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Case Report

An Unusual Presentation of Congenital Megacolon with A Rare Vascular Anomaly

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ABSTRACT

Congenital megacolon also referred to as Hirschsprung's disease or aganglionic megacolon is characterized by the absence of nerve plexus in the colon, it rarely affects the small intestine. Whereas acquired megacolon is a constant dilatation of the large intestine in the absence of disease but occurs due to various factors such as infections, emotional disturbances, stress, or secondary to inflammatory bowel diseases (IBD) such as ulcerative colitis, Crohn's disease, and proctocolitis. In the present case, we report here an unusual form of megacolon. The sigmoid colon was significantly enlarged but twisting or volvulus was not detected. The ascending, transverse, and remaining part of descending colon and mesentery appeared normal, The possibility of false rotation of the gut was excluded. Interestingly, the artery supplying the left one-third of the transverse colon and descending colon originated from a branch of the middle colic artery. This anomalous artery was found to form a communication with the trunk of the inferior mesenteric artery, the Arc of Riolan, and gave branches that supplied the descending colon. The left colic artery was absent. The transverse colon showed a stricture between its right 2/3rd and left 1/3rd, confirming with vascular accidents, resulting in anomalous blood supply and aganglionosis. The dilated part of the sigmoid colon was found to be with ganglion cells, while the distal part after the megacolon, lacked ganglion cells in all layers of the sigmoid colon. This case documents a very rare finding which will make the surgeons aware of a newer arterial pattern associated with congenital megacolon.

KEYWORDS: Sigmoid colon, Hirschsprung's disease, Arc of Riolan

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INTRODUCTION

Congenital megacolon was described by Hirschsprung in 1888. A dilated colon with the absence of ganglionic cells is a classical feature of Hirschsprung's disease (HD) [1]. Due to the absence of ganglionic cells, there is

a mass contraction of the colon without propulsive activity. During the development of the fetus, the neural crest cells derived from the neural folds migrate to the gut and are responsible for the formation of the enteric nerve plexus [2]. Vascular accidents leading to interruption of blood supply to the gut in utero may lead to failure of migration of neural crest cells to the gut resulting in Aganglionic bowel, the type of megacolon depends on the ischemic type and duration. The scar tissue formed as a result of interfering with the normal blood flow to the gut narrows the lumen [3].

CASE REPORT

A 60 yr. old male cadaver, on opening the peritoneal cavity, a large sigmoid colon was noticed (Fig.1A). Greater omentum, transverse colon, and coils of intestine were hidden from view. On further examination, the caecum and transverse colon were also larger than normal size (Fig 1B, 1C). The diameter of the caecum, transverse and sigmoid colon were 10.8 cm, 7.9 cm, and 12.8 cm. A constriction was noticed at the junction of the right 2/3rd and left 1/3rd of the transverse colon (Fig.1C). Further dissection of mesentery revealed superior and inferior mesenteric artery and its branches (Fig. 2). Interestingly, the left colic artery was absent, with a branch from the middle colic artery supplying the left 1/3rd of the transverse colon. An anomalous vessel connecting the middle colic artery and the inferior mesenteric artery supplied the proximal part of the hindgut (splenic flexure & descending colon) (Fig.2). This artery is the Arc of Riolan or the meandering mesenteric artery and this is the first time in literature, where branches arise from it and supply the gut. There are two aberrations, wherein, the artery of midgut supplies the hindgut and the presence of branches from Arc of Rioloan. The

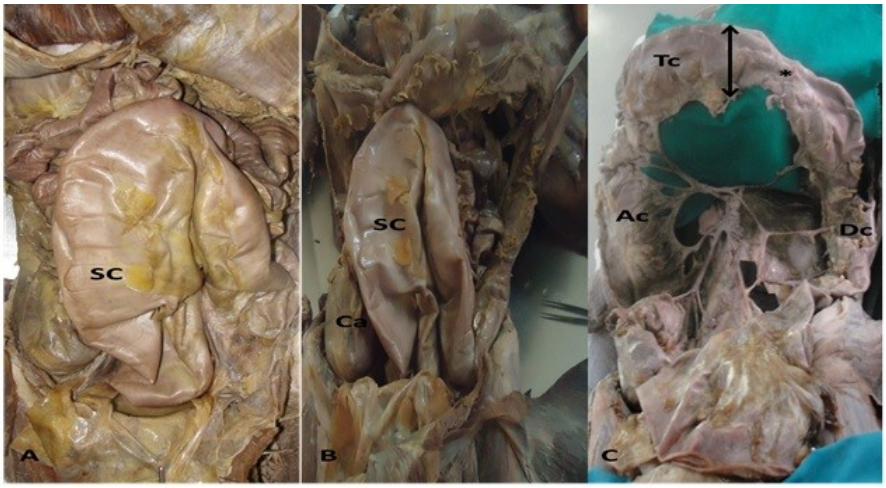

Fig. 1: A & B. Sigmoid megacolon (SC) insitu. 1B. Sigmoid megacolon and caecum (Ca). 1C. Sigmoid colon retracted to show normal ascending (Ac) and descending colon (Dc). The transverse colon is (⇕) is seen and a stricture (*) is seen at the junction of its right 2/3rd and left 1/3rd.

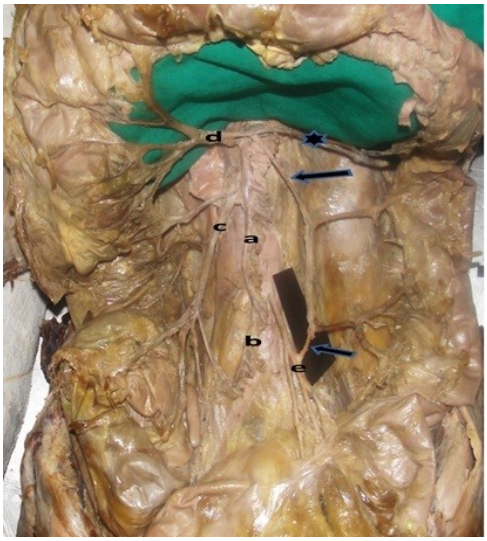

Fig. 2: Absence of left colic artery & Arc of Rolon. A branch (✱) from the middle colic artery (d) and communicating loop(arrows) between middle colic (d) and inferior mesenteric artery (e) supplies the Left 2/3rd of the transverse colon, descending colon, and upper part of the rectum. The superior mesenteric artery (a) and other branches, Ileocolic artery (b), and right colic artery (c) are also seen.

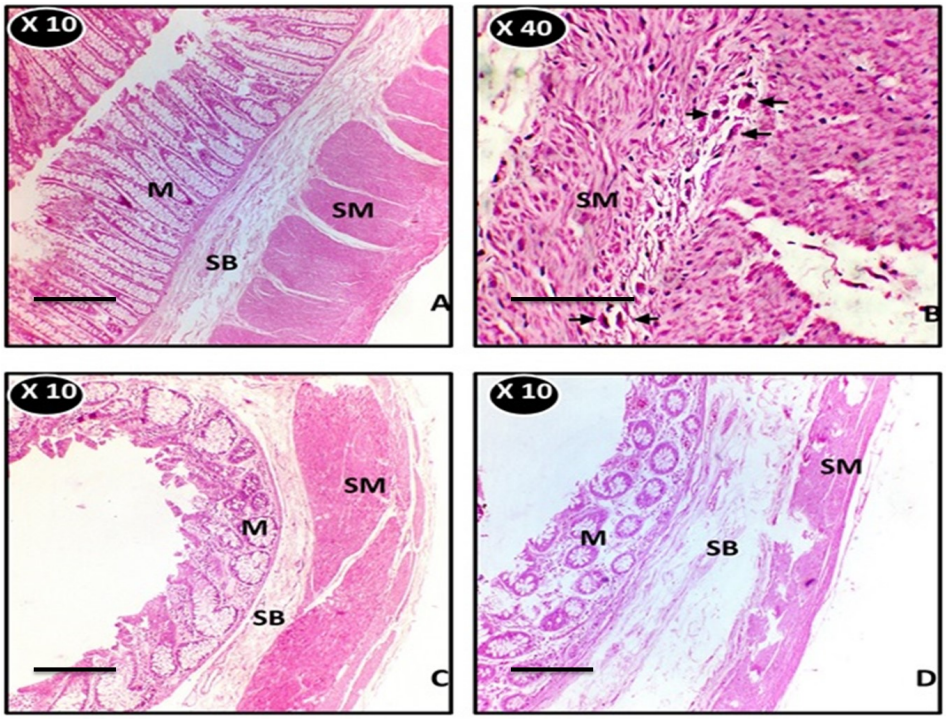

Fig. 3: A Photomicrograph of the transverse colon showing normal mucosal (M), submucosal (SB), and smooth muscle layer (SM). Fig. 3B. Presence of nerve plexus and ganglionic cells (arrows) in the smooth muscle of the transverse colon. Fig. 3C. Smooth muscle layer (SM) is reduced in size in caecum Fig. 3D. The smooth muscle layer (SM) of the sigmoid colon is greatly reduced in size. Figures A,C,D Scale bars=100 µm , Figure B Scale bars=50 µm.

Histopathological examination revealed a normal transverse colon (Fig. 3A & B). Whereas the smooth muscle layer of dilated sigmoid colon was reduced to a greater extent and ganglionic cells were absent in the distal part after the megacolon was visualized (Fig.3C&D).

DISCUSSION

The myenteric plexus is a part of the enteric nervous system. The neural crest cells migrate to the region of the gut to form ENS [4]. The ganglionic cells of the myenteric plexus of Auerbach, reach the colon in the 7th week, complete innervation by the 12th intrauterine week. The circular muscle first appears caudally at the 9th week and the longitudinal muscle layer starts cranially at the 10th week along the mesenteric border and progresses caudally, reaches caecum by the 11th week, and covers the full colon by the 16th week. The neural crest cells move to the esophagus along with the vagus nerve till the end of the gut usually. In the region of the hindgut, in addition to vagal neural crest cells (somites 2-8) the sacral neural crest cells (somites 22-28) also contribute to ENS. Only 4% of the aganglionic colon is seen proximal to the splenic flexure of the transverse colon. This can be attributed to the neural crests migrating till the right 2/3rd of the transverse colon along with the vagus nerve. The sacral nerves are seen as a poor guide to the migrating neural crest cells. Recent studies state that vagal cells initially colonize the submucosal area and migrate to the smooth muscle layer. It is exactly the opposite in the case of sacral cells. Failure of expression of various receptors such as Tyrosine kinase receptor (RET), KIT, Endothelin-3, and Endothelin B leads to aganglionic megacolon [5].

HD associated with intestinal atresia might be due to vascular accidents [6]. The role of the ischemic hypothesis cannot be excluded. This is observed in the present case with abnormal arterial supply and stricture at the junction of the midgut and hindgut. The arterial and nervous accidents or mishaps seem to have played a role in the aganglionosis. The defect in migration of neural crest cells has been accompanied by stricture and anomalous arterial supply by Arc of Riolan is very unique in this particular case.

The newer arterial pattern supplying the transverse colon and the descending colon,

with branches from Arc of Rioloan, is of utmost surgical interest. This anomaly, associated with stricture and the congenital megacolon points to vascular pathology in addition to a neural pathology. The surgeons need to be aware of such anomalous arterial supply when performing procedures of the large gut.

CONCLUSION

The exact pathway of neural crest cells to the gut is still obscure. It is assumed that the cells migrate along the blood vessels. Vascular anomalies or intrauterine vascular accidents to the gut may also contribute to Hirschsprung's disease. This case documents a very rare finding which will make the surgeons aware of a newer arterial pattern associated with congenital megacolon.

Author Contributions

Thotakura Balaji: manuscript writing and overview of the study

Jyothi Ashok Kumar: Manuscript Preparation and analysing the study findings

Vaithianathan Gnanasundaram: Collecting the literature and data interpretation

Hannah Sugirthabai Rajila Rajendran: Concept & Design of the study

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Conflicts of Interests: None

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