

Case Report

Jejunal atresia presenting as meconium cyst with polysplenia in a newborn: an extremely rare association

Rashi¹, Kalyani Saha Basu^{2*}, Samiran Biswas³, Shibsankar Barman⁴

Department of Paediatrics Surgery, ¹All India Institute of Medical Sciences, Patna, Bihar, ²Nil Ratan Sircar Medical College, Kolkata, ³B C Roy Post Graduate Institute of Paediatric Sciences, Kolkata, ⁴Institute of Post Graduate Medical Education & Research, Kolkata, India

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*Correspondence:

Dr. Kalyani Saha Basu,

E-mail: drksahabasu@gmail.com

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ABSTRACT

Jejunal atresia (JA) presenting as meconium cyst with polysplenia is an extremely rare condition. We reported a case presenting with abdominal distension and feculent vomiting. Exploration revealed a type IV JA with dilated proximal segment presented as a large cyst filled with air and meconium. Polysplenia diagnosed incidentally.

Keywords: Jejunal atresia, Polysplenia, Meconium cyst

INTRODUCTION

The embryogenesis of jejunoileal atresia (JIA) is thought to be due to intrauterine ischemic insult to the midgut affecting intestinal segments after their development.¹ Polysplenia refers to presence of two or more spleens. When associated with various organ abnormalities in the abdomen and chest, it is referred as Polysplenia syndrome. It was first described by Helwig in 1929.² The incidence of both anomalies occurring simultaneously is unknown and has only been reported in scattered cases.

CASE REPORT

A day 5 full term newborn male baby weighing 2000 gm at birth was referred to our emergency department from outside. Patient presented with abdominal distension (Figure 1) followed by repeated episodes of bilious vomiting in last 48 hours. He did not pass meconium since birth. He was first baby born by caesarean section. On examination, the neonate had normal and stable vital signs. Per abdomen examination revealed huge distension and per rectal showed no meconium but mucus present

on passing a red rubber tube. Straight X-ray abdomen showed a huge translucent shadow occupying central abdomen and a few scattered air fluid levels (Figure 2). Hemoglobin was 11.1 gm%, other biochemical profile were within limit.



Figure 1: Huge abdominal distension.



Figure 2: Straight X-ray abdomen showed a huge translucent shadow occupying central abdomen and a few scattered air fluid levels.



Figure 3: Peritoneal cavity, a cyst filled with air and meconium measuring about 20 cm × 20 cm found.

After initial resuscitation the patient was operated after about 12 hours of admission. Under general anesthesia, the right supraumbilical transverse incision was made. On exploration of peritoneal cavity, a cyst filled with air & meconium measuring about 20 cm×20 cm was found (Figure 3). Careful separation of cyst wall from underlying bowel loops and undersurface of liver was done. On careful dissection, atretic segment approximately 15-20 cm from DJ junction was found. It was type IV atresia. End to end jejunojejunal anastomosis was done with transanastomotic feeding tube. A thorough search of abdominal cavity was done. Polysplenia found incidentally (Figure 4). Specimen sent for histopatho-

logical examination. Post-op period was uneventful. Tube feeding was started on 2nd post-op day. Patient was allowed breastfeeding from 6th post-op day and discharged on 12th post-op day.

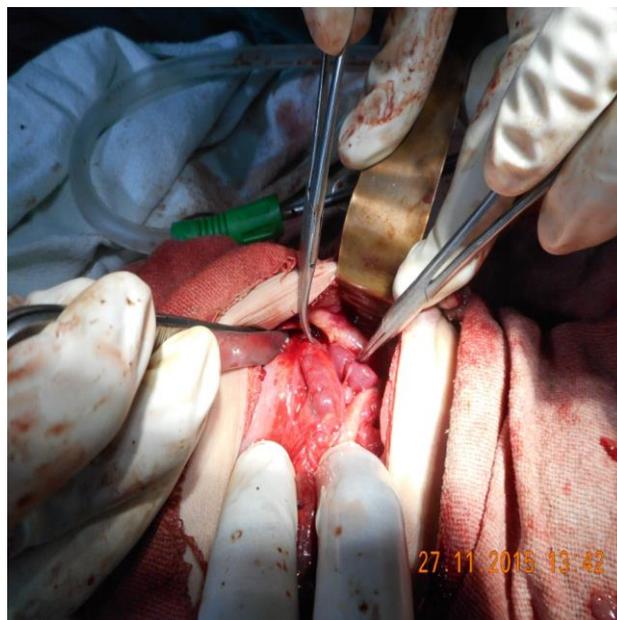


Figure 4: Polysplenia found incidentally.

DISCUSSION

The incidence of JIA varies from 1:330 and 1:400 live births in some reports to between 1:1500 and 1:3000 live births in others and JA is said to account for about 40-50% of these.³

The clinical features of jejuna atresia include bilious vomiting and upper abdominal distension. Our patient had presented with both. Maternal hydramnios could be associated with it antenatally, but it was not present in this case. JA can present with various complications. Dilatation of proximal segment to an extent of huge cyst filled with meconium is one of such rare complication.

Jejunal atresia has been associated with a number of other congenital malformations such as cystic fibrosis, malformation, congenital heart disease, biliary atresia, total colonic aganglionosis etc.^{4,5} We reported presence of polysplenia. There was two spleen of equal size with separate hilum at their normal location. A similar case report revealed an association of polysplenia with type 4 intestinal atresias⁶ but dilatation of proximal segment as huge meconium cyst was not there.

Simultaneous occurrence of jejuna atresia with meconium cyst and polysplenia is an extremely rare. A successful management of a neonate suffering from intestinal obstruction depends solely on making early diagnosis and treatment. Non-specific symptoms and rare incidence like this creates a diagnostic and therapeutic challenge to surgeons.

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