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Cystic Fibrosis Related Metabolic Syndrome (CRMS): An Under diagnosed Condition

Dr. Ahmad Khursheed[1], Dr. Pankaj Garg[2]

DESCRIPTION

M is a new-born male delivered at 35 weeks of gestationto a 28 year old mother. The antenatal history was not significantexcept for Gestational Hypertension which was controlled onoral antihypertensive (Labetalol). Birth weight was 2200gm and ApgarScore were 8,10 at 1 and 5 minutes respectively. Baby was noticed to haverespiratory distress for which baby was put on Nasal CPAP at FiO2 of 30% and PEEP of 6cm of H20 and was started on Orogastric tube feeds .The baby developed Bilious vomiting and abdominal distension for which baby was made Nil per oral and X-ray was done which revealed air under diaphragm(Figure 1) for which emergency glove drainage was done prior totransfer our neonatal intensive care unit(NICU).

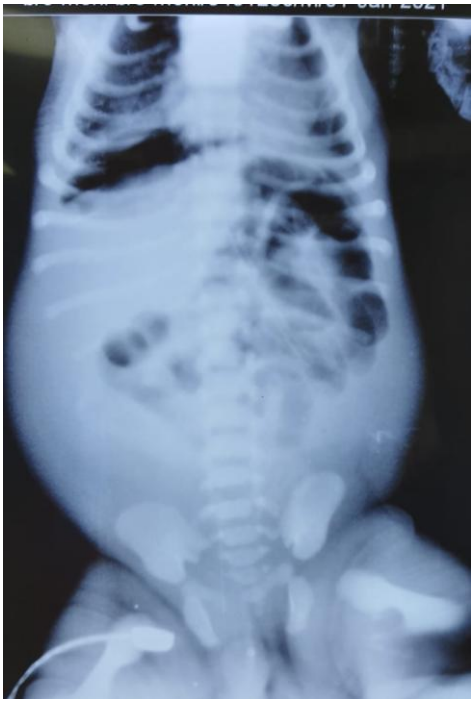

Figure 1:

On arrival at NICU ,the baby was lethargic with cold peripheries with delayed CFT of >3 seconds, abdominal distension, erythema, pus discharging from drain, tenderness and absent bowel sounds. Baby was electively ventilated ,started on ionotropes and was empirically started on antibiotics after taking sepsis screen including blood culture. X-ray was done which showed air under diaphragm. Baby was planned for urgent exploratory laparotomy which showed large small bowel perforation at ileum, microcolon filled with meconium like cast. A portion of necrotic small bowel was removed with construction of an ileostomy. After surgery baby was gradually weaned off from ionotropes and ventilator. Feeds were reintroduced on Post-op day 8 which baby tolerated well and hiked gradually and was discharged home on day 24 of hospital stay.

Baby was evaluated for new-born screening for cystic fibrosis with Immunoreactive trypsinogen(IRT) which was positive and was followed by Sweat Chloride test was done which was suggestive of Cystic Fibrosis

related metabolic disease. Baby was also screened for most common mutation causing Cystic Fibrosis(Delta 508) which was negative. Repeat Sweat chloride test was done which was negative and Expanded DNA analysis was planned.

DISCUSSION

Meconium ileus is an intraluminal obstruction of the terminal ileumby abnormal meconium. Cystic fibrosis (CF) accounts for 90%of patients presenting with meconium ileus, while up to 20% ofpatients with CF present as newborns with meconium ileus.1 Meconium ileus maybe "Simple" and result only in bowel obstruction, or itmay be "Complex" when the obstruction leads to prenatalperforation or twisting and ischemia 2.MI can be picked up antenatally by Ultrasonography by presence of Echogenic Bowel although it may be present in other conditions like>>>>>3.If not identified antenatally, the most common clinicalpresentation of MI is intestinal obstruction, which is oftenseen within hours of birth,when feedings are initiated, biliousemesis occurs with or without abdominal distention as was noticed in index case.In Complex MI peritonitis arises following prenatal gastrointestinalperforation.Meconium undergoes dystrophiccalcification intrauterine, which results in theclassic eggshell calcifications seen on X-ray4.

Neonate born with MI either Simple or Complex always should be screened for CF as early diagnosis of disease will avoid delayed intitation of disease specific therapy. Neonatal outcomes have improved over time as a result ofbetter diagnosis antenatally and with modern advances in paediatricsurgery and perinatology.5 Surgery remains the definitivetreatment with procedures including multiple drainage procedures,stoma creation and final ostomy closure.6

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