CASE REPORT

ULTRASOUND FINDINGS OF CONGENITAL POUCH COLON, A VARIANT OF ANORECTAL MALFORMATION

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ABSTRACT

SUMMARY: Congenital pouch colon (CPC) is a variant of anorectal malformation in which all or part of the colon is replaced by a pouch-like dilatation that communicates distally with the urogenital tract by a large fistula. It is associated with variable length of proximal normal colon. This case report illustrates the postnatal sonographic findings of a baby with type IV CPC, a rare presentation. The aim of this report is to highlight the importance of a careful examination of the morphology as well as the location of the fetal bowel.

Keywords: congenital pouch colon, anorectal malformation

Background

Congenital pouch colon is categorized into four subtypes (Types I-IV) based on the length of normal colon proximal to the colonic pouch.[2] Over the years, incidence of type IV CPC, which is less severe form, is increasing with low ARM.[3-6] This condition has a male preponderance with a male to female ratio of 2.25:1.[3] Type IV CPC has a near normal colon with only its terminal portion converted into a pouch.

The pathognomonic feature of type I and II congenital pouch colon best seen on anteroposterior xrays, is the presence of a large gas bubble of the colon, which occupies more than 50% of the abdominal width in a child with anorectal agenesis.The small bowel loops are displaced to the right.[5] Mostly in Type III and IV CPC, a smaller air-fluid level is seen on a plain X-ray, In males with Type IV CPC, several reports suggest that the colonic pouch ends in a colovesical fistula, so a distal sigmoid colostogram can demonstrate a CVF opening just above the bladder neck, and is designated as “true” Type IV CPC, often with gas in the bladder on plain X-ray. A prone cross-table or lateral film also better visualizes gas within the bladder. Though, in most cases, diagnosis is possible by abdominal radiographs alone. However, It is sometimes difficult to differentiate X-ray findings from the terminal rectosigmoid dilatation in anorectal malformation, although, the air-fluid level in type IV CPC is usually better defined. Barium enema reveals dilated sigmoid and rectum with abrupt transition to normal colon.[5] On ultrasound, there are signs of cystic enlargement of the colon.[5]

Pouch colon may be a cause of intractable constipation in children operated for low anorectal anomaly. Excellent results can be obtained by excision of the pouch.[6] Tactics of surgical correction and prognosis are largely dependent upon the type of CPC and of associated ano-rectal malformation.

Case Presentation

A one day old male baby, born at term through SVD was brought to the pediatric outpatient department of Liaquat National Hospital with antenatal scan done at 33 weeks showed a large cystic structure at the left side of fetal abdomen. On examining the neonate

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at birth, imperforate anus was noticed. His ultrasound abdomen was advised which showed large cystic area behind the bladder extending upto the left hypochondrium with air specks and echoes. This represent dilated large bowel segment most likely rectum and sigmoid colon.

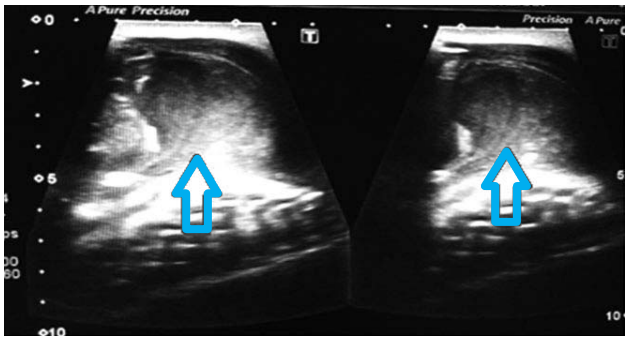

Figure 1: Shows dilated cystic structure with echoes on left side of abdomen (blue arrow)

He went for laprotomy which showed large pouch of sigmoid colon (blue arrow) with a broad base communication with normal descending colon proximal to it (*). These finding represent type IV pouch colon. End colostomy proximal to the pouch was done and a biopsy was sent from the hypertrophied sigmoid segment.

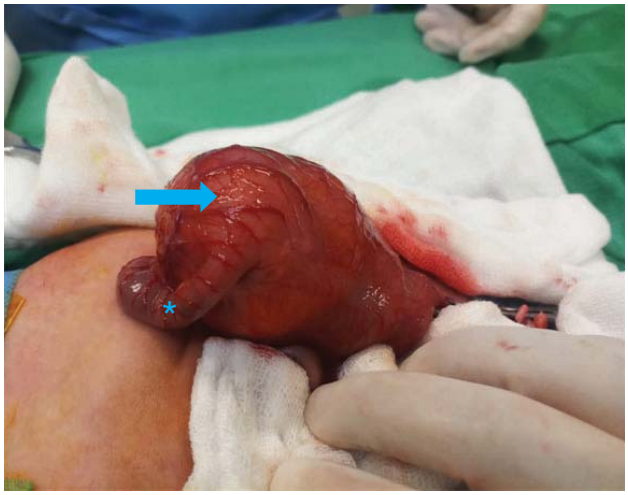

Figure 2: Shows excision of dilated pouch of colon (blue arrow) with normal colon proximally (*)

Later histopathology of the specimen revealed benign large bowel wall exhibiting folds of mucosa with intact crypt artichecture and intact epithelium with mild chronic inflammation and unremarkable muscle layer. Multiple dilated and congested vessels seen in subserosal layer. All these features are compatible with clinical diagnosis of pouch colon.

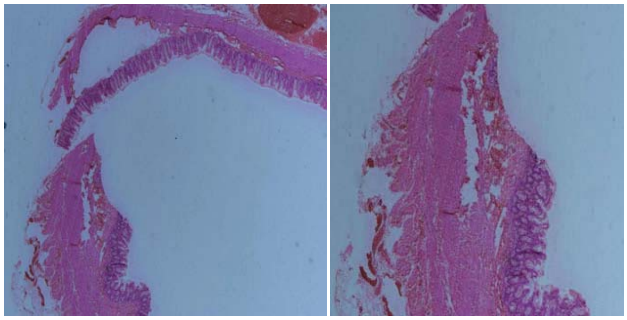

Figure 3: Shows normal large bowel wall mucosa with intact crypt artichecture and intact epithelium with congested vessels seen in subserosal layer

Discussion

Congenital pouch colon (CPC) is an unusual abnormality in which a pouch-like dilatation of a shortened colon is associated with an anorectal malformation.[1,5] CPC is an extremely rare variant of anorectal malformation in which all or part of the colon is replaced by a pouch-like dilatation that communicates distally with the urogenital tract by a large fistula. They are divided into four types.[2]

Type I - Normal colon is absent and the ileum opens directly 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 a significant length of normal colon between the ileum and the colonic pouch.

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

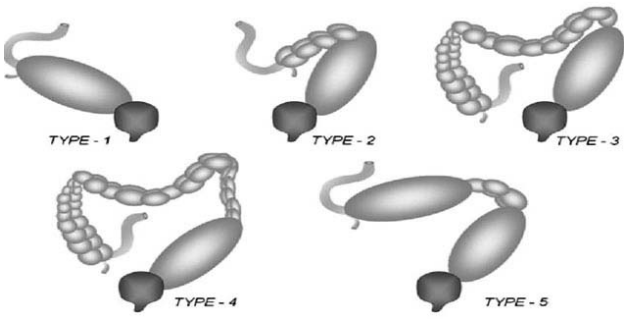

Figure 4: Shows types of pouch colon

Major malformations are uncommon associations with CPC however if major abnormalities are present,

suggesting an early, severe error in embryogenesis.[2] The more severe Types I/II CPC can usually be diagnosed by a large gas shadow or air-fluid level on X-Ray abdomen, occupying more than half of the total width of the abdomen and displacing the small intestine to one side (usually right).[3,4]

The embryogenesis of congenital pouch colon is still unknown. Various theories were proposed including chronic obstruction, obliteration of the inferior mesenteric artery early in fetal life, primary disorder of the proximal end of the hindgut or postsplenic gut, faulty rotation and fixation of the colon, vascular insult at the time of the partitioning of the cloaca by the urorectal septum, and the combined effect of defective development of the splanchnic layer of the caudal fold and failure of rotation of the gut. These factors may affect the in utero development of hind gut and differentiation into urinary and intestinal tracts.[4]

Its treatment involves a diversion colostomy at birth with or without the excision of the pouch followed by a pull-through.[4] For all sub types of CPC, it is preferable to preserve a segment of the pouch by fashioning a narrow colonic tube for pull-through, the technique known as coloplasty or tubular colorraphy.[5]

Conclusion

Congenital pouch colon is a rare variant of anorectal malformation, but it is necessary to carefully examine the fetal bowel anomaly and its location in abdomen, in order to do surgery in earliest postnatal period.

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