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Neonatal sigmoid volvulus

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ABSTRACT

Background: Neonatal sigmoid volvulus is a rare entity. It is associated with Hirschsprung's disease. Presentation is acute abdominal distention, vomiting and obstipation. Abdominal radiograph will show the "coffee bean" sign, but this is frequently missed and the diagnosis requires a high index of suspicion. Treatment options include contrast enema, colonoscopy or laparotomy, depending on the condition of the baby and local availability.
Population and results: During the last 6 years, 6 infants with sigmoid volvulus were treated in our department. Four presented during the first 48 h since birth, and 2 presented at the age of 2 and 7 weeks of age. One child was operated and 5 had primary contrast enema with radiologic de-volvulus. Rectal biopsy was performed in all cases; three children had Hirschsprung's disease. Those with normal biopsies responded well to rectal washouts. Two patients had early one stage transanal pullthrough and one had 2 further occasions of sigmoid volvulus prior to definitive surgery. All three recovered with an uneventful course.
Conclusions: Neonatal sigmoid volvulus requires a high level of suspicion. Contrast enema is efficient for primary de-volvulus. Rectal biopsy should be performed and if positive for Hirschsprung's disease, surgery should be performed sooner rather than later.

Sigmoid Volvulus (SV) is a life-threatening condition. Although fairly frequent and well described in the adult population, in neonates this is a very rare and frequently missed condition. Only sporadic case reports are found in the English literature [1, Table 1]. The volvulus is associated with the presence of a redundant sigmoid colon and a narrow sigmoid mesentery [2,3]. The presentation of SV is often abrupt with abdominal distention, acute obstipation and an empty rectal ampulla. The diagnosis requires a high level of suspicion and the treatment should not be delayed, since procrastination may lead to bowel necrosis and serious sequelae [2]. The mainstay of treatment is de-volvulus, achieved by contrast enema or surgery In neonates, the option of colonoscopic de-volvulus is not practical. Among the reasons for missed or delayed diagnosis in neonates are that the physical examination and radiologic signs may be misleading. Occasionally, it can be associated with Hirschsprung's Disease (HD) [3–6]. The treatment options vary according to the patient's condition and the available expertise in treating neonates.

During the last 6 years we treated 6 infants with SV, 5 of them neonates (less than 30 days of age) and one at the age of 7 weeks, and we have learned important lessons from the retrospective study of these cases.

1. Methods

All infants treated for SV between 2010 and 2015 were studied. Two of the cases presented at the Emergency Room, one at the age of 7 weeks of age and the second at the age of 16 days. The other 4 were referred from the neonatal ward at the average age of 2.6 days of age. All patients were males, born at term, with no known diseases or family history of diabetes (including gestational diabetes) or gastrointestinal problems. All infants passed meconium during the first 24 h of life.

Abdominal radiograph was obtained in all cases. One patient, the 7 week old baby, had abdominal tenderness and dehydration at presentation and was taken promptly to surgery. The operative findings were sigmoid volvulus with viable bowel and a manual de-volvulus was performed. Rectal biopsy performed 1 week following surgery was normal.

The other five infants had a contrast enema as a diagnostic procedure and as the primary course of treatment. Rectal biopsy was performed in all patients following symptom resolution.

2. Results

The presentation in all cases was of abrupt discomfort with abdominal distention and bilious vomiting. A "coffee bean" sign was present in abdominal radiographs (Fig. 1) in all patients. Contrast enema was performed in 5 cases and demonstrated SV in all (Fig.2). De-volvulus was achieved in all infants, either surgically (in one case)

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Table 1
Previous reports in English literature.

Study	Age/Sex	Symptoms	Plain film	Diagnosis	Treatment	Outcome
Carter & Hishaw[8]1961	14 d/M	Abdominal distension, bile-stained vomiting	Severe colonic distension	Ba enema, Exploratory laparotomy	None, spontaneous detorsion	Postoperative peritonitis, death after2 days
Srouji et al.[9]1974	2 d/M	Abdominal distension, bile-stained vomiting No meconium passage	Moderate distension, Air fluid levels, No air in rectum	Ba enema, Exploratory laparotomy	Sigmoid resection, Colostomy, Closure of colostomy at 3 months	Enterocolic Fistula, TPN sepsis, Death at 8 months
Valla et al.[10] 1982	5 d/M	Acute distal bowel obstruction	Air-fluid level	Laparotomy	Sigmoid resection	Hirschsprung's disease, Well
	6 d/M	Acute distal bowel obstruction	Air-fluid level	Ba enema	Laparotomy, Detorsion	Persistent constipation, Encopresis
Janik et al.[11]1983	1 d/M	Normal gas pattern, No meconium passage, Imperforate anus	Increasing distension, No air in rectum, Pneumoperitoneum	Exploratory laparotomy	Loop colostomy	Abdominoperineal pull-through
McCalla et al.[12]1985	2 d/M	Abdominal distension, Vomiting, No meconium passage	Dilated small and large bowel	Exploratory laparotomy	Detorsion, Sigmoid colostomy at 14 days	Hirschsprung's disease, Soave procedure at 11 months, Well
Venugopal et al.[13]1997	1 d/M	Abdominal distension, Non-bilious vomiting	Dilated bowel loops	Ba enema	Detorsion	Hirschsprung's disease, Soave procedure as neonate months, Well
De Caluwé et al.[1]2001	2 d/M	Abdominal distension, Non-bilious vomiting, Fight anus	Dilated bowel loops	Omnipaque enema	Detorsion	Anal dilatation at 4 day, Well
Pastor et al.[6]2013	Preterm/male	Poor feeding, constipation, abdominal distension, vomiting	Distended bowel loops, Air-fluid level	Laparotomy	Detorsion	Doing well
Arbell et al.[14]2010	49 d/M	Abdominal distension and vomiting	Distended bowel loops, Air-fluid level	Laparotomy	Detorsion	Doing well
Current report (6 cases)	53 d/M, 1.25 d/M, 1.25 d/M, 2 d/M, 21 d/M	all have Abdominal distension and vomiting	all have coffee bean sign	abdominal X ray	detorsion	Doing well

or radiologically (in all other five infants). Three of the patients had evidence of HD on biopsy and 3 had normal biopsies. During the same period of time, 98 children were treated for HD in our department (making the incidence of SV as the presenting symptom for HD (3%)). The results are summarized in Table 2.

Those with normal biopsies were treated with rectal washouts for one month and did not require further treatment. Two patients with HD had early surgery (transanal pull-through), at 35 and 45 days of age. In the third patient, diagnosis of HD was delayed because of technical problems with the biopsy, and during the period between presentation and diagnosis he had 2 further episodes of SV; he was operated at the age of 18 weeks. The average length of HD segment was 20 cm from the dentate line, approximately at the mid sigmoid colon with resection of 24–27 cm of bowel. All three operated infants had an uneventful recovery. The patients are still under routine follow-up, for a period of 1–6 years. All children are well and thriving.

3. Discussion

Sigmoid volvulus (SV) is a life threatening emergency needing immediate care. It is a well-known entity in the elderly population, where colonoscopy or hydrostatic de-volvulus has become standard practice, leaving surgical treatment as an option in cases of questionable bowel viability [7]. Sigmoid volvulus is usually attributed to a redundant (dolico) sigmoid colon with a narrow mesentery. It is uncommon in children, and exceedingly rare in neonates or very small infants [4] Since the first case of neonatal SV reported by Carter in 1961 [8], we found only 8 cases in the English literature (Table 1) [4,6,9–14]. Therapeutic suggestions vary and included laparotomy with detorsion with or without a colostomy or hydrostatic reduction utilizing contrast enema. Colonoscopy is not mentioned, and is not relevant to the neonates, especially in an emergency setting. The association with Hirschsprung's disease is mentioned in all manuscripts, although, as can be seen in the table, only 3 out of 8 cases had a proven HD. Although previous reports conclude that the incidence of SV because of HD is about 0.6% of the HD population [5], in our series we had 3 out of 98 HD patients presenting with SV (3%). As all series, including ours, are small, this may be without significance. It is mentioned that this condition is easily missed in children, either because of a subtle presentation or because it is not expected at this age group [15]. In this series, all children had a clinical and radiological presentation that is similar to SV in adults, i.e., abrupt abdominal distention and a "coffee bean" sign on plain films. Therefore, in a previously healthy neonate or infant with abrupt-onset abdominal distension, SV should be suspected and radiological signs should be sought for on the abdominal radiograph.

Our series is the largest one in a single center, with 5 neonates (2–21 days old) and one 7 week old infant. In this series, only one child was operated, because of abdominal tenderness at presentation. Five of the six neonates responded well to contrast enema. Enema performed under fluoroscopy with a water-soluble contrast agent confirmed the clinical diagnosis and easily achieved de-volvulus. Although previous reports found only 77% success in enema reduction in children [4], we had a 100% success rate in all our attempts. This may be because of sheer luck or perhaps because of the fact that our