J Interdiscipl Histopathol 2014; 2(3): 173-178 *ISSN: 2146-8362*

Case Report

Congenital Pouch Colon: A Rare Variant of Anorectal Malformation: Histopathological Perspective with Brief Review of Literature

Dhiraj B Nikumbh[1,2], Kishor H Suryawanshi[1,2], Sudhir Singhavi[3], Poonam Pagare[2], Akshay Surana[2]

Department of Pathology[1], ACPM Medical College[2], and Consultan Pediatric Surgeon[3], Dhule, Maharashtra, India

Received: January 16, 2014

Accepted: February 23, 2014

Published Online: February 24, 2014

DOI: 10.5455/jihp.20140223111355

Corresponding Author:
Dhiraj B Nikumbh, MD
Associate Professor
Department of Pathology,
ACPM Medical College,
Dhule, Maharashtra, India
drdhirajnikumbh@rediffmail.com

Keywords: Congenital pouch colon; anorectal malformation; congenital anomaly

Abstract

Congenital pouch colon is an extremely rare variant of anorectal malformation (ARM) in which all or part of colon is replaced by a pouch like dilatation that communicates distally with the urogenital tract by a large fistula. Congenital pouch colon differs from normal colon structurally, histologically and functionally. The incidence of congenital pouch colon among all cases of anorectal malformation in northern India has been reported to be between 5-10%. Rest of the India and around the globe handful of documented case reports was noted. The mortality from congenital pouch colon has decreased from 40% to 15%, if identified and managed properly.

Herein, we report a case of 10 days male child presented with congenital pouch colon in view of a rare congenital anomaly. We enlighten the detailed histopathological findings in this case as there is paucity of literature of congenital pouch colon.

© 2014 GESDAV

INTRODUCTION

Congenital pouch colon (CPC) is a rare condition in which there is a short, pouch like dilated colon and is associated with anorectal malformation (ARM) [1, 2]. The pathogenesis and embryology of the CPC is not well understood but dietary, environmental and familial inheritance may be contributing factor for this pathology [2]. In CPC all or part of the colon is replaced by a pouch like dilatation that communicates distally with the urogenital tract by a large fistula [3].

This condition is more common in northern population of India and neighboring countries like Pakistan and Nepal with 90% cases reported from India [4]. With the growing awareness there were few case reports around the globe of the CPC [4]. The mortality has been decreased from previously 40% to 15%, if identified and managed properly [5].

Though enough literature is available on the descriptive and management aspect of CPC associated with ARM, there is paucity of its detailed histopathological findings [6]. Herein, we report a case of 10 days male child presented with congenital pouch colon with detailed histopathological findings. We report this case due to its rarity and paucity of literature regarding pathological perspective.

CASE REPORT

A 10-days-male child presented to the pediatric department of our hospital with distension of the abdomen and absent anal opening. On physical examination there were no obvious other congenital anomalies. Clinically it was diagnosed as high anorectal malformation. Plain X-ray abdomen revealed dilated bowel loops. Hematological and biochemical parameters were within normal limits. Emergency laparotomy was performed by pediatric surgeon. On exploration, pouch colon was noted with rectovesical fistula. Excision of the pouch colon with end colostomy was done and the specimen was sent for histopathology. Postoperative period was uneventful.

Gross examination: The specimen was measured as 6x4x2 cms in dimension. The external surface is cyst/pouch like dilated with congested thick walled blood vessels. Haustrations and taenia coli were not seen (Figure 1). On cutting the colon, fecal matter was observed in the lumen of the specimen. The mucosa was generally attenuated with focal areas of congestion and hemorrhages (Figure 2).

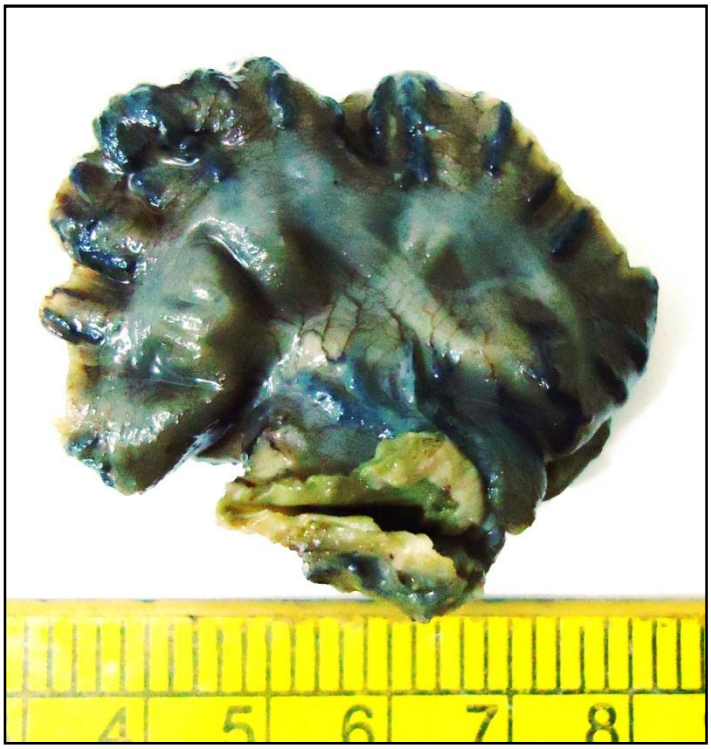


Figure 1: Gross specimen of resected pouch colon with loss of haustration and taenia coli with dilated and congested thick walled blood vessels.

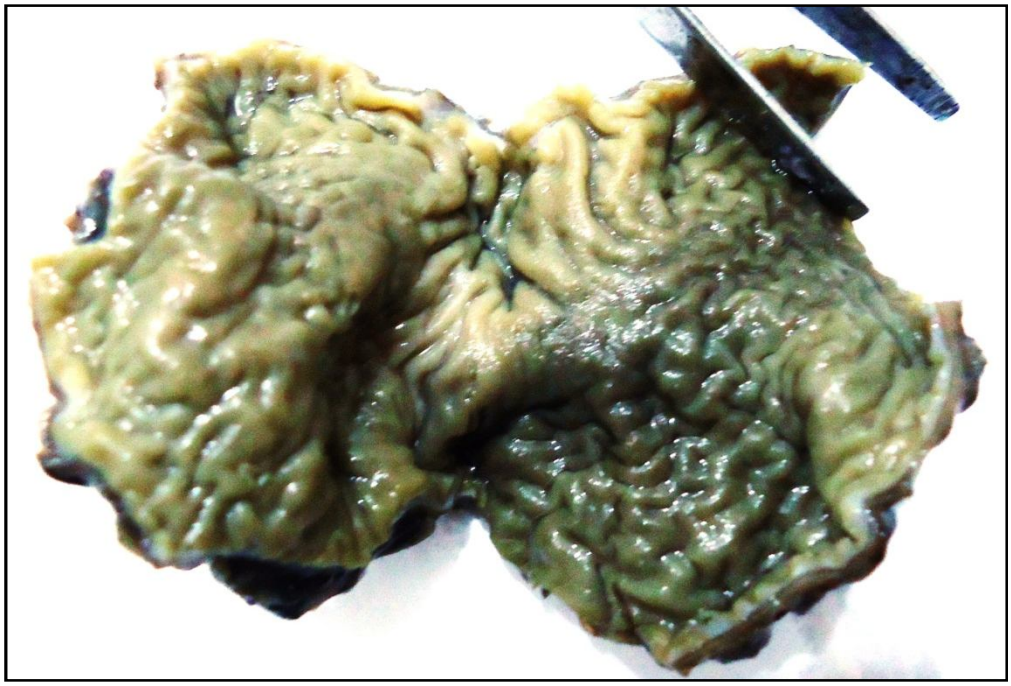
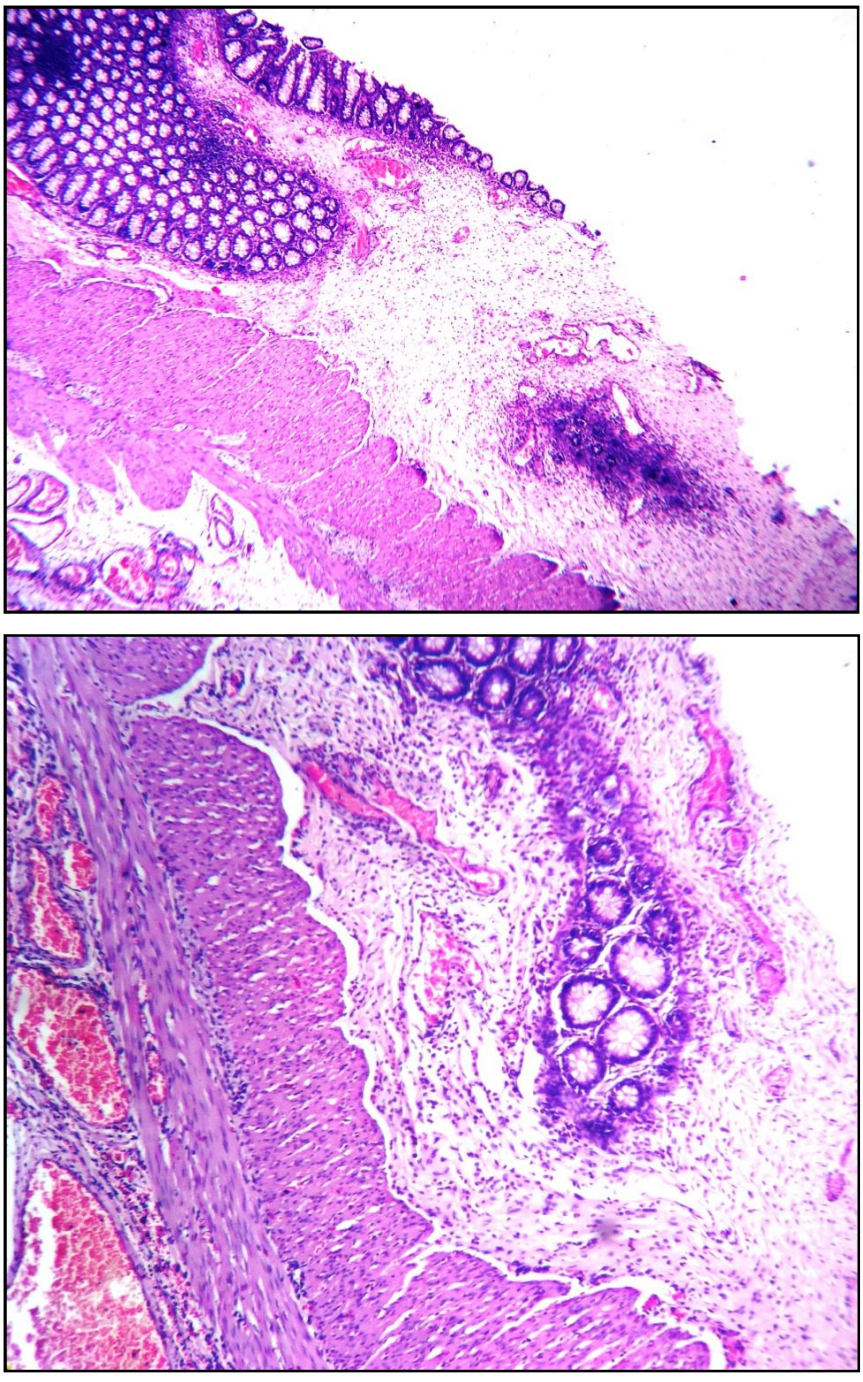
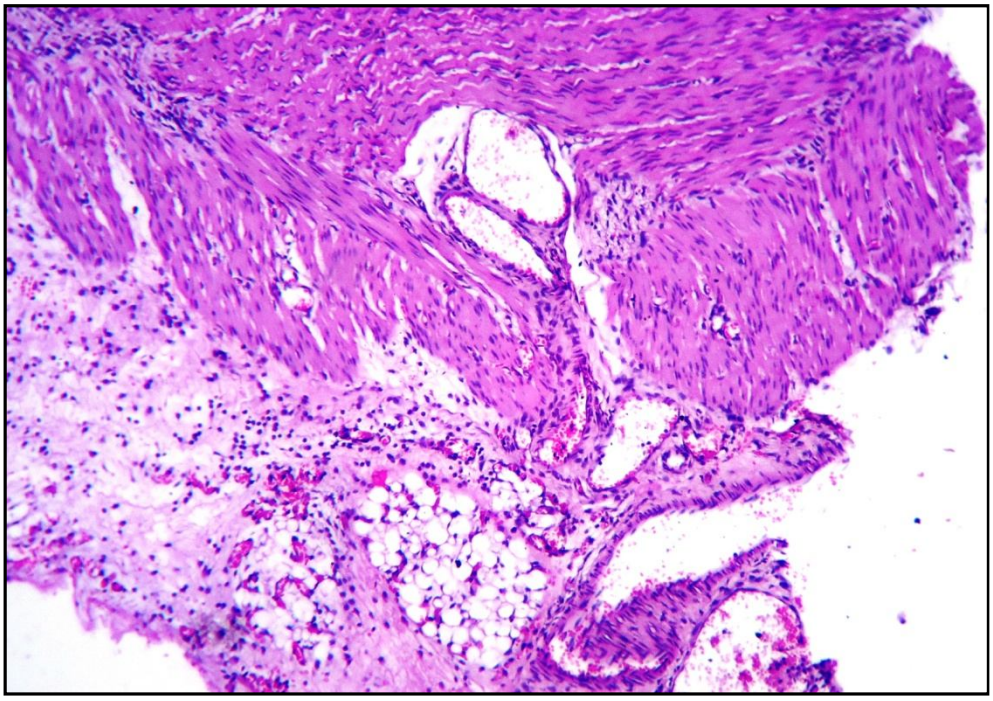


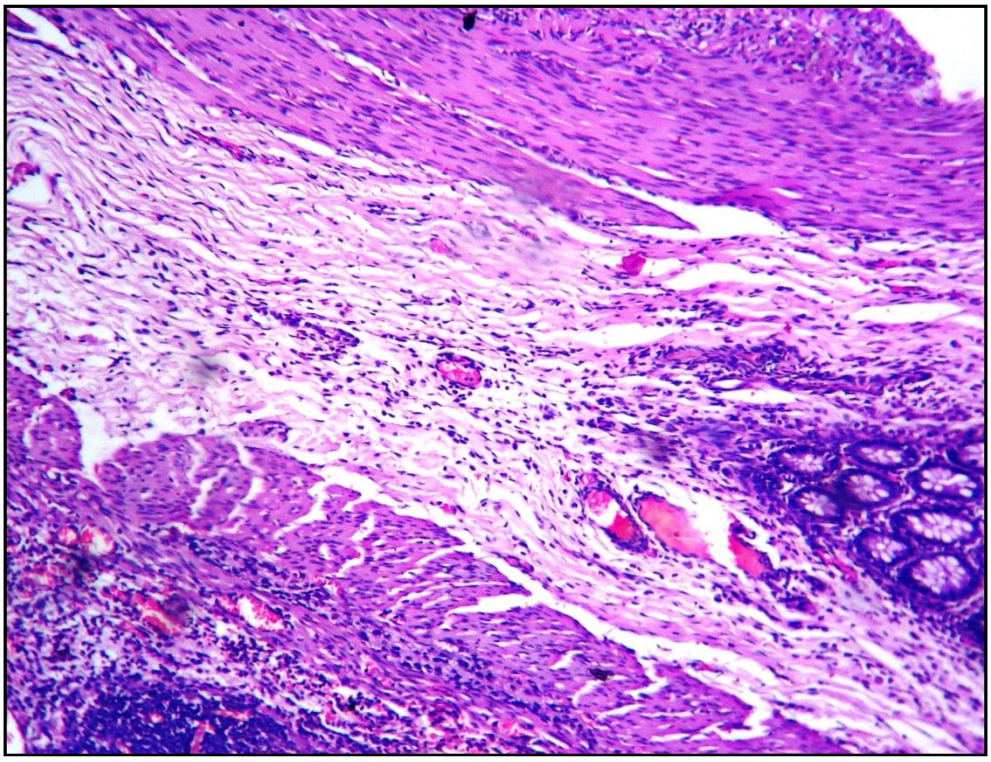
Figure 2: Cut surface of CPC with thickened wall and flattening of mucosa with focal hemorrhagic areas.

Light microscopy: Histologic sections revealed large intestinal mucosa with focal areas of congested blood vessels and focal dense and mild diffuse mononuclear cells infiltration (Figure 3, 4). Prominent lymphoid follicles were also noted in the histologic sections. Congested and thrombosed thick blood vessels leading dilatation were seen in submucosa. The muscular layers demonstrated focal areas of fibrosis with hypertrophic muscle fibers. Disarray of muscle coats was noted. Focal dipping and narrowing of the muscular coat at inner levels was evident (Figure 5). Epithelium was noted in subepithelial areas with fibrous stroma (Figure 6). The ganglions in between muscle fibers were immature and hypertrophied. Some of them had giant forms. Congested and thrombosed blood vessels were noted in serosa. Few lymphoid aggregates were noted in subserosal area. Final histopathological impression from resected specimen of colon (anorectal malformation) was congenital pouch colon.



Figures 3 and 4:
Photomicrograph showed large intestinal mucosa with congested blood vessels and diffuse mononuclear inflammatory cell infiltration (H&E, x100).





Figures 5 and 6: Photomicrograph showed disarray of muscle coats with focal dipping and narrowing. Epithelium was noted in subepithelial areas with fibrous stroma (H&E, x400).

DISCUSSION

Congenital pouch colon was first described in 1912 by Spriggs in a London hospital museum specimen with the absence of the left colon and rectum. Trusler in 1959 described a pouch like dilatation of shortened colon associated with high ARM. Spencer in 1965 reported 53 cases, out of which 43 were with extrophy of the bladder and intestines were called typical and atypical "extrophia splanchanica". Blunt in 1967 discussed this condition as an absence of colon and rectum. Shafie in 1971 described this condition as a cystic dilatation of the colon [7].

In India, CPC was first reported by Singh and Pathak (1972) in a series of six cases with name "Short Colon". Gopal (1978) called it a colonic reservoir in a case with rectovaginal fistula [7]. In 1981, Li from China named it as "Congenital atresia of anus with short colon malformation". Narsimha Rao et al [8] in 1984 suggested the name "Pouch Colon Syndrome" and proposed an anatomical classification of this condition which has been widely accepted.

The incidence of CPC varies in different parts of the world. Apart from the northern part of the Indian subcontinent, there are only sporadic case reports from the other parts of the world including China, Japan, Sweden, UK and USA [3, 4, 7]. In the northern India, the incidence of CPC among all the cases of ARM has been reported as 5-10% [7]. The incidence in tertiary centers is 10-15% of all cases of ARM and even up to 20% of all cases of high anomalies. CPC is more common in males [7].

The etiopathology and embryology of CPC is not known, but environmental factors with deficiency of iodine and Vitamin B12 are some of the possible factors contributing to this anomaly in north Indian belt as per Gupta et al [7].

CPC is classified into four types based on the length of abnormal colon [8]:

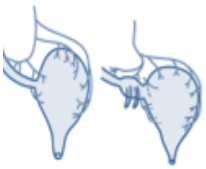
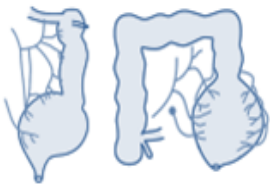
Type I: Normal colon is absent and ileum directly opens into the colon pouch.

Type II: The ileum opens into the short segment of caecum which then opens into the colonic pouch.

Type III: Presence of a significant amount of normal colon between the ileum and the colonic pouch.

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

The classification of congenital pouch colon [3]:

Complete CPC	Incomplete CPC
	
If there is either no or little normal colon left that is not enough for performing the pull through. In this situation, a coloplasty procedure would be required to retain only 15 cm length of pouch colon in the form of a tube, to be brought out as an end colostomy. A pull through procedure at the time of performing coloplasty should not be preferred in the newborn stage as it is associated with high morbidity and mortality	Where the length of the normal colon is adequate enough for performing the pull through, without the need for doing a coloplasty. The procedure would involve excision of pouch with an end colostomy at birth definitive pull through later. A single stage pull through in the newborn stage can also be undertaken if the condition of the baby permits.

CPC should have following anatomical criterion [7]:

1. There is anorectal agenesis.
2. Total length of colon is short.
3. Colon has a pouch formation for a varying length-Saccular or Diverticulum formation with the collection of meconium or fecal matter.
4. The blood supply of pouch is abnormal. The colon wall is thick and muscular with hypertrophied mucosa.
5. The fistula with the genitourinary tract is large muscular and long. It is closely adherent with the bladder wall.
6. There is no transition zone between the pouch and the normal bowel. The pattern changes suddenly and sharply.

The above anatomical features were present in our case.

Pathological Aspect: The CPC has a short pouch and poorly developed mesentery. The wall of the pouch is thick and taenia coli, haustration and appendices epiploicae are absent or ill-defined [7]. Microscopically, the mucosa shows congestion and focal hemorrhages in CPC. The muscles are also arranged in haphazard and decussating pattern [7]. The most salient feature was disorganization of the muscle coat in an arborizing manner [7, 8]. The gross and microscopic features were concordance with above findings in our case.

Clinically most of the patients of CPC presented in the early neonatal period with an absent anal opening and distension of the abdomen with or without meconium. Early gross distension of abdomen and bilious vomiting in a case of ARM is strongly suggestive of CPC [7, 8]. Associated genitourinary malfunction (cloacal anomalies, double vagina and extrophy) are common in girls [7].

When a fistula connection with urogenital tract is large, prognosis is bad in some instances. In our case, 10 days old male baby presented with abdominal distension and imperforate anus with rectovesical fistula and CPC.

Regarding treatment of CPC, the aim of surgery is to utilize the available length of colon for absorption and storage capacity as well as capability for propelling the fecal matter onwards with continent anal opening [7, 8]. The treatment choices are excision in incomplete CPC and coloplasty in complete CPC. In our case, excision and end colostomy was done.

To summarize, CPC associated with ARM is an abnormally developed tissue and needs to be resected for better functional outcome of the remaining gut. The crux of the treatment lies on timely diagnosis and planned management. Hence it is important for gastroenterologists and pediatric clinicians to be aware of the pathological perspective for proper diagnostic evaluation and surgical management. We present this case, in view of the paucity of the literature regarding pathological perspective.

CONFLICTS OF INTEREST

Authors declare that they have no any conflict and disclosure.

REFERENCES

1. Chadha R, Gupta S, Mahajan JK, Bagga D, Kumar A. Congenital pouch colon in females. Pediatr Surg Int 1999; 15(5-6): 336-342.
2. Chadha R. Congenital pouch colon associated with anorectal agenesis. Pediatr Surg Int 2004; 20(6): 393-401.
3. Gupta DK, Sharma S. Congenital pouch colon. In: Hutson J, Hulschnider A (eds) Anorectal malformations. 1st edition, Springer Berlin Heidelberg, New York, pp 211-222, 2006.
4. Arestis NJ, Clarke C, Munro FD, Micallef C, O'sullivan MJ. Congenital pouch colon (CPC) associated with anorectal agenesis. A case report and review of literature. Pediatric Dev Pathol 2005; 8(6):701-705.
5. Sharma S, Gupta DK, Bhatnagar V, Bajpai M, Agrawal S. Management of congenital pouch colon in association with ARM. J Indian Assoc Pediatr Surg 2005; 10:s22.
6. Ganagopadhyay AN, Patne SC, Pandey A, Aryya NC, Upadhyaya VD. Congenital pouch colon associated with anorectal formations-histopathological evaluation. J Pediatr Surg 2009; 44(3):600-606.
7. Gupta DK, Sharma S. Congenital pouch colon-Then and now. J Indian Assoc Pediatr Surg 2007; 12(1): 5-12.
8. Narsimha Rao KL, Yadhav K, Mitra SK, Pathak IC. Congenital short colon with imperforate anus (Pouch colon syndrome). Ann Pediatr Surg 1984; 1: 159-167.

This is an open access article licensed under the terms of the Creative Commons Attribution Non-Commercial License which permits unrestricted, non-commercial use, distribution and reproduction in any medium, provided the work is properly cited.