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Neonatal Isolated Focal Intestinal Perforation in a Preterm with Hypoganglionosis

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Abstract

An interesting rare case of focal intestinal perforation in a preterm baby with hypoganglionosis which was managed conservatively by an innovative abdominal paracentesis under ultrasound guidance and endosurgical transanal procedures respectively successfully has been reported. The neonatal rectal suction biopsy showed few ganglion cells suggestive of hypoganglionosis. Baby was treated conservatively but in infancy presented with functional distal intestinal obstruction and limited lower gastrointestinal contrast study showed a transitional zone in the rectum for which the patient underwent transanal endosurgical procedures uneventfully. The patient is asymptomatic, thriving well with excellent anatomical, cosmetic and functional results at long term follow up at 6 years.

Keywords: Anorectal myomectomy; Bowel perforation; Congenital motility disorder endoscopic; Focal; Hypoganglionosis; Ileal; Infant; Intestinal; Isolated; Neonate; Pneumoperitoneum; Preterm; Rectal; Transanal; Variant Hirschsprung's disease

Introduction

Neonatal isolated focal intestinal perforation (FIP) is a very rare but life-threatening potentially lethal condition that occurs when a newborn baby's bowel wall perforates [1]. It is a rare complication that mainly affects preterm babies with low birth weights and involve terminal ileum, appendix and cecum in most instances [2]. Previously it has been labelled as spontaneous intestinal perforation (SIP) which we now avoid as there is usually an underlying functional or organic cause for it [3]. We wish to report a neonate who initially presented with FIP and treated conservatively. Subsequently, patient needed intervention for the underlying partial functional distal intestinal obstruction due to hypoganglionosis and treated well with endosurgical transanal procedures successfully with excellent results.

Case Report

A-preterm (36+3/40), low birth weight (2050 grams) female baby, born to a 2nd gravida mother having normal prenatal scans after an uneventful pregnancy and normal spontaneous vaginal delivery. Patient has passed meconium soon after birth and established breast feeding very well. Patient developed sudden severe abdominal pain with persistent crying, abdominal distention, constipation and vomiting and refusal to feed on day 10 of life. The referring hospital did abdominal radiographs and referred to us after initial resuscitation with nasogastric decompression, intravenous fluids and antibiotics (Figure 1). On examination, baby was settled and vital signs were stable, Abdominal was distended tender with no discoloration. The anus site and size were normal. We could pass rectal catheter for washouts without any grip around it and explosive gas and there was some changing stool staining on the catheter. The laboratory tests showed high white cell and neutrophil counts with CRP of 80 mg/dl. Abdominal ultrasound guided paracentesis using size 16 intravenous cannula in left lateral decubitus position on the right flank vented large amounts of free gas. The abdomen went scaphoid and the respiratory compromise went away immediately. Baby's general condition improved, bowel sounds returned and

passed stools after which slow feedings were started which were tolerated well.

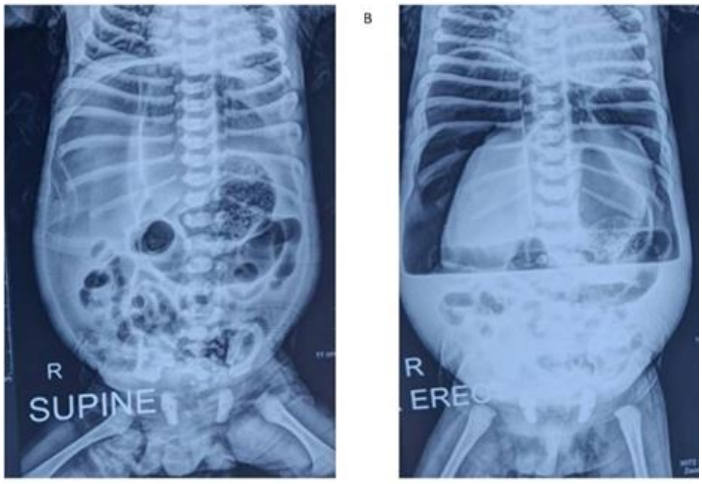

Figure 1: *Abdominal radiographs at presentation with massive pneumoperitoneum with football sign. A. Supine with dilated bowel loops in the background. B. Erect with air fluid levels.*

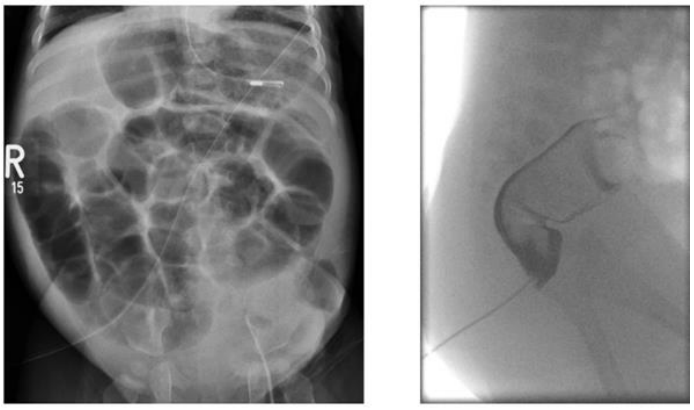

Figure 2: *Radiographs at subsequent presentation. A. Dilated bowel loops with gasless rectum B. Limited contrast enema with transition zone at rectum.*

A rectal suction biopsy showed few ganglion cells without any hypertrophied nerve fiber bundles and acetyl cholinesterase enzyme staining was normal. The neonatal conservative management was instituted for hypoganglionosis with micronutrient supplements, holobiotics, laxative and alternate day or twice a week warm salted water enemas 10ml per kg if baby does not pass stools spontaneously. The parents and grant parents were trained and baby discharged home. After couple of months parents stopped conservative treatment and baby gradually developed chronic constipation especially when weaning foods started. Infant presented to us with distal intestinal obstruction. Abdominal radiograph showed dilated bowel loops in the abdomen with no gas in the rectum (Figure 2A). A limited lower gastrointestinal water-soluble contrast study showed a tradition zone at lower rectum (Figure 2B). Parents wanted a permanent solution as they were unable to cope with the conservative management. Baby underwent transanal endosurgical procedures consisting of examination under anesthesia, anorectal examination, proctoscopy, on table large bowel preparation using normal saline with hydrodistension of left colon, transanal extended anorectal and circular muscles myectomy with full thickness biopsy uneventfully. The postoperative period was uneventful. Full thickness rectal wall biopsy showed few ganglion calls and no hypertrophied nerves suggestive of hypoganglionosis. Patient remains under annual follow up and at 6 years long term follows up; patient is asymptomatic, well, having on no medications or scars. with excellent anatomic, functional and cosmetic results.

Discussion

FIP is different from necrotizing enterocolitis (NEC), another gastrointestinal complication of prematurity. Both conditions are postnatal, has prematurity and low birth weight as risk factors, can present in the same way but their subsequent management and underlying conditions including outcomes are different. Our patient had colostrum and regular breast feeding from birth and therefore has done well despite a potentially fatal condition with mild prematurity and relatively higher weight. These benefits of colostrum and continuous exclusive breast feeding are well known for better gut health and body immunity [4]. Our team has recently published the effects of prematurity, low birth weight, partial functional hind gut obstructions and their sequalae in causing back pressure leading initially to colonic blind loop obstruction. With ileocecal reflux and backwash ileitis and inflammation of payer's patches, perforation of terminal ileum can explain the etiopathogenesis of the FIP [5]. The watershed areas of vascular supply in the ileocecal region may explain its location in terminal ileum at the sites of these patches [6]. Neonates have amazing healing power with serosal patch from parietal peritoneum following aspiration of air and fluid by cannula or a drain.

A neonatal intestinal perforation is a major life-threatening condition with high morbidity and mortality despite improvements in surgical and medical treatments that requires emergency expert management. We have very dedicated neonatology team and an enthusiastic neonatal and pediatric interventional radiologist with pediatric surgical background which helps us taking innovative strategies for management as most neonatal surgeons would have done laparotomy at the initial presentation. Based on our extensive fetal surgery and functional partial distal intestinal obstruction for decades allows us to discover and implement innovative approaches to the traditional management of these disorders using natural orifice transanal corrections in one stage with very satisfying results to patient, parents and professions alike [7]. The enterocolitis remains a risk

in severe degree of hypoganglionosis in young infants especially when embarking on conservative treatment and appropriate safety netting measures to reduce this serious and potentially lethal complication remains the mainstay for neonates and infants [8]. We prefer conservative non-interventional management of most congenital colorectal motility disorders but those with severe symptoms and presentation in neonatal period with surgical complications are best managed with advanced endosurgical procedures especially for patients in developing countries with limited resources and rural background with poor transportation systems.

Conclusion

FIP and hypoganglionosis association is very rare. Conservative management of DIF is possible, safe, effective but needs to be individualized in selected patients. Hypoganglionosis may be treated conservatively and when it fails, endosurgical transanal procedures are very safe, effective and successful with excellent long-term prognosis.

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