Hirschsprung’s Disease Masquerading as Obstructing Congenital Band in a Premature Neonate: A Double Pathology

Prematüre Yenidoğanda Konjenital Bandın Tıkanması Olarak Maskelenen Hirschsprung Hastalığı: Çift Patoloji

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ABSTRACT

Chronic feeding intolerance in preterm infants is often associated with necrotizing enterocolitis rather than Hirschsprung’s disease (HD) or intestinal obstruction due to congenital band. We report a case of a 26-day-old male born premature at 30 weeks of gestation presenting with recurrent episodes of desaturation and feeding intolerance which were initially thought to be due to NEC. However, the pathology was found to be a congenital band that caused partial intestinal obstruction and suction rectal biopsy subsequently showed it had characteristic features of HD. This dual pathology is a rare condition that required a high index of suspicion to be able to get a correct diagnosis especially in a premature baby with non-resolving chronic feeding intolerance or chronic partial obstruction.

Key Words: Premature Neonate, Feeding intolerance, necrotizing enterocolitis, Hirschsprung

INTRODUCTION

Neonatal Hirschsprung’s disease (HD) is not an uncommon disease. The incidence is 1 in 5000 new live birth. Prematurity is reported in as many as 10% of those children with HD(1). Pathophysiological basis of the disease is failure of neural crest cells to reach the distal bowel in utero resulting in an aganglionic segment of the colon. These abnormal innervations of the bowel lead to aperistaltic segment of bowel causing functional obstruction. We report a case of a 26-day-old male born premature at 30 weeks of gestation which was initially thought to be due to necrotizing enterocolitis (NEC), but later confirmed histologically to have HD.

CASE REPORT

A 26-day-old girl, delivered at 30 week of gestation, developed recurrent episode of desaturation, abdominal distension, and feeding intolerance for 2 days duration followed by no bowel movement for 3 days. Plain radiograph of abdomen showed dilated bowel, without pneumatosis, or pneumoperitoneum. The baby was treated by paediatric team as NEC grade 2 with antibiotic and the kept nil by mouth for 5 days. Daily monitoring of the baby’s abdominal girth showed persistent abdominal distension.
On further history, it was found that she was born vigorous with good APGAR score. Birth weight was 940 gm. Feeding was started at day 2 of life, found to be well tolerated and gradually stepped up to full feeding. There was normal meconium passage within the first 24 hours. There was no history of disease, drug taking or polyhydramnios in the mother. She had no physical or congenital abnormality and there was no family history of congenital defects. Initial physical examination revealed a dehydrated and crying baby with tachycardia and tachypnoea. She was afebrile. The abdomen was grossly distended, tympanic, with superficially dilated veins, however there was no evidence of peritonitis. There was neither external hernia nor peripheral oedema. Perineal examination revealed a normal anal canal, rectum, vagina and perineum.

She was resuscitated and improved with conservative management by means of nasogastric decompression, intravenous fluids, analgesia and antibiotics and urine output monitored via a urethral catheter. A full blood count was normal but urea and electrolytes showed evidence of dehydration. An erect chest radiograph was normal but a supine abdominal radiograph showed distended small and large bowel with air in the rectum.

Lower contrast study reported a smooth flow of contrast from rectum into sigmoid, descending and distal transverse colon. There was abnormal position of ascending colon and caecum. Caecum position is at mid upper abdomen. Both ascending and caecum appear dilated compared to the distal bowel. Suspicious transitional zone was seen that may suggest HD (Figure 1).

**DISCUSSION**

In our case, the diagnosis of HD was delayed as the presenting symptoms of feeding intolerance and chronic bowel obstruction were attributed to a concurrent pathology of partial obstruction by a congenital band. Interestingly, she developed again episode of abdominal distension and feeding intolerance post release of the congenital band. Abdominal distension and feeding intolerance is a common problem in neonates. Usually, the cause is NEC rather than HD. Commonly, children with HD will present with failure to pass meconium within the first 48 hours of life, but this was not seen in our case. Delayed passage of meconium was reported to be found in 17% of premature infants with HD(1). However, early passage of meconium does not rule out the possibility of HD(1).

The prevalence of HD was found to be significantly less common in premature infants (5%) when compared to term infants (50%) with obstructive symptoms (1). Although HD occurs in premature infants, HD as the aetiology of obstructive symptoms is far less common that in term infants(1). A previous report explained that history, physical examination, and contrast enema should be used to reliably identify patients unlikely to have HD and spare them from a suction rectal biopsy(2). There is no exact diagnostic algorithm that has been proposed for the diagnosis of HD for numerous reasons. Although lower contrast enema study was done earlier in this case, it failed to detect any features of HD. According to previous literature report, the sensitivity of contrast enema ranged between 65% and 80% and the specificity to ranged between 66% and 100%(3).

In our case, an anomalous congenital band and small bowel herniating through the band was initially found to be the cause of intestinal obstruction. The aetiology of the band was unclear as it could be congenital or post NEC causing partial small bowel obstruction. However, the presence of an anomalous congenital band is very rare occurrence(4). Due to the extremely low prevalence of HD in premature infants, suction rectal biopsy should be used selectively in this group of patients only after additional workup has failed to yield a diagnosis as was done in our case.

**CONCLUSION**

This case emphasizes the importance of having high index of suspicion in managing a premature infant with unresoved NEC and chronic small bowel dilatation as it can also be a secondary to coexisting HD as a second pathology.

**Conflict of interest**

No conflict of interest was declared by the authors.

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