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**Introduction**

Hydrops fetalis (HF) is the result of an imbalance in the regulation of fluid, leading to an increase in interstitial fluid production or a decrease in lymphatic return. HF can be diagnosed prenatally by ultrasound and defined by the presence of more than two abnormal fluid collections (ascites, pericardial/pleural effusion, skin edema) in the fetus. Non-immune hydrops fetalis (NIHF) comprises the subgroup of cases not caused by red cell alloimmunization [1]. The prevalence of NIHF in the general population is estimated to be 1 in every 2500-3500 neonates and 1 in every 1600-7000 fetuses [2]. Wide variations in reported prevalence are due to differences in definitions, populations, thoroughness of evaluation, and whether late pregnancy terminations were included. Despite extensive investigations, the etiology of NIHF remains unknown in 15-25% of these cases. Though NIHF has got poor prognosis, several etiologies can be treated with potentially good results. One very rare but treatable condition is Meconium peritonitis (MP). MP was first discovered by Morgagni in 1761, but the first corrective surgery was executed successfully in 1943 by Agerty [3]. MP is a rare fatal disease characterized by sterile inflammatory reaction, secondary to extravasation of meconium into the peritoneal cavity. The pathological event described for the occurrence of MP is intrauterine bowel perforation that may occur during antenatal or postnatal period [4]. The key element for the management consists of prenatal diagnosis and excluding chromosomal disorders, congenital infections and cystic fibrosis. Early surgical procedures to reduce systemic and abdominal inflammation just after birth may improve the outcome of severe MP cases. Recently, the survival rate for MP increased to over 90%. This improvement is the result of an advance in fetal diagnostic techniques, timely intervention and intensive care after birth [5].

**Case Report**

A 32 year, G5P2L1(IUD)A2, non-consanguineous marriage, was referred to our hospital at 33 weeks 2 days of gestation in view of isolated fetal ascites, diagnosed on antenatal scan at 32 weeks. Mother’s blood group was “A” positive, ruling out the possibility of incompatibility. First trimester aneuploidy screen was indicative of low risk. Second trimester serum screening (quad test) and targeted imaging for fetal anomalies were normal. Immuno-hematological work up including indirect coombs test, irregular antibody screening by 3 cell panel was negative. There was no history of congenital anomalies in family or in her previous babies. Work up done to rule out other possible causes including syphilis, cytomegalovirus (CMV), parvovirus B19 and toxoplasmosis, and the reports were normal. Fetal echo was done, which showed structurally normal heart with mild pericardial effusion and echogenic foci in both ventricles. One week later, repeat ultrasound was done which showed moderate fetal ascites, fluid collection in infra diaphragmatic space with echogenic bowel loops floating within it, also few areas of calcification in the bowel loops with prominent Inferior vena cava (IVC), and associated polyhydramnios was noted. Dopper middle cerebral artery peak systolic velocity was < 1.5 multiples of median which rules out the probability of fetal anemia (Figure 1). There was no evidence of hydrocephalus, hydrothorax or skin edema. The probable diagnosis of MP was made. Neonatologist and pediatric surgeon’s opinion were taken regarding fetal prognosis and further management. She was planned for conservative management with an aim to prolong the pregnancy till 37 weeks. Dexamethasone was given for fetal lung maturation. Subsequently, she went into spontaneous labour and a preterm hydropic female baby of birth weight 3.05 kg was delivered at 35 weeks, cried immediately after birth, the baby had ascites, and vulval edema (Figure 2). Immediate intubation was done in view of respiratory distress and shifted to neonatal unit for further workup and ventilator support. Placental examination revealed large placenta weighing 1.1 kg, however no other placental abnormality seen (Figure 3). Placental tissue was sent for histopathological examination, Polymerase chain reaction (PCR) for Treponema pallidum, CMV and parvovirus DNA, which all reported negative. Umbilical cord blood sent for cytogenetic analysis and revealed normal chromosomal complement. Post natal ultrasound of the neonate was done which showed gross thick particulated ascites, a giant cyst compressing the IVC, and minimal bilateral pleural effusion. X-ray whole abdomen revealed few spots of intra-abdominal calcification. Emergency exploratory laparotomy was performed on day three of life. Intra-operative findings consisted of clumps of collapsed small bowel loop, a giant meconium cyst of 8 x 10 cm containing approximately 150 ml of thick meconium, which was drained out, there was also terminal ileal perforation of about 10-14 cm proximal to ileo cecal junction which was brought out as loop stoma (double barrel ileostomy). Postoperatively baby was extubated and was maintaining saturation with oxygen by nasal prongs, edema started resolving and over all neonatal condition improved (Figure 4). On postoperative day five, baby had one episode of febrile seizure and was started on injection levetiracetum. Ileostomy started...
duced drastically due to universal use of immunoprophylaxis for red cell isoimmunization. Consequently, NIHF accounts for almost 90% of cases of HF. MP has been reported as one of the curable etiologies of NIHF. With the evolution in the imaging technologies increasing number of fetuses with MP are being diagnosed prenatally.

**Discussion**

The occurrence of immune hydrops has been re-

Diagnosis of MP is rare before 20 weeks’ because peristalsis rarely commences before this time. The median gestational age at initial diagnosis of MP was 24 weeks [6]. According to the study by Uchida k, et al.
Figure 1: Antenatal ultrasound showing Fetal ascites, Echogenic bowel loops, Polyhydramnios and Normal doppler middle cerebral artery peak systolic velocity.

Figure 2: Immediate post-natal period-picture of Hydropic female baby.

Figure 3: Large placenta.

Prenatal diagnosis was made in 73% of patients. The ultrasound (US) findings with suspected MP were polyhydramnios (100%), bowel dilatation (53%), ascites (33%), and pseudocyst (13%) [5]. In another study by Ping LM, et al. Fetal ascites (93.3%) was the most common prenatal US finding [6]. Antenatal US has high specificity (100%) but low sensitivity (22.2%) in detecting meconium pseudocyst. Prenatal Magnetic Resonance imaging can improve the low diagnostic yield of prenatal US scan.
Secondary to in utero bowel perforation has been shown to activate immune cells including macrophages. Infiltration of macrophages into the peritoneum will lead to activation of cellular functions, including phagocytosis, liberation of chemical mediators, and antibody-dependent cell-mediated cytotoxicity. The intense inflammatory reaction leads to the formation of a dense, adherent membrane that practically seals off the intestine at the site of perforation. However, if the sealing is incomplete, a thick-walled cystic space is formed, and meconium will continuously keep collecting in this cystic pocket. Any cause of small bowel ischemia or associated mechanical obstruction such as intestinal atresia, volvulus, intussusception, congenital bands, and meconium plug syndrome, as in cystic fibrosis, may result in the genesis of meconium peritonitis [5].

Depending on the degree of inflammation, three pathological variant of MP can be seen: Fibro-adhesive, cystic and generalized. The most common variant is fibro-adhesive type, resulting from enormous fibroblastic reaction; cystic type is seen when the perforation site is not completely sealed and thus forming a thick-walled cyst. Generalized MP presents as diffuse bowel thickening of the affected segment, peritoneal fibrosis and calcium deposits.

Recent studies do not provide clear guidelines concerning surgical strategies for MP. Enterostomy, T-tube ileostomy, primary anastomosis, Bishop-koop, santulli and Mikulicz are common procedures for MP. Although the type of surgical procedure seems to depend upon clinical manifestation, general condition of the patient and surgeon’s preferential technique, few comparative studies have been performed. Miyake, et al. consider primary anastomosis as safe option for almost all patients with MP except for those with very low birth weight and in an unstable condition. The advantages of primary anastomosis are reduced hospital stay, avoidance of stoma related morbidity and second laparotomy for stoma closure. Nam, et al. reported a preference for primary resection and anastomosis of the intestinal segment involved. However severe complications related to the surgical procedure itself, such as peritonitis from anastomotic leakage and perforation caused by frequent manipulation are more often seen in primary anastomosis. It is difficult to assess objectively the viability and condition of the intestine, therefore karimi, et al. recommended resection with temporary double barreled enterostomy as the safest treatment [9]. The surgical strategy for our case was two stage approach with abdominal drainage or temporary enterostomy and elective reconstruction of intestinal continuity (stoma closure) at fifth month of life. However stoma closure could not be done in our case as the baby succumbed on fourth week of life due to sepsis. Recently, the first choice for cystic type MP was abdominal decompression by catheterization closed drainage.

Infants usually present with tense abdominal distension, edematous wall with shiny skin and visible veins, respiratory distress, bilious aspirates/ vomiting, failure to pass meconium and features of peritonitis [3]. Abdominal plain X-ray reveals the calcifications in the peritoneal cavity occasionally; the calcifications may extend to the scrotum. On US, MP produces multiple discrete, very highly reflective foci, with acoustic shadowing or occasionally diffuse peritoneal reflectivity (referred to as a ‘snowstorm’ appearance). Postnatal contrast Computed Tomography (CT) scan is required to define persistent intestinal perforation invisible with prenatal US scan. In most cases surgical intervention is required immediately after birth; Spontaneous healing is reported in rare cases. Caro-Dominguez, et al. reported that postnatal imaging findings that are predictive of the need for surgery include intestinal obstruction, ascites, pneumoperitoneum, and volvulus; however, the presence or distribution of peritoneal calcification was not predictive of the need for surgery [7]. Neonatal sepsis is reported by several authors to be one of the most common causes of mortality as was in our case also.

The incidence of chromosomal abnormalities and genetic syndromes is not increased in cases with MP however a relatively strong association with cystic fibrosis is seen in between 8%-40% of patients [8]. Therefore amniocentesis & DNA studies for cystic fibrosis should be done if both parents are carriers. However we could not evaluate in our case for this due to non affordability issues.

Meconium is a composite mixture of bile salts, cell debris, and proteins. Spillage of these constituents sec-
Informed consent
Informed consent was obtained from the patient.

Conflict of interest
The authors declare that they have no competing interests.

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Conclusion
Prenatal diagnosis is crucial for the first step of perinatal therapy for MP. Management and the need for surgery depend on the clinical presentation and the overall condition and gestational age at birth of the newborn. Surgery is required when signs of intestinal obstruction are present. Early diagnosis, use of higher antibiotics and active management of acid base imbalance, superimposed bacterial peritonitis, and septic shock can prevent mortality. Timing of delivery should rely on composite decision of gynecologists, neonatologists, and neonatal pediatric surgeons in perinatal and maternal care centers. Surgery performed within 24 hours in newborns with bowel obstruction may also improve their outcome.

Declarations

Ethical approval and consent to participate
Not applicable.