



Vascular malformation of ileum: A possible cause of neonatal intestinal obstruction

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ABSTRACT

Vascular lesions of gut are rare and they may rarely cause luminal obstruction in neonates. A baby boy born at 36 weeks of gestation presented with small bowel obstruction on day 3 of life. X-ray suggested small bowel obstruction. Contrast enema showed microcolon with multiple filling defects. Exploration revealed thin floppy gut with multiple segments of stenoses and dilatation with surface bluish discoloration but no perforation, two areas were resected. Proximal anastomosis was done and distally a stoma was formed. Histopathology showed vascular malformation in all the layers of gut. The diagnosis was impossible preoperatively and could be established after surgery only.

1. Introduction

Meconium disease of the newborn, Hirschsprung's disease, intestinal atresia and Necrotizing enterocolitis (NEC) are common causes of neonatal intestinal obstruction [1]. Surface hemangiomas and vascular malformations are quite common in children. However, Visceral vascular malformations are extremely rare and poorly understood [2]. Intestinal vascular malformations in association with neonatal intestinal obstruction has not been reported to the best of our knowledge. Here in line with the SCARE criteria [3] we report a neonate who had features of localized NEC and an extensive area of thin walled floppy and aperistaltic ileum, which on histology came out to be intestinal vascular malformation, leading to functional obstruction. Through this report we aim to highlight the points to be kept in mind in order to diagnose and manage such rare cases.

2. Presentation of case

A baby boy born at 36 weeks of gestation (Birth weight 2400 g), who was well at birth but did not pass meconium for 3 days, developed abdominal distension and bilious Nasogastric aspirates. There was no significant antenatal or past family/pharmacologic history associated. Plain X-Ray abdomen suggested distal small bowel obstruction which raised a suspicion of differentials related to neonatal intestinal obstruction like meconium ileus, ileal atresia or total colonic

aganglionosis. Gastrograffin enema being a diagnostic as well as therapeutic modality in such conditions was performed. It showed microcolon with multiple filling defects (Fig. 1). The dye did not reach the small bowel. He passed small amount of contrast mixed meconium immediately after the dye study, but very little subsequently. Abdominal distension increased and there were fixed loops on serial X Rays (Fig. 2). Based on extensive operative experience and the outcomes related to neonatal intestinal obstruction, exploratory laparotomy was performed on day 4 of life. It revealed multiple areas of stenoses and dilatations in the ileum, which were thin walled and floppy with bluish discoloration on the surface. No transition zone was there. Bowel rotation was normal. The surface of the involved bowel showed prominent lacteals, and no peristaltic movements were seen. This involved segment was about 40 cm long. About 25 cm proximally, there was a 5 cm segment of localized enteritis with impending perforation. Colon and rectum were small in caliber (microcolon) (Fig. 3a). The segment with impending perforation was resected and end to end anastomosis performed (Fig. 3b). The abnormal looking bluish segment was resected and a Bishop Koop (BK) anastomosis performed.

Post-operative course was smooth, Feeds could be started on day 2. Initially there was substantial output from the BK stoma, but gradually he started passing stools per rectum and stoma output was minimal.

Histopathological examination of the resected gut showed thin walled blood vessels of variable caliber within the submucosa, muscularis and the serosa. Abnormal anastomoses were also seen which

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Fig. 1. Contrast enema showing Microcolon with multiple filling defects (arrow).

confirmed the lesions as Vascular malformation (Fig. 4.). The proximal resection (5 cm) showed features of necrotizing enterocolitis (NEC).

After discharge the child was followed initially weekly and then every month at the outpatient department (OPD). The growth was monitored by anthropometric measurements and it was adequate as per the age. The stoma outputs were also monitored, it reduced gradually and most of the stools were passed per rectally. It ensured the normal functioning of the distal limb of the BK stoma. The stoma was closed at 7 months of age, with uneventful recovery.

3. Discussion

Hemangioma and vascular malformation are different entities with respect to their clinical course and the histological features. For description and diagnosis of these lesions, Mulliken's classification is followed most commonly [4]. Though the classification was developed for superficial lesions it is also applicable to visceral lesions [5]. Cutaneous and soft tissue lesions has been commonly described. Gastrointestinal vascular lesions are very rare, incidence being 0.05% [6]. Gut involvement is usually localized and segmental, mostly in the region supplied by superior mesenteric artery i.e. midgut, however, diffuse involvement is also reported [7,8]. The common clinical presentations vary from subtle signs like anemia resulting from slow intractable painless gastrointestinal bleeding to emergencies like acute intestinal obstruction, intussusception or perforation [7–9]. Our case had no blood in the stools and there was no luminal obstruction on account of the malformation. However, there was functional obstruction because of poor muscular action as the malformation was transmural. This explains the floppy appearance, lack of peristalsis, and fixed loop on the plain X ray. The impending perforation and NEC in the more proximal segment could be secondary to functional distal obstruction or maybe it was incidental - it happened 25 cm proximal to the vascular malformation.

Diagnosis of gut vascular malformations is difficult even with advanced imaging techniques such as Doppler Ultrasound, MRI, CT and endoscopy [2,10,11]. Chronic lower gastrointestinal bleeding may invite specific investigations to pick up a vascular cause, but neonatal intestinal obstruction invites differential diagnosis such as atresia, meconium disease of newborn, NEC and Hirschsprung's disease. Usually plain abdominal film, and at times, contrast enema is enough to make a decision for surgery. Our case probably had a long standing (probably from antenatal period) functional obstruction because of the muscle layer involvement. The proximal NEC could be related to this obstruction or be an unrelated pathology.

4. Conclusion

Visceral vascular anomalies are rare and seldom diagnosed preoperatively. Infantile cutaneous hemangiomas are quite common but isolated vascular malformations involving gut has been seldom reported, our case being the rarest case to the best of our knowledge.

Provenance and peer review

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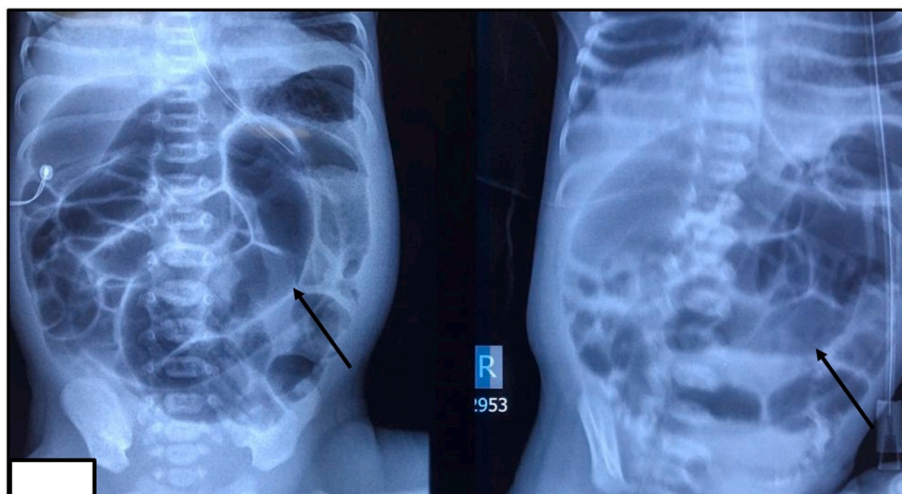


Fig. 2. Serial Plain X rays showing fixed loop.

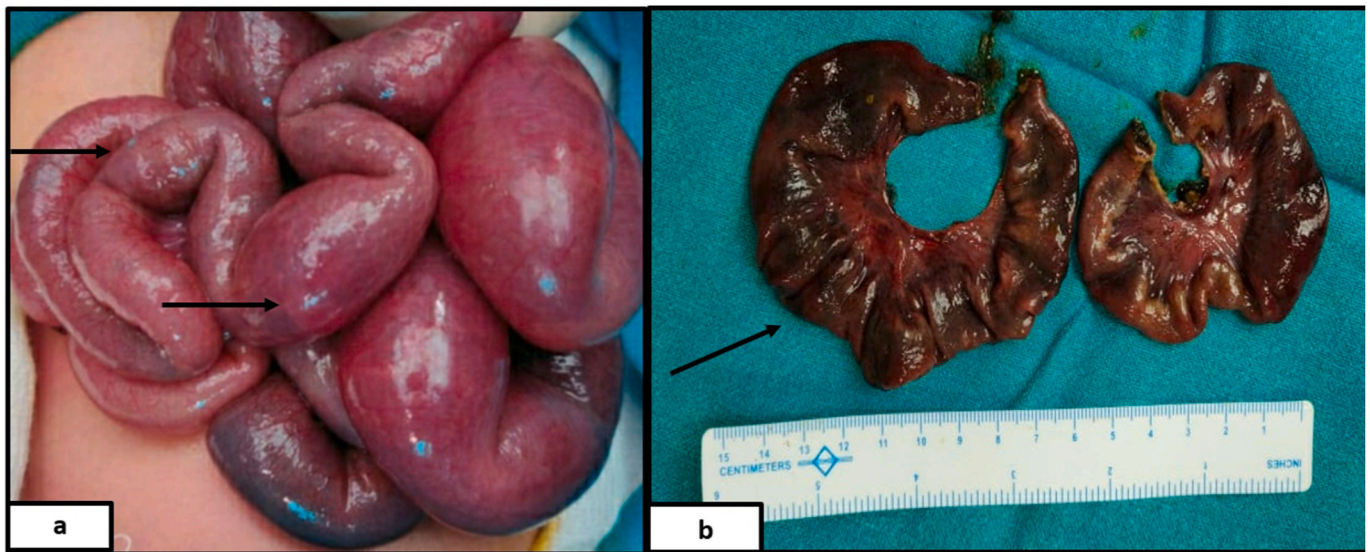


Fig. 3. (a) Multiple bluish thin walled, dilated and stenosed segments with surface lacteals.
Fig. 3(b) Resected abnormal segment of ileum.

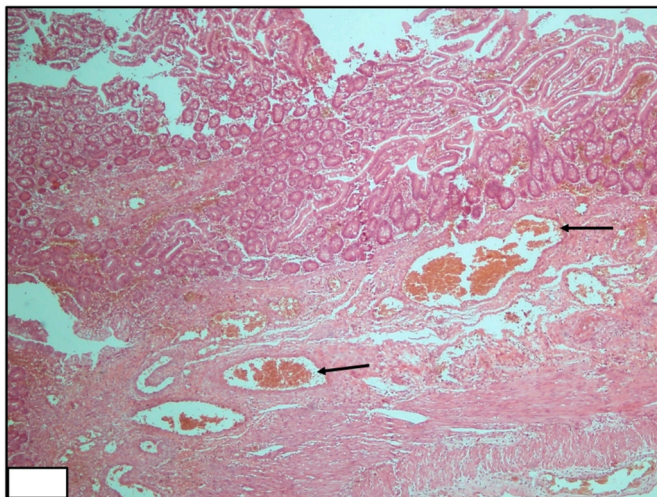


Fig. 4. Photomicrograph of Histology (H and E Stain, 40X) showing abnormal blood vessels and anatomoses of variable calibers in all layers of gut.

Patient consent

Written informed consent was obtained from the parents of the baby for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Ethical approval

The study is exempt from ethical approval in my institution.

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Author contribution

Dr Satish Kumar Aggarwal: writing the manuscript, study design, final review.

Dr Gaurav Singh and Dr Rupa Banerjee: data collector, writing the manuscript.

Research Registration number

In accordance with the Declaration of Helsinki 2013, all research involving human participants has to be registered in a publicly accessible database. Please enter the name of the registry and the unique identifying number (UIN) of your study.

You can register any type of research at <http://www.researchregistry.com> to obtain your UIN if you have not already registered. This is mandatory for human studies only. Trials and certain observational research can also be registered elsewhere such as: ClinicalTrials.gov or ISRCTN or numerous other registries.

Guarantor

Dr. Satish Kumar Aggarwal.

Declaration of competing interest

The authors declare conflicts of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2020.11.048>.

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