Congenital pouch colon: an unusual case report

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Abstract

Introduction. Congenital pouch colon (CPC) is a rare malformation. It causes variable dilatation of the colon associated with anorectal malformation (ARM), usually presenting a fistula towards the genitourinary tract.

Clinical case. 2-day-old female patient, with no relevant medical history. She had abdominal distension and imperforate anus. She underwent colostomy. She had an irregular evolution with high colostomy debits. Contrast imaging studies were performed, which demonstrated an erroneous exteriorization of the jejunal loop. She underwent an exploratory open surgery, which confirmed the previous diagnosis and incidentally found colonic agenesis, with ileum entering in a pouch of 6 cm of diameter that connects with the bladder.

Discussion. CPC is a common pathology in certain eastern countries and extremely rare in western countries. In Ecuador, no records of reported cases were found. A correct pre-surgical analysis of ARM patients should be carried out to achieve an adequate planning and surgical approach, thus reducing morbidity and mortality.

Key Words: Congenital pouch colon; Anorectal malformation; Segmental dilatation of the colon.

Bolsa colónica congénita. Reporte de un caso inusual

Resumen

Introducción. La bolsa colónica congénita (BCC) es una malformación poco común en la que se produce una dilatación variable del colon asociada a malformación anorrectal (MAR), generalmente presenta una fistula hacia el tracto genitourinario.

Caso clínico. Paciente femenino de 2 días de vida, sin antecedentes médicos de relevancia, presenta distensión abdominal y ano imperforado, es sometida a colostomía, presenta evolución irregular con débitos altos a través de la colostomía, se realizan estudios contrastados de imagen donde se observa exteriorización errónea de asa de yeyuno, se somete a laparotomía exploratoria donde se comprueba lo descrito y además se reporta como hallazgo incidental agenesia colónica con desembocadura del íleon en una bolsa de 6 cm de diámetro que se conecta con la vejiga.

Discusión. La BCC es una patología común en ciertos países orientales y extremadamente rara en países occidentales; en Ecuador, no se encontraron registros de casos reportados. Se debe realizar un correcto análisis prequirúrgico de los pacientes con MAR para conseguir una adecuada planificación y abordaje quirúrgico disminuyendo con ello la morbimortalidad en el paciente.

Palabras Clave: Bolsa colónica congénita; Malformación anorrectal; Dilatación segmentaria del colon.

INTRODUCTION

CPC is a rare malformation causing a variable dilatation of the colon, which is in turn shortened and associated with ARM. Typically, it presents a fistula towards the genitourinary tract, but sometimes it can end in a blind pouch[1-5]. CPC is more common in certain eastern countries. According to some studies, India represents 92.2% of the total cases reported globally[5]. In terms of sex, various studies have found a male-female relationship of 2.5:1[5-9]. CPC is one of the rare variants according to Krickenbeck's ARM classification[6]. In 1984, Narasimharao et al.[1] classified CPC into four subtypes (I-IV) according to the length from the proximal normal colon to the colonic pouch (Fig. 1), types I and II being the most severe ones. IAPS classification from 2007 mentions a V subtype encompassing the rarest variants of CPC[10].

Within this classification, type IV CPC is the most common presentation[11]. CPC incidence is eastern countries is believed to be higher than reported as a result of type IV CPC cases being erroneously considered as rectal ectasia, megasigmoid, or megarectum[5].

Clinical presentation varies according to sex. In newborn boys, it presents with imperforate anus, abdominal

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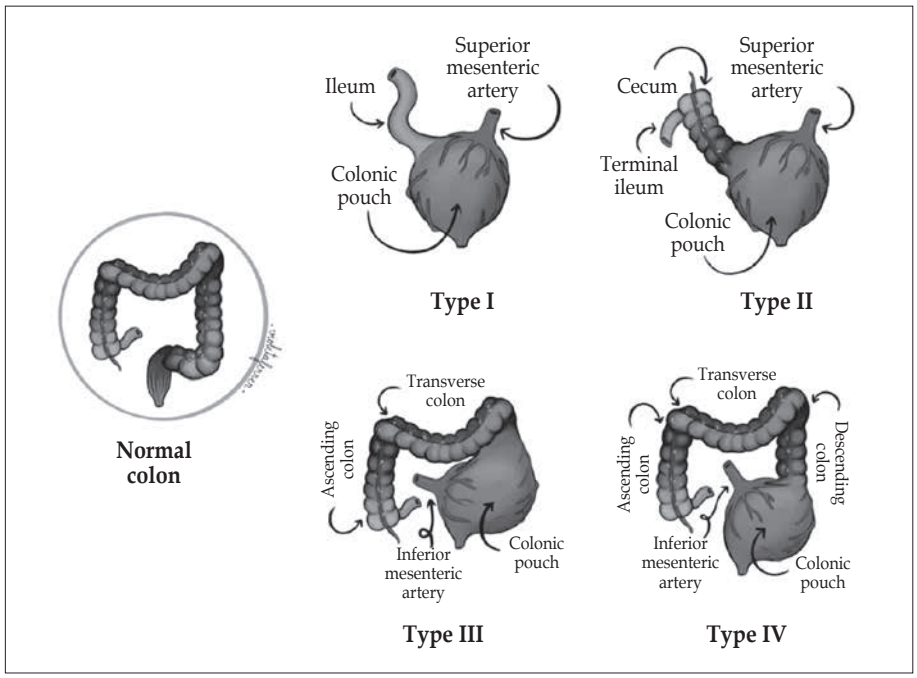

Figure 1. Classification of congenital pouch colon.

distension, and fecaluria in 50% of patients. Abdominal distension can be less evident in milder types (III and IV). In newborns, there is a high incidence of CPC perforation when it has thin walls, causing pneumoperitoneum, peritonitis, and sepsis[1,2,4,12].

In girls, most cases present with abdominal distension, and also with constipation and episodes of enterocolitis and urinary incontinence[12-15]. Diagnosis can be achieved based on the exploration of the external genitalia[13-15]. In case of doubt, a contrast can be placed through the vestibular fistula, if present[3,11].

CLINICAL CASE

2-day-old girl born full-term, without remarkable prenatal history. She presented abdominal distension and imperforate anus, so she underwent colostomy. She was then referred to a Pediatric Hospital for high debit colostomy management. No further findings were described in the surgical protocol. No previous imaging studies were available either.

An intestinal transit test was performed. It showed that the ostomy had been carried out at the level of the jejunum instead of the colon, so an exploratory open surgery was scheduled, which demonstrated jejunostomy and, incidentally, colonic agenesis, with ileum entering in a pouch of 6 cm of diameter that connects with the bladder – these findings were compatible with type I congenital pouch colon. End-to-end anastomosis was performed according to algorithms for jejunostomy closure purposes. A double-opening ileostomy was carried out 18 cm away from the colonic pouch described (Fig. 2), followed by tubular colorraphy, colovesical fistula repair, posterior sagittal anorectoplasty, and intestinal transit restoration.

DISCUSSION

In imaging studies (simple abdominal X-ray with anteroposterior ray), type I and type II CPC patients typically present a gas shadow or hydroaerial level on the left side occupying up to 50% of the cavity, with small bowel loops being displaced to the right[1,2,12]. In the lateral projection or prone position, gas can be detected inside the bladder in case of colovesical fistula. In most cases, diagnosis can only be achieved using abdominal-X-ray[16]. In case of CPC perforation, a pneumoperitoneum is found instead of the image previously described.

In type III and type IV CPC cases, similar images are found, but with a lower airflow as they are less severe. The dilated type IV CPC loop has a different position owing to its greater mobility, so it may be difficult to distinguish from terminal rectosigmoid dilatation[16].

This case was aimed at reporting our experience with a rare pathology, which has no reported cases in Ecuador. Clinical presentation in this patient was similar to that of an ARM with imperforate anus and abdominal distension at birth. Pre-surgical imaging studies did not allow the pathology to be diagnosed, so colostomy was attempted, but if failed. Therefore, the patient was referred to a specialized hospital. New imaging studies and an exploratory open surgery were carried out, which incidentally demon-

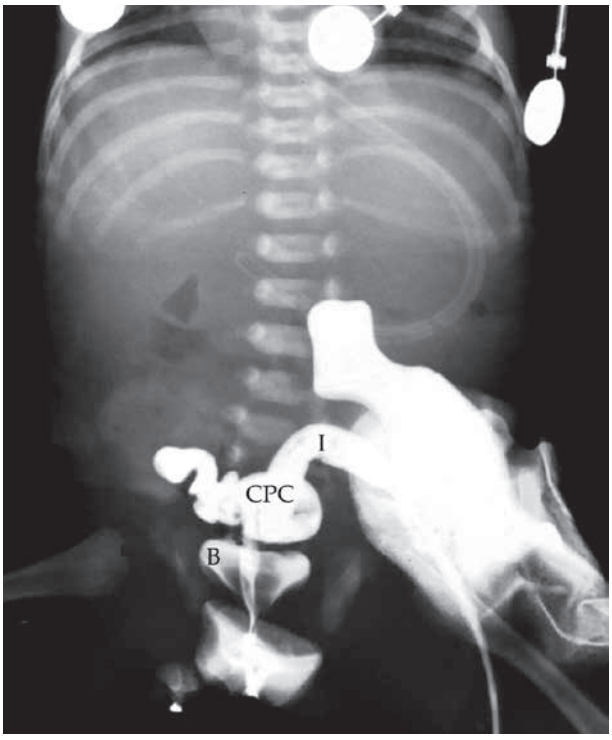

Figure 2. Post-surgical contrast imaging study through ileostomy (I). A type I congenital pouch colon (CPC) with a fistula towards the bladder (V) is observed.

strated the aforementioned findings, compatible with type I congenital pouch colon.

Congenital pouch colon is an extremely rare pathology, and even more in Ecuador. An appropriate pre-surgical analysis of ARM patients should be carried out to achieve an adequate planning and surgical approach. The objective is to accomplish optimal results with the smallest number of surgical procedures, thus reducing morbidity and mortality.

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