Neonatal Multisystem Inflammatory Syndrome (MIS-N) Presenting as Necrotizing Enterocolitis and Cardiac Dysfunction

Multisystem inflammatory syndrome in neonates (MIS-N) associated with perinatal severe acute respiratory syndrome 2 (SARS-CoV-2) exposure is increasingly being reported in recent times [1-3]. MIS-N presents with a variety of clinical presentations, requires a high index of suspicion, and is a diagnosis of exclusion.

We report a female neonate of 2.64 kg, born late preterm at 35 weeks gestation, by cesarean section due to placenta previa and preterm labor. The baby was vigorous at birth, treated initially for transient tachypnea of the newborn (TTNB), and shifted to the mother’s side on day 2 with exclusive breastfeeding. The baby developed abdominal distension with bilious vomiting on day 3 of birth and was shifted to NICU. She developed fever spikes of 38.5°C and shock within 12 hours of abdominal symptoms. On examination, she was febrile, drowsy, had tachycardia (heart rate, 202/min), had hypotension (mean blood pressure:32 mm of Hg) with cool peripheries, tachypnea (respiratory rate:72/min) with chest retraction, and oxygen saturation of 96% on 30% FiO2 with 5 cm of CPAP support. The precordium was hyperdynamic with a systolic murmur of grade 3/6 over the left infraclavicular area, hepatomegaly of 3 cm below right costal margin and tender abdomen. X-ray abdomen showed dilated bowel, right iliac soap bubble appearance and left sided pneumatosis intestinalis (Fig. 1). An echocardiogram revealed moderate biventricular dysfunction with poor contractility (left ventricular ejection fraction of 42%), dilated right atrium and right ventricle, large patent ductus arteriosus (4 mm) with a left to right shunt, dilated inferior vena cava, and normal coronaries. The sepsis screen was positive with elevated CRP (61.6 mg/dL) and neutrophilic leukocytosis (total 22×10^9/L, 78% neutrophils). A diagnosis of probable sepsis with hemodynamically significant PDA (HS-PDA) and necrotizing enterocolitis (NEC) was made. Treatment included nil per oral, intravenous fluids, orogastric decompression, antibiotics, frusemide, inotropic support, and mechanical ventilation. On reviewing the history, mother was unvaccinated for coronavirus disease 2019 (CoVID-19) and had a SARS-CoV-2 infection three weeks before delivery.

The baby had high titers of IgG antibodies 241.13 AU/mL (<10.0 non-reactive) with negative IgM titers for SARS-CoV-2. The SARS-CoV-2 reverse transcriptase – polymerase chain reaction (RT-PCR) of the neonate was negative. Inflammatory markers done on day 4 of birth were elevated, (IL6 19.9 pg/mL, ferritin:244 ng/mL, NT-Pro BNP: >35000 pg/mL), serum LDH 1086.00 U/L, D-dimer: 4259 ng FEU/mL. Blood culture was sterile. The baby was treated with dexamethasone (0.15 mg/kg/dose 12-hourly for 3 days followed by oral prednisolone 1 mg/kg/day for 4 days) and intravenous immunoglobulins (1 g/kg/day of IVIG for 2 days). The baby gradually improved in the next 48 hours with improvement in cardiac function, closure of the PDA, resolution of abdominal distension, shock, and normalization of inflammatory markers. The baby was extubated on day 5, full feeds were achieved on day 8, and was successfully discharged on day 10.

Acute deterioration on day 3, shock needing inotrope support and ventilation, NEC requiring supportive care, cardiac dysfunction, negative blood cultures, high inflammatory markers, positive COVID serology with maternal COVID history led us to the possibility of MIS-N in this newborn, which meets the modified CDC criteria for MIS-N.

The exact pathogenesis for MIS-N remains elusive with the most possible hypothesis being immune dysregulation and endothelial injury. Neonates with MIS-N present with respiratory (respiratory distress), cardiac (cardiac dysfunction, coronary aneurysms, thrombus, conduction abnormalities), gastrointestinal (NEC), central nervous system (encephalopathy, stroke), dermatological (vasculitis rash), and sepsis-like (fever, hypothermia, shock) manifestations. Immunomodulator therapy (IVIG, steroids) forms the crux of management of MIS-N [5].

We conclude that MIS-N should be considered in any neonate with unexplained necrotizing enterocolitis and/or cardiac dysfunction, after ruling out the common causes.

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Rapunzel Syndrome: Diagnosed After Laparotomy in a Young Girl

Rapunzel syndrome is an unusual and rare form of trichobezoar found in patients with the habit of hair pulling (trichotillomania) and swallowing it (trichophagia), consequently leading to collection of bezoars in stomach and intestines. We describe this syndrome in a very young girl who presented to us as a case of moderate acute malnutrition with intestinal obstruction.

Bezoars have been known to occur in the form of undigested masses found in the stomach but Rapunzel syndrome involves the presence of a gastric trichobezoar with a long tail extending beyond the duodenum and till terminal ileum [1]. Around 100 cases have been described in the literature since then, with a mean age of presentation of 10.8 years [2,3]. It is usually seen in young girls with or without known psychiatric disorders [4].

A 5-year-old girl presented to us with poor growth and not eating well. There was history of pain abdomen and occasional vomiting after meals. On examination, the child looked pale, emaciated, and stunted. Her hemoglobin level was 9 g/dL. Her weight was 11.2 kg (< 1st centile according to WHO weight-for-age chart) and her height was 92.5 cm (<1st centile WHO height-for-age chart). The abdomen was mildly distended with normal bowel sounds. The patient’s mother admitted that she had a habit of picking hair from floor and secretly swallowing it (trichophagia), consequently leading to the formation of bezoars. A diagnosis of moderate acute malnutrition with moderate anemia with subacute intestinal obstruction was made. The patient was kept nil per orally and received intravenous fluids, and surgical opinion was sought. After two days, the child passed stool mixed with hair strands with relief of abdominal distention. The child was allowed to eat semi-solid food, which resulted in vomiting of strands of hair and recurrence of abdominal distention. CBC showed moderate anemia with dimorphic picture. X-ray abdomen showed dilated gut with multiple air fluid level at various level. Serial USG abdomen failed to detect any intra luminal mass in the stomach or duodenum and ileum. Computed tomography was not available at that time.

Exploratory laparotomy was performed through a supraumbilical midline abdominal incision. A longitudinal 4 cm gastroscopy made on the anterior surface of the corpus of the stomach revealed an intraluminal smooth contour mass occupying bulk of stomach with post pyloric extension. There was a continuous thin strand of trichobezoar in duodenum with thick tail of 2-3 cm diameter along jejunum and terminal ileum, which were removed with separate enterotomy.

After discharge child was provided nutritional rehabilitation. Five months post-surgery, the child has gained 3 kg weight and is on iron supplements. She is receiving Behavioural modification advice but not on any psychiatric treatment.

Rapunzel syndrome is rare but should be taken into consideration while investigating a malnourished child with intestinal obstruction. Trichotillomania and trichophagia as a diagnosis are considered on finding some bald patches on scalp or history of habit of pulling out hair and swallowing it [4]. This child did not have bald patches on her scalp.

Upper gastrointestinal endoscopy is the gold standard diagnostic modality but was not available at this hospital [3]. USG abdomen, although is said to be specific for trichobezoar, but in our case serial ultrasound abdomen failed to detect the nature or presence of any intra luminal mass. Ultrasound as an imaging modality has often missed subtle finding of a trichobezoar [5]. CT scan reveals the nature and the extent of the trichobezoar, and is regarded as the best modality for diagnosis. Obstruction of the upper digestive tract is the most common clinical manifestation of this disorder [1].

In the present case, surgery was attempted as an exploratory method due to uncertain cause of intestinal obstruction. Psychiatric follow-up is important, and care should be extended to family members, who should be vigilant with patients since recurrences of the problem have been described [4]. The need for adequate follow-up should be emphasized to avoid recurrences, although these are rare since the trauma of surgery may prevent the patient from provoking another episode.

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