CASE REPORT

Overcoming Obstacles: A Case Report of Preterm Neonatal Respiratory Distress Syndrome with Coexisting Hydrocephalus, Aqueductal Stenosis, and Neonatal Hyperbilirubinemia

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Abstract

Aim: The aim of present report is to describe the diagnosis and management of a preterm neonate with coexisting respiratory distress syndrome (RDS), hydrocephalus, aqueductal stenosis, and neonatal hyperbilirubinemia (NNH).

Background: A 31-day-old preterm male infant with RDS, neonatal hyperbilirubinemia, Intrauterine growth restriction (IUGR), aqueductal stenosis, and hydrocephalus was admitted to the neonatal intensive care unit (NICU) and received oxygen support and double volume exchange transfusion. Physical examination showed yellow skin discoloration, triangular facies, moderate hepatosplenomegaly, and hypertonia. A hematological workup revealed anemia, leukocytosis, *Staphylococcus haemolyticus* growth, and elevated bilirubin levels. Ultrasonography (USG) brain showed dilated ventricles and aqueduct effacement, leading to the diagnosis of hydrocephalus due to aqueductal stenosis, and a positive toxoplasmosis, rubella cytomegalovirus, herpes simplex, and HIV (TORCH) test.

Case description: A 31-day-old preterm male infant with multiple medical conditions, including RDS, hyperbilirubinemia, IUGR, aqueductal stenosis, and hydrocephalus, was admitted to the NICU for 30 days. The baby showed yellow skin discoloration, triangular facies, moderate liver/spleen enlargement, hypertonia, and anemia. USG brain showed dilated ventricles and aqueduct effacement, leading to a diagnosis of hydrocephalus due to aqueductal stenosis. The baby was also diagnosed with TORCH (Toxoplasma) positive.

Conclusion: The present case report highlights the complexity and challenges associated with managing a preterm male infant with multiple medical conditions. The diagnosis of RDS, neonatal hyperbilirubinemia, IUGR, aqueductal stenosis, and hydrocephalus required a comprehensive approach, including oxygen support, double volume exchange transfusion (DVET), and hematological workup. The management of the infant's conditions emphasized the importance of prompt and accurate diagnosis and aggressive treatment strategies. The successful outcome in this case, highlights the potential of multidisciplinary approaches in improving the prognosis of infants with multiple medical conditions and emphasizes the need for continuous monitoring and intervention to ensure optimal outcomes.

Clinical significance: Clinical significance refers to the practical importance or real-world impact of the findings or information presented in the case report. It highlights how the case report contributes to the current understanding of a medical condition, its diagnosis, and treatment, and how it can help improve patient care. In a case report, the clinical significance is often summarized in the conclusion and highlights the important takeaways for healthcare professionals.

Keywords: Aquedutcal stenosis, Case report, Hydrocephalus, Neonatal-hyperbilirubinemia, Preterm infant, Respiratory distress syndrome.

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Background

We report a case of a premature infant showing hydrocephalus and aqueductal stenosis by antenatal USG, interpretating that respiratory distress syndrome (RDS) requires oxygen support and Neonatal hyperbilirubinemia requires double volume exchange transfusion (DVET).

A prominent cause of respiratory illness in preterm infants is RDS, frequently referred to as hyaline membrane disease. Infants whose mothers had diabetes when pregnant also have RDS. RDS is brought on by a lack of alveolar surfactant, which raises alveolar surface tension and causes micro-atelectasis and small lung capacities.[1] The development of air bronchograms and the etiology of RDS are both heavily influenced by pulmonary edema. In premature babies, excessive lung fluid is linked to epithelial damage in the airways, a decrease in sodium-absorbing channels in the lung epithelium, and relative oliguria in the first two days after delivery.[2] The clinical evaluation, oxygen saturation (SpO_2)

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Table 1: Downes score

Score	*Respiratory rate*	*Cyanosis*	*Air entry*	*Grunt*	*Retraction*
0	<60 breaths/minute	Nil	Normal	None	Nil
1	60–80 breaths/minute	In room air	Mild decrease	Audible with stethoscope	Mild
2	>80 breaths/minute or apnea	In >40% oxygen	Marked decrease	Audible without stethoscope	Moderate-to-severe

Table 2: Silverman Anderson score (SAS)

Score	*Upper chest**	*Lower chest#*	*Xiphoid retractions*	*Nares dilatation*	*Grunting*
0	Synchronized	No retractions	None	None	None
1	Lag on inspiration	Just visible	Just visible	Minimal	Heard with stethoscope
2	See-saw	Marked	Marked	Marked	Heard without stethoscope

*Upper chest - If a newborn is predominantly using their upper chest muscles to breathe, it suggests that they might be experiencing significant respiratory distress. This can indicate difficulty in effectively moving air into their lungs.
#Lower chest - If a newborn is primarily using their lower chest and abdominal muscles for breathing, it indicates more efficient breathing and suggests better lung function. This is generally considered a positive sign

and alveolar-arterial diffusion gradient of oxygen (A-aDO_2), fraction of inspired oxygen (FiO_2) requirements, oxygenation index (OI), oxygen saturation index (OSI), and arterial blood gas parameters are all valuable in determining the severity of RDS in a term infant.

The severity of RD can be objectively assessed using a variety of clinical scoring systems, the two most used of which are the Silverman-Anderson and Downes scoring systems (Tables 1 and 2). Preterm infants frequently utilize the SAS score, while term neonates typically use the Downes grading system.

Scores of 0 indicate no distress, 1–4 indicate mild respiratory distress, 5–7 indicate moderate respiratory distress, and >7 indicate severe distress or imminent respiratory failure.[3]

We report a 31-day-old male baby with RDS, neonatal hyperbilirubinemia, IUGR, Aqueductal stenosis, and hydrocephalus are rare associations.

The baby was diagnosed with RDS due to hydrocephalus and aqueductal stenosis.

Case Description

A male neonate was born at 34 weeks gestation (preterm) via vaginal delivery to 21-year-old gravida-1 and a para-1 woman. Birth weight was 1500 gm, and Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. Fetal growth was abnormal with hydrocephalus, aqueductal stenosis, neonatal hyperbilirubinemia, and IUGR. The baby cried after birth and weighed 1500 g. The baby had RDS required oxygen support and neonatal hyperbilirubinemia require DVET and was admitted to NICU for 30 days from the labor room. Also, antenatal USG suggested aqueductal stenosis. The baby showed yellow discoloration of the skin, which was gradually progressive with the passage of high-colored urine since 30 days of life, and given DVET.

Physical examination revealed that the baby appeared unwell, irritable, and afebrile, with stable vital values. The infant was 41 cm long, weighed 1500 g, and had a head circumference of 29.5 cm. The infant had triangular facies, profound jaundice, and slight anemia. The head size was average. The anterior fontanel was diamond-shaped, open wide, and pulsatile. There was a patent posterior fontanel.

On general examination baby were HR-134/min, RR-50/min, Temp-98.1% and SpO_2-95.

On CNS examination, the baby was alert and active, pupils were reactive to light bilaterally, and ophthalmic evaluation revealed microphthalmia and mature cataract. The baby also had hypertonia and hyperreflexia. The baby had moderate hepatosplenomegaly, a smooth surface, firm consistency, and well-defined margins associated with mild splenomegaly.

Hematological testing revealed leukocytosis (14.7 k/μL) and anemia (8 g/dL). Blood culture confirmed the presence of *Staphylococcus haemolyticus* species, and liver function tests revealed a total bilirubin of 24.4 mg/dL, with direct and indirect bilirubin of 10.7 mg/dL and 13.7 mg/dL, respectively, and SGPT and SGOT of 38 μ/L and 156 μ/L. On the antenatal USG, hydrocephalus caused by aqueductal stenosis was discovered. Toxoplasmosis, rubella cytomegalovirus, herpes simplex, and HIV tested positive for toxoplasma using the ELISA method (IgM).

On eye examination:

- Right eye-microphthalmia with mature cataract
- Left eye-panuveitis with mature cataract

The baby was diagnosed with respiratory distress syndrome with hydrocephalus due to aqueductal stenosis.

USG brain suggests:

- Bilateral lateral ventricle, third ventricle, and fourth ventricle appear dilated with aqueduct effacement possibility of aqueduct stenosis with hydrocephalus.
- Posterior horn of lateral ventricles is more dilated than the anterior horn causing effacement of adjacent brain parenchyma.

Discussion

This preterm neonate, born at 34 weeks of gestation and weighing 1.5 kg, is facing several medical challenges.

Managing premature neonates with multiple co-occurring conditions, including small for gestational age (SGA), hydrocephalus, symmetrical IUGR, hyperbilirubinemia, aqueductal stenosis, and microphthalmia, requires a carefully orchestrated approach to achieve optimal results.

Analysis of respiratory distress in SGA neonates is still debatable. Due to the lungs' enhanced development, SGA children are typically thought to be in better shape than appropriate gestational age (AGA) infants of the same weight. This is essential for identifying potential problems in infants with SGA and AGA. However, every investigation shows that SGA newborns are actually no better than AGA infants for the same gestational age, same-sex, and

same race. According to several research, SGA newborns were significantly more likely to experience RDS, respiratory issues, and even mortality.[4] Additionally, SGA newborns that have respiratory distress during the neonatal period are prone to die and suffer from morbidity.[5]

Hydrocephalus is a condition characterized by the abnormal buildup of cerebrospinal fluid (CSF) in the cerebral ventricles. This accumulation can occur due to factors such as obstruction in the normal CSF flow, inadequate absorption into the venous system via the Pacchionian arachnoid granulations, or excessive production of CSF.[6]

Intrauterine growth restriction (IUGR) refers to a condition where the rate of fetal growth is below what is considered normal given the individual infant's growth potential. Infants may be diagnosed with IUGR if they exhibit signs of limited growth and malnourishment during their time in the womb and at the time of birth.[7] The most common cause of neonatal death was respiratory distress syndrome (44.4%).[8] It is because IUGR fetuses have reduced lung volume and impaired surfactant production, which increases their risk of developing RDS.

Elevated levels of bilirubin in the blood can lead to jaundice and, in severe cases, may cause bilirubin-induced brain damage (kernicterus). This, in turn, can lead to respiratory distress and make it difficult for the neonate to breathe.[9] In addition to the elevated bilirubin levels, premature neonates may also have a deficiency of surfactant, which helps to keep the lungs open, leading to the collapse of the air sacs (alveoli) and making it difficult to breathe.[10]

The cerebral aqueduct is a narrow 15 mm conduit that allows for CSF to flow between the third ventricle and the fourth ventricle.[11] Aqueductal stenosis can cause an increase in intracranial pressure, leading to hydrocephalus, which in turn can contribute to RDS. As mentioned previously, elevated intracranial pressure can compress the brainstem, which controls breathing, and make it difficult for the neonate to breathe. In such cases, prompt surgical intervention to widen the aqueduct may need to manage the aqueductal stenosis and prevent or treat respiratory distress. However, it is important to note that RDS is primarily caused by a deficiency of surfactant, which helps keep the lungs open, rather than by aqueductal stenosis.

In this case, prompt recognition and treatment of all challenge the child had crucial in ensuring a favorable outcome for the patient.

Conclusion

The preterm neonate with RDS has multiple health challenges including low birth weight, hydrocephalus, symmetrical IUGR, neonatal hyperbilirubinemia, and aqueductal stenosis. These conditions require specialized care and close monitoring to ensure the best outcomes for the neonate.

Clinical Significance

The case report "Overcoming Obstacles: A case report of preterm Neonatal RDS with Coexisting Hydrocephalus, Aqueductal Stenosis, and Neonatal Hyperbilirubinemia" holds immense clinical significance for several reasons. Firstly, the concurrent presentation of multiple complex health conditions in a preterm neonate is a challenging scenario. The report serves as a testament to the resilience of such patients and the critical role of interdisciplinary collaboration in ensuring their survival and well-being.

Moreover, the report provides a wealth of information and insights that can inform the practices of healthcare providers, including neonatologists, pediatricians, and other specialists, in the management of similar cases in the future. The report's documentation of the medical interventions, treatments, and outcomes can help healthcare providers identify best practices, improve patient outcomes, and advance the field of neonatology.

Finally, the reports contribution to the body of medical literature on preterm neonatal RDS, hydrocephalus, aqueductal stenosis, and neonatal hyperbilirubinemia, can inform future research and ultimately improve the care and quality of life for such patients.

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