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Case Report

A rare triad: complicated meconium ileus with type 3b jejunal atresia and antenatal mid jejunal perforation complicated by late-onset sepsis induced hemophagocytic lymphohistiocytosis

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ABSTRACT

Complicated meconium ileus (MI) involving mid jejunal perforation with congenital adhesions bands with type IIIB jejunal atresia (apple-peel deformity) is a high-risk surgical emergency. While surgical management is often the primary focus, the postoperative course can be derailed by systemic inflammatory syndromes. We report a rare case of a 35-week neonate with antenatal perforation and Type IIIB atresia who, following successful surgical repair, developed late-onset culture-proven sepsis. This subsequently triggered hemophagocytic lymphohistiocytosis (HLH), a life-threatening hyperinflammatory condition. Through aggressive multidisciplinary management, the neonate survived and is currently a healthy two-year-old. This case emphasizes the need for early recognition of HLH in neonatal sepsis cases that fail to respond to standard therapy.

Keywords: Jejunal atresia, Antenatal perforation, Neonatal immune system, Hyperinflammatory condition

INTRODUCTION

Jejunal atresia (JA) occurs in approximately 1 in 5,000–14,000 live births.[1] Grosfeld has classified JA into five anatomical types of which type 3b JA, or "apple-peel" atresia, is particularly challenging due to significant mesenteric defects and compromised vascular supply and is an uncommon variant.[2] When associated with meconium ileus and antenatal mid jejunal perforation, the surgical complexity increases.

Furthermore, the neonatal immune system is prone to dysregulation; sepsis can serve as a trigger for secondary hemophagocytic lymphohistiocytosis (HLH). It is rare in neonates, often misdiagnosed as refractory sepsis, and carries a high mortality rate without prompt immunomodulatory intervention.

HLH is a clinical syndrome characterised by the uncontrollable activation of macrophages and T lymphocytes. It may have hereditary causes due to abnormalities in the genes that controls how naturally occurring killer cells and cytotoxic T lymphocytes functions. Infections, cancers, autoimmune and autoinflammatory illness as well as acquired immune deficiency state are other situations in which HLH may develop.

CASE REPORT

A female neonate (2400 gm) was born by normal vaginal delivery at 35 weeks' gestation to a gravida 3, para 2, live 2 elderly multigravida (age 36 year). Antenatal ultrasound at 34 weeks suggested mid-jejunal perforation and dilated bowel loops. The neonate was admitted to neonatal intensive care unit immediately following birth on 10 April

2024, with respiratory distress and gross abdominal distension. Ryles tube contained 110 ml bilious fluid aspirate just after delivery. Baby was put on non-invasive positive pressure ventilation (NIPPV) respiratory support with 25% FiO_2. Bilious drainage from stomach was replaced by Ringer lactate (volume by volume). Initial blood investigations were within normal parameters. Abdominal X ray revealed proximal dilated bowel loops (Figure 1) and ultrasound abdomen was suggestive of small bowel obstruction, hence surgical intervention was planned and baby was put on ventilatory support with minimal settings. Baby passed meconium pellet at 48 hour of life after giving glycerine suppository (Figure 2).

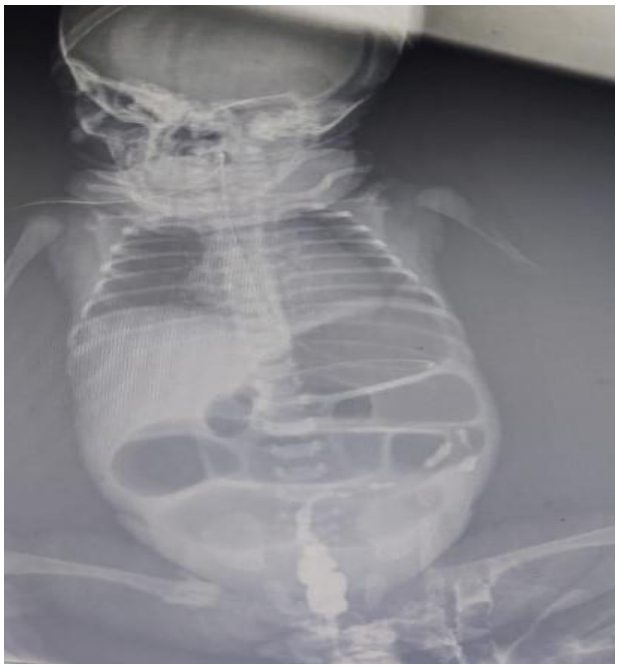

Figure 1: Abdominal X-ray showing proximal bowel obstruction.

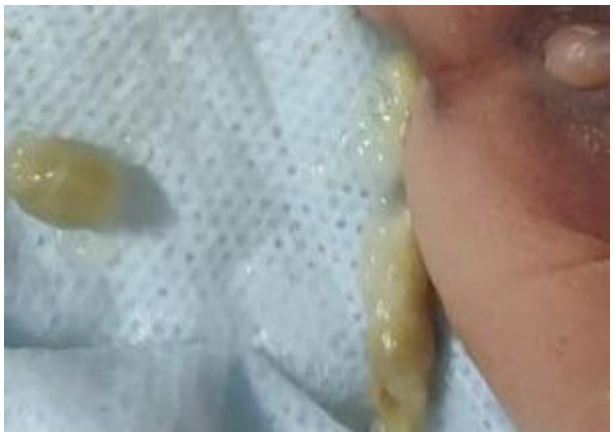

Figure 2: Hard pellet of stool.

An exploratory laparotomy was performed at 50 hours of life which revealed complicated meconium ileus. Key findings included the following.

Antenatal mid jejunal perforation

Evidence of a mid-jejunal perforation that had occurred in utero, leading to localized meconium peritonitis.

Congenital adhesion bands

Additional mechanical obstruction was noted due to congenital bands.

Type IIIb jejunal atresia

A proximal jejunal atresia with a large mesenteric defect; the distal small bowel was found coiled around a precarious marginal artery (apple-peel appearance) (Figures 3 and 4).

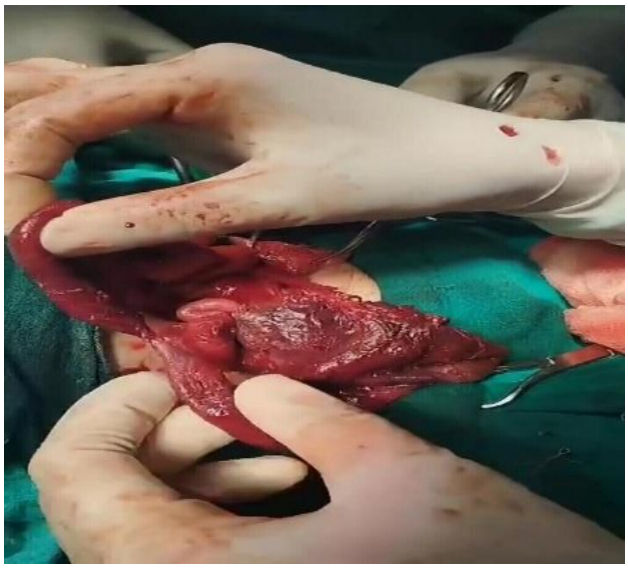

Figure 3: Intraoperative picture showing jejunal atresia.

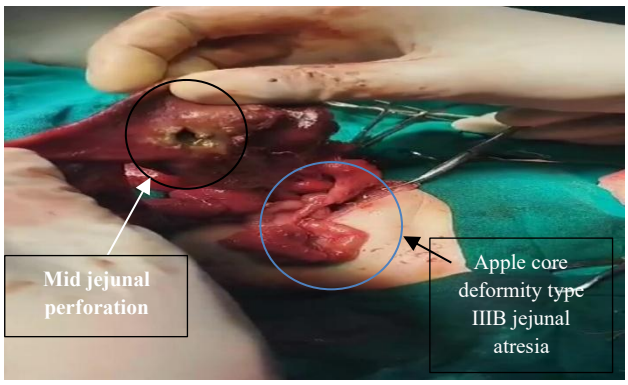

Figure 4: Intraoperative picture showing mid jejunal perforation and Apple core deformity.

The surgical team performed primary repair for perforation, adhesiolysis, resection of the atretic jejunal segments present at distance of 60 cm from DJ (duodeno-

jejunal) flexure and patency was made by primary end to side jejuno-jejunal anastomosis.

The initial postoperative recovery was focused on establishing bowel function and nutritional support (total parenteral nutrition-10% Aminoven @3.5 gm/kg/day). Baby was extubated to NIPPV after 24-hour (Figure 5).

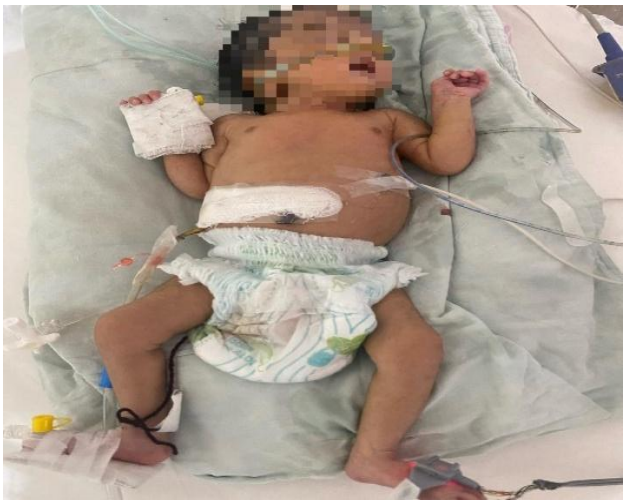

Figure 5: Day 2 post-operative.

Subsequently after 48-hours baby developed cyanosis, poor peripheral perfusion, urine output was <1 ml/kg/hour, ABG showed mild metabolic acidosis, hence was mechanically ventilated with 35% FiO_2 with inotropic support after giving normal saline bolus @10 ml/kg over 30 min. Antibiotics were started as per unit protocols, however initial blood culture was sterile. Relevant investigations showed features of pancytopenia. Packed red blood cells were transfused in view of anaemia. Gradually ventilator settings were decreased and baby was extubated to NIPPV support.

At day thirteen of life baby developed high grade unremitting fever and subsequently massive spleno-hepatomegaly (splenic notch was palpable) with progressive cholestasis with late-onset culture-proven (pseudomonas aeruginosa) sepsis. sensorium was normal and there was no evidence of bleeding manifestations CSF study (routine, microscopy and culture) was normal. Despite appropriate antibiotic escalation, the clinical state deteriorated into a hyperinflammatory profile.

Initially liver function test was normal but later on showed features of mild transaminitis (SGOT/SGPT-145/60), low albumin, mildly increased Prothrombin time and total bilirubin of 21 (direct was 19). Significant cytopenias (Hb 99 g/l, TLC-4100 N1×10^7/l platelet count 1.4×10^6/l), hyperferritinemia (763 µg/l), lactate dehydrogenase (LDH) (663 U/l (250-450), hypofibrinogenemia (qualitative) and hypertriglyceridemia (358 mg/dl), fulfilling the HLH-2004 diagnostic criteria of 5 out of 8 as shown in Table 1. The diagnosis of acquired HLH was made, triggering factor being the septic insult.

Table 1: Diagnostic criteria for HLH fulfilled 5 out of 8 criteria.

HLH	Patient
Fever	101 ^{0}C
Splenomegaly	Present
Cytopenia (affecting ≥2/3 lineages in peripheral blood)	Present
Hemoglobin <100 g/l	99
Neutrophils <1.0×10^9/l	1×10^7/l
Platelets <100×10^9/l	1.4×10^9/l
Hypofibrinogenemia and/or hypertriglyceridemia	Present
Hypofibrinogenemia ≤1.5 g/l	Qualitative
Fasting triglycerides ≥265 mg/dl	358
Ferritin >500 µg/l, max 1185.50 µg/l	763
Hemophagocytosis in bone marrow or spleen or lymph nodes, no evidence of malignancy	Not done
Low or absent NK-cell activity	Not available
Sil-2r ≥2400 U/ml, max 2415 U/ml	Not available

We couldn't conduct genetic test to rule out possibility of familial HLH due to high cost and serum soluble interleukin IL-2 (CD 25) was not available at our hospital.

Peripheral smear for malaria (haemoparasite) was negative. Coombs test was negative and reticulocyte count were also normal. Mother TORCH profile was normal and baby urine CMV PCR was negative. Urine for fungal hyphae (KOH preparation) and culture was sterile. Viral PCR test for influenza was negative and not available for other viruses like Herpes simplex virus (HSV), Epstein Barr virus (EBV). Eye examination was normal with no evidence of chorioretinitis and cataract. 2D ECHO and cranial USG were normal. On X-ray abdomen at birth there was no evidence of intraperitoneal calcification and USG abdomen didn't show features of cystic meconium peritonitis. There was no history of consanguinity of marriage, blood group incompatibility, maternal diabetes mellitus and pregnancy induced hypertension.

General examination showed no evidence of microcephaly, bulging fontanel, dysmorphic facial features, café-au-lait spot, hemangioma, micropenis, tuft of hair over spine, ecchymotic patches, hernia or limb deformities.

Treatment included intravenous immunoglobulin (IVIG) given in a dose of 1 gm/kg/day for 2 days (partial response) then methyl prednisolone 10 mg/kg/day was added for 3 consecutive days and supportive care to dampen the cytokine storm.

Outcome and follow-up

The neonate gradually responded to treatment after 3 weeks. Clinically fever subsided, rapid regression of spleen and liver size and subsequent extubation to NIPPV

support with minimal setting and later started maintaining oxygen saturation at room air. Hemogram gradually normalized (Figure 6). Optimum weight gain of 15-20 gm/kg/day began till birth weight was achieved, subsequently followed by 20-30 gm/day. She was successfully transitioned to full oral feeds. She was discharged at 34-day of life in stable condition with normal lab parameters.

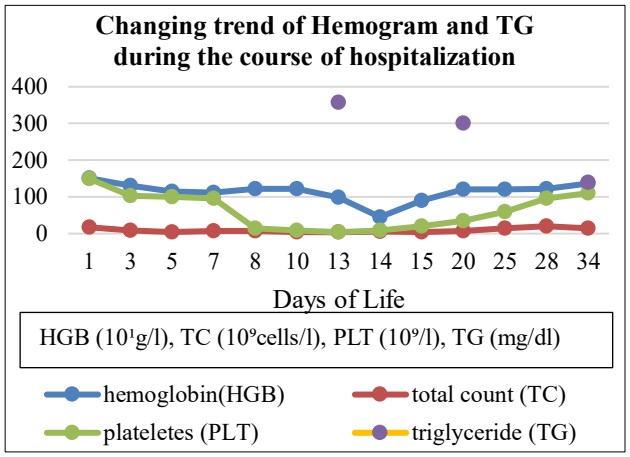

Figure 6: Changing trend of hemogram and TG during the course of hospitalization.

Now, at two years of age, the child is thriving, showing normal growth and developmental milestones, with no long-term sequelae from the "apple-peel" atresia or the HLH episode (Figure 7).

Figure 7: At 2 years of age.

DISCUSSION

We present a unique case of premature neonate of surgical and medical pathologies.

A neonatal jejunal atresia is a form of intestinal obstruction characterized by a blockage in the jejunum, which can result from narrowing, complete closure or absence of a segment of the jejunum. The incidence of intestinal atresia varies from 1 in 5000 to 1 in 14000 live births.[3]

There are five main classifications of jejunoileal atresia: type I involves a membrane that fully obstructs the intestinal lumen while maintaining the integrity of the intestine. Type II is characterized by a discontinuity in the intestine with a fibrous cord connecting the proximal and distal segments. Type III A is characterized by a mesenteric gap without any linkage between the segments. Type III B presents jejunal atresia with the absence of the distal superior mesenteric artery, resulting in the distal small bowel coiling like an apple peel and a shortened gut as observed intra operatively in this case (figure). Type IV comprises multiple atretic segments, resembling a string of sausages.[4]

The diagnosis of neonatal ileal atresia relies on clinical history, physical examination, and imaging studies like abdominal X-rays and ultrasound. Clinical manifestations may include bilious vomiting, abdominal distension, and failure to pass meconium, as observed in this case report.[5] In a significant number of cases (29-50% involving jejunoileal atresia), prenatal detection is possible through fetal ultrasound, revealing signs such as polyhydramnios, ascites, dilated bowel loops, and increased bowel echogenicity.[6] Like in this case report, the mother underwent an obstetric ultrasound examination antenatally and was diagnosed with mid jejunal perforation.

Supportive treatment for neonatal jejunal atresia includes intravenous fluids, nasogastric decompression, and antibiotics. Definitive treatment involves a surgical intervention to remove the obstruction and restore normal intestinal continuity, this may be achieved either with primary anastomosis or stoma creation first.[7,8] A decision to do a resection and anastomosis was made in this case (Figure) due to the location of the lesion. Managing 3b atresia requires preserving every centimetre of the distal "apple-peel" segment, as these neonates are at high risk for short bowel syndrome (SBS).

The mortality rate after surgery for neonatal jejuno-ileal atresia is relatively low, with most studies reporting a mortality rate of less than 5% attributed to advancements in neonatal intensive care, anaesthesia, surgical techniques, and the utilization of total parenteral nutrition (TPN).[9]

Although HLH can occur across all age groups, HLH with neonatal onset within the first 4 weeks of life is rare, accounting for only 4% of cases.[10] Our case was having onset very early within the 2nd week of life.

In HLH, there is a dysregulation of the innate immune system, particularly involving natural killer (NK) cells and CD8+ cytotoxic T-cells. Normally, cytotoxic T-cells produce two key cytolytic enzymes – perforin and granzyme. Perforin creates destabilising pores in the membranes of target cells, enabling granzyme to enter and trigger cell destruction. However, in HLH, this process is disrupted due to genetic mutations or immunogenic stimuli. As a result, additional cytotoxic cells are recruited

but fail to eliminate the pathological antigen, leading to a massive increase in circulating cytokines. This hypercytokinemia causes widespread macrophage activation, hemophagocytosis and severe organ-damaging inflammation.[11]

Diagnosis of HLH can be established if a molecular diagnosis is consistent with HLH, or 5 out of total 8 diagnostic criteria for HLH are fulfilled (HLH-2004 criteria).[12] Our case had splenomegaly, cytopenia, hypertriglyceridaemia, hypofibrinogenaemia and raised ferritin. Because of financial constrain, we were not able to do genetic testing as also special tests such as NK cell activity and soluble CD25.

The cornerstone of treatment for HLH is immunosuppression and cytotoxic therapy. The recommended approach follows the HLH-2004 protocol developed by the Histiocyte society. This protocol includes a combination of dexamethasone, cyclosporine A, etoposide and intrathecal methotrexate (for patients with neurological involvement), followed by a bone marrow transplant.[13]

As neonatal HLH carries a grim prognosis, high level of suspicion and early diagnosis is very important. It should be included in the differential diagnosis for cytopenias and multi-organ dysfunction, particularly when no infectious or metabolic cause is identifiable.[14] Worsening thrombocytopenia of unknown origin in newborns should prompt consideration of neonatal HLH. Without treatment, children with HLH invariably face a fatal outcome.

In this case, the culture-proven sepsis acted as the "first hit," leading to a pathological immune activation. The successful outcome here was dependent on recognizing that the neonatal decline was no longer just due to infection, but due to an uncontrolled immune response.

CONCLUSION

The survival of a neonate with type 3b atresia complicated by antenatal perforation and subsequent HLH is rare. It highlights that even in the face of extreme surgical and immunological challenges, a multidisciplinary approach combining meticulous neonatal surgery with early haematological intervention can lead to excellent long-term outcomes.

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