Case Report

Type II Congenital Pouch Colon in a Female Child

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Abstract

Congenital pouch colon (CPC) is a condition where the colons are dilated and pouch-like and lack tenia coli and haustration. CPC is also known as ‘pouch colon syndrome.’ Large gas shadow on the left side of abdomen occupying more than half of the abdomen and pushing residual bowel shadow to the right is the characteristic radiological finding of type II congenital pouch colon. We report a case of 4-day-old female child with congenital pouch colon underwent surgery but died because of overwhelming sepsis along with brief review of the literature.

Keywords: Anorectal Malformation; Congenital Pouch Colon; Coloplasty

Introduction

CPC is an uncommon congenital abnormality in which a pouch-like dilatation of a colon is associated with an anorectal malformation. It is also termed as ‘pouch colon syndrome.’ It is categorized into four subtypes (Types I–IV) based on the length of normal colon proximal to the colonic pouch. CPC is much more common in India. It is more common in males as compared to females (M: F, 3.5:1). Large gas shadow on the left side of abdomen occupying more than half of the abdomen and pushing residual bowel shadow to the right is factual radiological finding. Vascular insult during the gestational period from the obliteration of inferior mesenteric artery is the probable etiology for pouch colon. Early diagnosis and treatment may lead to improved outcome. This case report aims to bring attention to congenital pouch colon associated with anorectal malformation. We present a brief review of classification and management.

Case Report

A full term, four days old, 2.3 Kg weight, the female child was brought to an emergency room with the diagnosis of imperforate anus. An invertogram revealed high variety imperforate anus. The sacrum was normal. The abdomen was grossly distended, and bowel sounds were absent. Plain X-ray abdomen erect large gas shadow in the abdomen on the left side occupying more than half of the abdomen and pushing residual bowel shadow to the right.

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Figure 2: Large gas shadow in the abdomen on the left side occupying more than half of the abdomen and pushing residual bowel shadow to the right

Diagnosis of congenital pouch colon was made. On exploration, pouch lacked tenia coli and haustration. The pouch is a short segment of cecum proximal to the pouch (Type II). The whole pouch was excised, proximal ileostomy was constructed. The appendix was absent in this case. Postoperatively, unfortunately, baby succumbed to septic shock.

Discussion

Congenital pouch colon (CPC) is a condition where the colon is dilated and pouch-like and lacks tenia coli and haustration. The CPC is also known as ‘pouch colon syndrome’. Spriggs first described the congenital pouch colon (CPC) in 1912 [1]. CPC has now been accepted and included in the Krukenbeck Classification of Anorectal malformation (ARM) in 2005. 90% of the CPC was reported from Indian region predominantly from north India. It is more common in males as compared to females (M:F 3.5:1) [2]. The essential findings/peculiarities of CPC are [3]: Its association with the ARM, abnormal blood supply, an absence of haustration and appendices epiploicae, urogenital fistula, and gross VUR. Large gas shadow on the left side of abdomen occupying more than half of the abdomen and pushing residual bowel shadow to the right is characteristic radiological finding. Vascular insult during the gestational period from the obliteration of inferior mesenteric artery is the probable etiology for pouch colon. Low socioeconomic status, environmental factors, vitamin deficiency, and excess use of pesticides in the farm may also contribute to the development of CPC. There are two popular classifications for CPC.

Classification for CPC [4] is as follows:

Type 1 and 2: Single large pouch with the apex located in the left hypochondrium.

Type 3: The pouch apex was directed towards the right hypochondrium.

Type 4: The apex of the pouch was headed towards the right hypochondrium, and

Type 5: Radiograph was inconclusive.

Classified CPC into four types depending upon the length of colon involvement [5].

Type I: The Normal colon is absent, and the ileum directly opens into the colonic pouch.

Type II: The ileum opens into a short segment of cecum which then opens into the pouch.

Type III: Presence of large segment of the normal colon between the ileum and the colonic pouch.

Type IV: Presence of nearly normal colon with the only terminal portion of the colon (rectum and the different portion of sigmoid) converted into a pouch.

Management of CPC based on Saxena - Mathur classification [6]:

In type 1 and type 2: pouch excision and pull-through or 3-stage procedure: ileostomy, pouch-coloplasty with pull-through, and ileostomy closure.

Type 3 and type 4 (3 stages): pouch excision with colostomy, pull-through, and colostomy closure.

Type 5 (3-stages): Distal pouch excision with proximal pouch-coloplasty with ileostomy, pull-through, and colostomy closure.

In the review of large series of 566 cases, Most common anomaly reported is absent appendix (n=61) followed by hydronephrosis in 40 cases. Least commonly reported anomalies is hemivertebrae (n=4) [7].

Conclusion

CPC is a rare congenital abnormality and radiograph of the abdomen erect is diagnostic. Management depends on the types of CPC. A high index of suspicion and early diagnosis and treatment may lead to improved outcome.
References


