Case Report

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Two simultaneous intussusceptions in a neonate with DiGeorge syndrome: A case report

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KEYWORDS

Neonate,
Intussusception,
Multiple intussusceptions,
Hypoxia

ABSTRACT

Background: Double simultaneous intussusception is a peculiar and rare variety of intussusception with only 3 previously reported neonatal cases.

Case presentation: A 15-day-old male neonate with respiratory distress was found to have Tetralogy of Fallot and hypoplastic pulmonary stenosis. Small bowel intussusception was diagnosed on ultrasound abdomen following hematochezia on the next day. Emergency laparotomy revealed two intussusceptions, ileocolic and jejunojejunal, with bowel gangrenous requiring resection and anastomosis. No pathological lead point was identified. He recovered with supportive care and was discharged.

Conclusion: Simultaneously occurring double intussusceptions are extremely rare in neonates, and thorough examination of the entire small bowel in cases of intussusception is key to the diagnosis.

INTRODUCTION

Intussusception is due to the invagination of one part of the bowel into the adjacent intestine. It can occur from the intrauterine period to adulthood but is usually a common cause of intestinal obstruction in children between 6 to 18 months of age. Neonatal intussusception accounts for only 0.3 to 1.3% of intussusceptions.[1] Double simultaneous intussusception is a rare variety in which there are two intussusceptions at different locations and very few cases of this entity are reported. We report one such case with associated hypoxia secondary to the cardiac anomaly. The hypoxic injury also has been proposed to be one of the aggravating factors for late-onset neonatal intussusception.[2]

CASE REPORT

A 15-day-old term male neonate, delivered by Caesarean section with a birth weight of 2.75 kg and normal antenatal history, was brought with respiratory distress; developed cardiac arrest at arrival thus received cardiopulmonary resuscitation twice. He was diagnosed to have Tetralogy of Fallot and hypoplastic central pulmonary arteries with collaterals. Echocardiography (ECHO) showed large subaortic ventricular septal defect (VSD) with a bidirectional shunt, patent foramen ovale (PFO), small atrial septal defect (ASD) with a bidirectional shunt, small restrictive patent ductus arteriosus with a left to right shunt, 50 % overriding of the aorta, small right ventricle, hypoplastic pulmonary artery, and right arch. Left ventricular function and dimensions were normal. His oxygen saturations were maintained between 75 to 80% with optimal ventilator support. There was a requirement of inotropes to support the circulation.

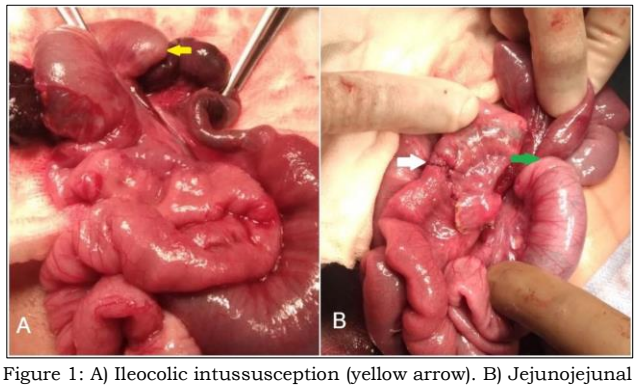

Figure 1: A) Ileocolic intussusception (yellow arrow). B) Jejunojejunal intussusception (green arrow) with distal anastomosis (white arrow).

Surgical consultation was sought on the next day for abdominal distension, bilious aspirate, and an episode of bleeding per rectum. The radiograph of the

abdomen showed dilated bowel loops. Ultrasonography (USG) of the abdomen showed features of small bowel intussusception. Emergency laparotomy revealed two intussusceptions, an ileocolic and jejunojejunal types (Fig.1). Both intussusceptions required resections of gangrenous intussusceptum with end-to-end anastomosis (Fig.2).

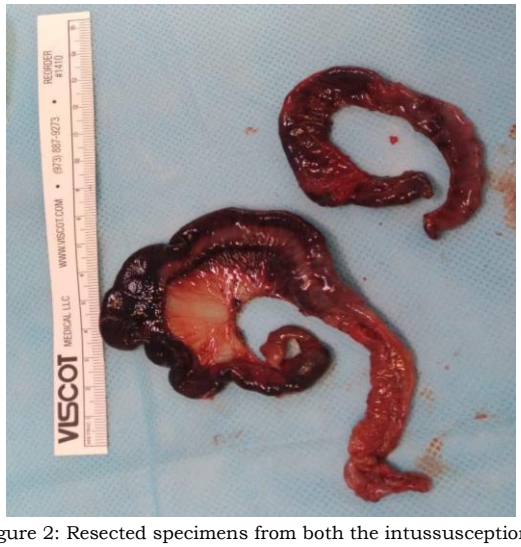

Figure 2: Resected specimens from both the intussusceptions with no pathological lead points grossly

There were no pathological lead points. Histopathology of excised segments showed full-thickness hemorrhagic necrosis only. Postoperatively, feeds were started on the fifth day and progressed gradually. He recovered well and was discharged. Subsequent evaluation for syndromic associations was done due to dysmorphic facies, post-axial polydactyly, cardiac anomaly, thymic hypoplasia, and hypocalcemia. Fluorescence in situ hybridization (FISH) analysis showed microdeletion at 22q11, diagnosing Di-George syndrome.

DISCUSSION

Diagnosis of intussusception in neonates (especially in preterm) is usually delayed as the symptoms mimic necrotizing enterocolitis (NEC) or other causes of intestinal obstruction.[3] USG abdomen is useful to diagnose intussusception. In our case also, there was a delay in diagnosis of one day, as symptoms were attributed to the cardiac condition. In view of this delay, most neonates have gangrenous bowel by the time of surgery, as happened in the index case.

Intussusception in neonates can be idiopathic or associated with a pathological lead point such as inspissated meconium, duplication cyst, Meckel's diverticulum, hamartoma, polypoid masses, etc.[4,5] In premature neonates, small bowel intussusception is the commonest type for which intestinal hypoperfusion, hypoxia, and dysmotility may be the contributory factors. Lead point is seen in only 5-8% of neonates.[1] Our case had no pathological lead point.

Simultaneously occurring double intussusception is a peculiar variety characterized by two separate segments of intussusceptions with an intervening normal segment of the bowel. The term is used interchangeably with double intussusception, which refers to an intussusception within intussusception.[6] Our patient had two simultaneously occurring intussusceptions. Only 3 previous cases of this entity have been reported in the neonatal age group in English literature.[6-8] These are summarized in Table 1.

Table 1: Summary of cases of simultaneous Intussusception in neonates reported in English literature.

Sr.no	Age at presentation	Number	Locations	Lead-point	Associated anomalies	Outcomes
1[8]	3 days	2	Jejuno-jejunal and ileo-colic	Idiopathic	Transposition of great arteries	Died postoperatively due to cardiac complication
2[7]	21 days (preterm)	6	Ileo-ileal	Meconium plug (one site). Rest-Idiopathic	Patent ductus arteriosus, Respiratory distress syndrome	Died on postoperative day 10 due to sepsis
3[6]	10 days	2	Ileo-ileal and Ileo-colic	Idiopathic	Nil	Died on postoperative day 5 due to sepsis
4	15 days (present case)	2	Jejuno-jejunal and Ileo-colic	Idiopathic	DiGeorge syndrome, Tetralogy of Fallot	Survived and discharged one month postoperatively

Cardiac anomalies were associated with this entity in two of the reported cases.[7,8] Our patient had complex cardiac anomalies as part of DiGeorge syndrome. Cardiac anomaly is probably not the causative factor, but the associated hypoxia may aggravate the pathology. Our child had persistent hypoxia (oxygen saturations of 75-80%) due to Tetralogy of Fallot with hypoplastic pulmonary stenosis which may have contributed to the gangrene of the intussusceptum. Furthermore, in most instances, the cardiac condition worsens with altered hemodynamics (as in our baby) and may distract treating doctors from the abdominal pathology contributing to delay in surgery.[8]

Preoperative detection of simultaneous intussusceptions has not been described in neonates though it is described in children.[9] In our patient, only one intussusception was reported on ultrasound. This may not affect the decision for operative management. However, diagnosing and treating the second intussusception during the first surgery is important to minimize the morbidity to the baby and hasten recov-

ery. Clues at the time of laparotomy for a second intussusception include:

1. The presence of altered blood in lumen proximal to the intussusception (the first delivered intussusception is the distal one in most instances)
2. Dilated bowel distal to intussusception (if the proximal one delivered first).

The outcome in these babies usually depends on the associated conditions though increased morbidity can be anticipated due to delayed diagnosis and bowel gangrene. The previously reported cases have all died in the postoperative period due to associated cardiac anomalies or sepsis; our neonate is the only reported survivor.

To conclude, intussusception is an uncommon cause of intestinal obstruction in neonates. Simultaneous double intussusception in neonates is a rare entity within this group. A high mortality rate is attributed to underlying cardiac anomalies as well as delays in diagnosis and treatment. Examination of the entire small bowel at the surgery in cases of intussusception is key to the diagnosis. Prompt surgical care can improve the outlook in these babies.

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