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Non-Immune Hydrops Associated with Urogenital Sinus

Key Words

Nonimmune hydrops
Urogenital sinus

Abstract

Objective and Importance: To report an unknown association between nonimmune hydrops and a case of urogenital sinus. **Clinical Presentation:** A newborn girl presented with gross fetal and neonatal hydrops and was managed successfully. She was then extensively investigated for the cause of hydrops, and was only found to have a urogenital anomaly that was managed by temporary external drainage. **Conclusion:** Urogenital sinus anomalies may be added to the list of causes and associations of nonimmune hydrops fetalis.

Introduction

Nonimmune hydrops fetalis is associated with many types of diseases and conditions [1]. Urethral obstruction and congenital anomalies of the uterus and vagina are among the genitourinary conditions of associations.

Urogenital sinus is defined as a common channel into which both the urinary and genital tracts open. It is one of the rare congenital malformations that has a wide spectrum of urogenital anomalies with variable modes of presentation caused by a lack of development of the urogenital sinus and its derivatives [2, 3, 5].

We report a case of fetal and neonatal hydrops that was associated with urogenital sinus in a newborn girl.

Case Report

A 22-year-old woman was referred to the Maternity Hospital at 32 weeks of gestation with polyhydramnios. An ultrasound scan on the referral day showed fetal hydrops with the suspicion of hydronephrosis and hydroureter. She had normal pregnancies previously ending in 2 normal children. Because of the gross ascites and hydrops the mother underwent lower segment cesarean section to give birth to a baby girl weighing 3,660 g with gross hydrops. The baby's Apgar score was 2 and 6 at 1 and 5 min, respectively.

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Fig. 1. Voiding cystourethrogram of the patient showing the filling of the vagina with dye.

The baby was resuscitated and then transferred to the neonatal intensive care unit (NICU) for further treatment. In the NICU, the baby was managed successfully with aggressive ventilation, surfactant therapy and careful fluid management with colloids in the form of 12-hourly albumin infusion, followed by low-

dose furosemide. She remained stable with regard to the vital signs and basic hematological and biochemical investigations including the renal function test. She lost 1,400 g over 7 days and could be extubated on the 10th day of life. The baby had a lower abdominal mass that was palpable only when the ascites was cleared,

Fig. 2. a The urogenital sinus found in our case. **b** when it got obstructed. B = Bladder; R = rectum; S = symphysis pubis; U = uterus; URS = urogenital sinus; V = vagina.

and at that point the absence of the vaginal opening was detected.

The baby had the following investigations: (1) Abdominal ultrasound which revealed a resolving hydronephrosis and hydroureter and an abdominal mass that could be not clearly defined. (2) Findings in the CAT scan of the abdomen were not helpful. (3) Micturating cystourethrogram of the patient showed that the dye filled the bladder initially. Upon voiding, however, the mass could be identified as having a fistulous connection with the bladder but it was not definable. There was no vesicoureteric reflux but it showed the filling of the vagina with the dye upon micturition (fig. 1). (4) Cytoscopy was done under general anesthesia which showed a vaginal opening into the proximal urethra just below the bladder neck area. The opening was cannulated and urine was drained from the vagina confirming the diagnosis of urogenital sinus. A percutaneous temporary tube vaginostomy was then inserted.

The baby continued to do well in spite of one episode of *Pseudomonas* sepsis. She responded well to systemic antibiotics, and during her stay in the NICU she was additionally investigated for other known causes of hydrops fetalis. These included blood film examination, urine analysis, liver function tests, hemoglobin electrophoresis, immunoglobulin levels, viral studies, α -fetoprotein, and α_1 -antitrypsin levels, all of which proved to be normal. An echocardiogram showed a normal heart, and a chromosomal analysis showed a karyotype of 46 XX. The baby had cholestasis which disappeared along with the hydronephrosis-

and the hydroureter once the mass was decompressed. This was confirmed by ultrasound examination.

On follow-up, the baby had been doing well but the external vaginostomy needed dilatation. She had urinary tract infection that was treated, and the baby was commenced on prophylactic antibiotics. The renal function remained normal.

Discussion

In the embryonic development, the primitive hindgut, or cloaca, is divided by the urorectal septum into an anterior urogenital sinus and the dorsal hindgut, which becomes the rectum and the anal canal [4]. The urogenital sinus gives rise to the bladder and urethra, and the distal vagina.

In the female there are three types of urogenital sinus abnormalities resulting from defective development in this location: (1) vaginal obstruction or atresia, (2) persistent urogenital sinus abnormality and (3) persistent cloacal anomalies. Our case fits with the second variety [3] (fig. 2).

On the other hand, nonimmune hydrops remains one of the challenging conditions in diagnosis and treatment in perinatal medicine

[1, 2]. Several causes and associations have been reported, and in most of the associations the mechanism of the hydrops formation is not fully understood [1]. Among the genitourinary conditions are the congenital nephrotic syndrome, urethral obstruction and renal

dysplasia, polycystic kidneys, renal vein obstruction and congenital abnormalities of the vagina and uterus. In 50% of the cases the causes remain unknown, and poor prognosis is seen in up to 80% of the conditions.

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