management, and the positive outcomes. This detailed case can serve as a useful reference for pediatricians and pediatric surgeons dealing with similar cases.

As for the limitations, the overall rarity of reported cases about there is limited data in terms of management and outcomes on this clinical condition.

Regarding limitations, due to the overall rarity of reported cases, there is a scarcity of data regarding the management and outcomes of this clinical condition.

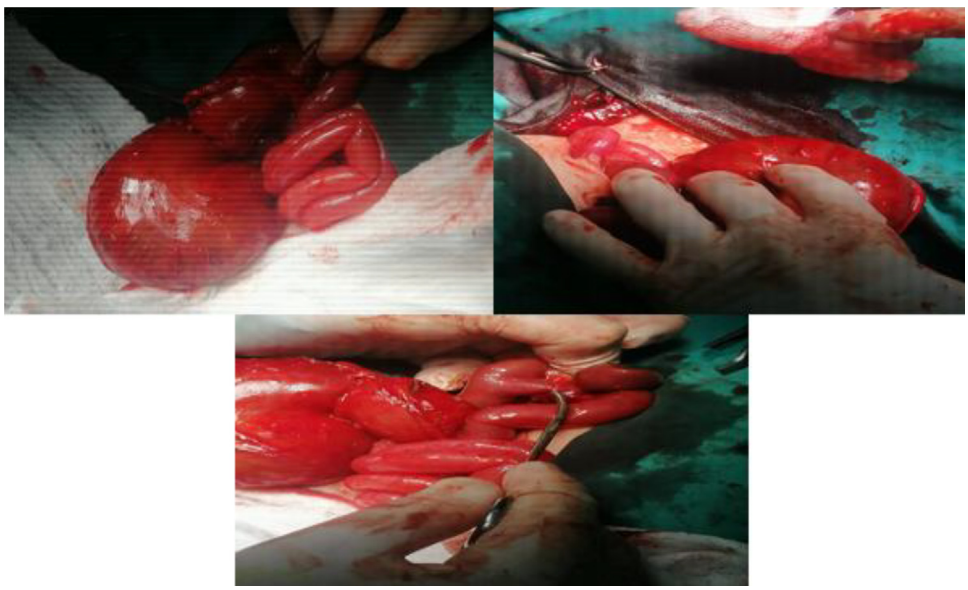

Fig. 4 – Surgical management of the congenital colonic pouch.

Conclusion

In summary, the purpose of this article is to report our encounter with an extremely rare congenital disease that has no reported cases in Lebanon. Congenital colonic pouch is a rare but serious condition that usually presents with imperforate anus and abdominal distention which might be associated with inability to pass stools and vomiting as in the case of this patient. Early imaging can help address the diagnosis to start the appropriate management, plan for surgery to prevent the development of a megacolon and therefore, perforation. The objective is to be aware and consider congenital colonic pouch diagnosis after encountering newborns with such clinical presentation in order to direct clinical investigations toward the diagnosis and treatment of the case early, thus reducing the risk of complications and improving the patient's quality of life.

Patient consent

The patient legal representative obtained an informed consent and granted approval of the publication.

IRB approval

The IRB at the hospital does not require further documentation if the data will be shared anonymously.

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