Case Report

Segmental dilatation of colon associated with anorectal malformation

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ABSTRACT

Children with segmental dilatation of the colon suffer severe constipation and are clinically indistinguishable from Hirschsprung disease. Segmental dilatation of colon is rare in neonates. In this report, a neonate with unusual combination of segmental dilatation of the colon and high anorectal malformation is presented.

KEY WORDS: Anorectal malformation, pouch colon, segmental dilatation

INTRODUCTION

Segmental dilatation of the colon (SDC) is a rare entity. The affected children usually suffer from chronic constipation and the clinical course and the radiological appearance of the colon may resemble that of Hirschsprung’s disease. Since its first description by Swenson and Rathhouser in 1959, the entity has been reported mostly in children beyond neonatal period. No segment of colon is aganglionic and the anorectum is normal unlike pouch colon syndrome (PCS) that presents in the neonatal age and is almost always associated with an anorectal malformation (ARM). A review of English-language literature found only eight cases of SDC in neonates and the association of ARM with SDC was seen in only four cases.

We report a neonate having ARM and a grossly dilated ascending, transverse and descending colon with normal caliber sigmoid colon distal to the dilated segment.

CASE REPORT

A full term, 2.4 kg, male baby, born to a primigravida mother by normal vaginal delivery, presented to us on 3rd day of life with bilious vomiting, gross abdominal distension, and an absent anal opening. Physical examination revealed an active, hemodynamically stable baby with bilious nasogastric aspirates and abdominal distension. Perineal examination showed an absent anal orifice and well formed buttocks. External genitalia were unremarkable. Abdominal X-ray showed pneumoperitoneum with distended bowel loops. There were no associated vertebral anomalies.

The baby was taken up for emergency laparotomy. At operation, the ileum was seen to enter into a normal looking cecum from which a single appendix originated. The cecum opened into a large pouch. The pouch lacked haustrations, appendices epiploicae and taenia coli. It occupied most of the right side and the upper abdomen and was perforated at its antimesenteric border. There was no ascending, transverse or the descending colon. The pouch continued at its distal end as a normal caliber sigmoid colon going deep into the pelvis behind the bladder. The vascular supply to the dilated segment originated from a branch of the superior mesenteric artery, whereas branches from the inferior mesenteric artery supplied the sigmoid colon. The length of the entire colon as such was short. The dilated segment was excised along with the cecum and appendix [Figure 1]. The distal ileum was brought out as an end ileostomy and the sigmoid colon as a mucous fistula. The baby was discharged on the 8th post-operative day. Post-operatively, ultrasonography of the abdomen showed hydronephrosis in both the kidneys, however, the biochemical renal function tests were normal. Baby will further undergo radionuclide renogram assessment at 4-6 weeks of age.

The histopathology examination of the specimen showed...
sparse distribution of muscle fibers, normal ganglion cells, and vasculature.

**DISCUSSION**

The salient feature of SDC is the presence of a single well-defined segment of dilated colon with out any evidence of intrinsic obstruction or innervation defect and abrupt transition to normal caliber intestine proximally and distally. Early onset of the symptoms indicates congenital origin of the malformation.

The exact embryogenesis of SDC has not been worked out. Impairment of intestinal organogenesis, primary dysplasia, and congenital damage to the myenteric plexus, intrauterine vascular catastrophe, have all been suggested as the etiologic factors responsible for abnormal dilatation of the bowel. Arrest of cloacal septation due to nondescent of the urorectal septum along with faulty organogenesis of the distal most intestine close to descending urorectal fold would lead to ARM with abnormal dilatation. From the anatomy of this case, it can be seen that the defective segment of the intestine, which undergoes dilatation, may not always be close to the urorectal septum. We speculate that the embryological process leading to the occurrence of PCS and SDC may have some similarity. The characteristic features of SDC such as abrupt transition from normal bowel to pouch, hypomotility, presence of ganglion cells, absence of taenia and abundant serosal vascularization fed by a large marginal colonic artery have been seen in PCS as well. In our case, the dilatation involved whole of the ascending, transverse and descending colon. In two cases reported earlier, the extent of colonic involvement was proximal and extensive as in the present case, whereas in other two patients, only the distal colon was involved in dilatation. Out of four patients, three had associated anomalies.

The treatment would follow the same principles as that for the other varieties of ARM. The dilated segment has been excised; the distal colon would be pulled down through PSARP route and the ileocolostomy closed at a later stage.

**REFERENCES**


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