CASE REPORT

HYDROMETROCOLPOS PRESENTING AS A HUGE ABDOMINAL SWELLING AND OBSTRUCTIVE UROPATHY IN A 4 DAY OLD NEWBORN: A DIAGNOSTIC CHALLENGE

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ABSTRACT

BACKGROUND: Abdominal swelling is an uncommon presentation in newborn babies. A combination of huge abdominal swelling, obstructive uropathy and imperforate hymen in newborns has not been reported in the medical literature.
CASE DETAILS: We report a 4 days old newborn with a rare presentation of hydrometrocolpos which posed a diagnostic challenge and consequently resulted in delays in diagnosis and treatment.
CONCLUSION: Hydrometrocolpos should be considered as a differential diagnosis in neonates who present with huge abdominal swelling.
KEYWORDs: Neonate, Abdominal Swelling, Hydrometrocolpos, Imperforate Hymen

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CASE REPORT

A 23 days old female neonate born to a 35 years old Para IV lady at gestational age of 38+6weeks was referred to the neonatal intensive care unit (NICU) of Hawassa University Referral Hospital from a nearby district hospital. Mother had antenatal followup and pregnancy was uneventful. Delivery was done by spontaneous vertex at a district hospital after 20hrs of labor; membrane was ruptured during labor. Her birth weight was 2500 grams; Apgar score was unknown but the baby cried immediately after delivery. Few minutes after delivery, the baby failed to suck and was admitted to newborn unitof the hospital. Vital signs were normal except for a depressed sucking reflex. Other systemic examination was non-revealing. Blood workup revealed a white cell count (WBC) of 22,400/$mm^3$and an absolute neutrophil count (ANC) of 13,216/mm^3. Based on these clinical clues, infection was suspected, and the newborn was started on Ampicillin (100mg/kg BID) and Gentamicin (5 mg/kg once daily). On the 4th postnatal day, a supra-pubic swelling and failure to pass urine were noticed. She was catheterized and 80cc of clear urine was evacuated. However, the mass did not decrease in size. Pelvic ultrasound revealed an intra abdominal cystic mass with bilateral moderate hydronephrosis.

Even though the neonate was referred on the 4th postnatal day to Hawassa University Hospital (HUH) NICU, the parents could not come immediately for social reasons and brought her on the 23rd postnatal day because of a progressively increasing abdominal swelling, failure to pass urine and difficulty of breathing. By then, her vitals were: pulse rate, 152/minute; respiratory rate, 82/minute; temperature, 37.2 Celsius. Weight, 2590 grams; length, 51 cm; and head circumference, 35 cm. She was noted to have emaciation, failure to thrive, tachypnia and labored breathing.A huge cystic abdominal mass with difficulty to delineate the lower border was appreciated. The genitalia looked grossly normal, but the hymeneal membrane was imperforate (noticed after the operation). Upon catheterization, 30cc of urine was evacuated.Repeat laboratory tests showed: WBC, 30,100/mm^3; ANC, 21,390/mm^3; hemoglobin, 15gm/dl; platelets, 412,000/mm^3; blood group/Rh, O/+; creatinine, 10.9 mg /dl; sodium, potassium and calcium within normal limits.

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Repeated ultrasound showed a distended fluid containing mass with echo-debris arising from pelvis with a conclusion of ascites, bilateral moderate hydronephrosis, and possibility of endometrial cavity abscess or adnexal cyst (Figure C). However, a third ultrasound suggested the possibility of teratoma. Since the abdominal swelling was noted to be huge and the imperforate hymen was noticed late and with the controversial ultrasound findings, decision was made to do exploratory laparatomy. Intra-operative findings were a 20x10cm distended uterus with the bladder adherent to the abdominal wall which caused inadvertent injury upon incising the abdominal wall. Uterine tubes, ovaries, bowel and peritoneum were all healthy. There was no free peritoneal fluid. Around 1250cc of whitish fluid with debris was drainedfrom the uterine cavity using a wide bore needle.

Following the procedure, the baby was persistently hypothermic with irregular breathing. Since infection was initially considered, antibiotics were continued; she was put on radiant warmer and was on oxygen. However, after 3 hours of the procedure, the baby started to gasp and was resuscitated which was not successful ultimately. The possible cause for death was uncontrolled sepsis and acute renal failure.

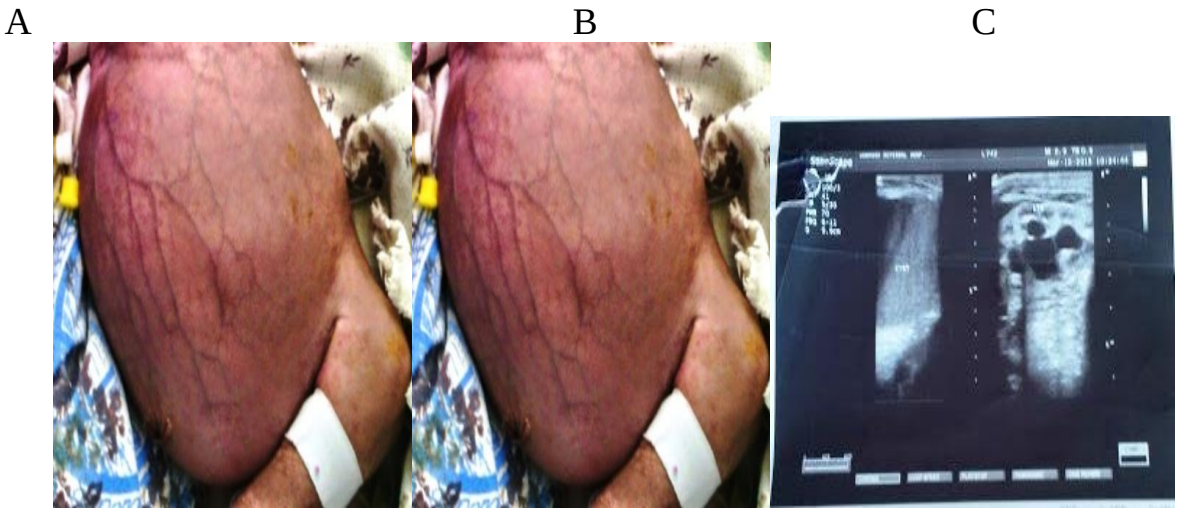

Figure 1:*A: Photo of the abdominal swelling on Day, 21 B: Photo of abdominal swelling on Day 23, C: Ultrasound examination on Day 21.*

DISCUSSION

Huge abdominal swelling is a relatively uncommon presentation for newborn babies. The common differential diagnoses to be considered in such babies include neuroblastoma, intra-peritoneal fluid collection following organ failure, ovarian cysts, intra-abdominal sacrococcygeal teratoma, mesoblastic nephroma, bowel duplication, genitourinary anomalies and anterior sacral meningocele (1). Congenital hydrometrocolpos as a cause of abdominopelic mass in neonates has also been characterized in few case reports in the past 2 decades (1-6). In the current case, the presentation with huge abdominal swelling with distended veins and compromised breathing (Figures A and B) prompted entertainment of various differential diagnosis and delayed the actual diagnosis of hydrometrocolpos which was made intra-operatively.

Hydrometrocolpos is a condition in which the uterus and vagina are distended by retained fluid other than blood in the presence of distal vaginal outlet obstruction. Secondary infection of hydrometrocolpos leads to pyometrocolpos (pyometra) (1,6). It is a very rare condition with incidence of 1 in 16,000 live births (7). The previously reported low incidence of hydrometrocolpos could be due to difficulty in

diagnosis and high mortality rates as suggested by Mittal et al (6). This difficulty in diagnosis was similarly observed in the present case. Associated anomalies may include obstructive congenital malformations of the genital tract such as vaginal atresia, transverse vaginal septum and imperforate hymen. McKusick–Kaufman syndrome, an autosomal recessive disorder characterized by vaginal atresia with hydrometrocolpos, week of gestation; the incidence is 0.0014–0.01 % in full-term newborns (1, 8). Previous authors described that hydrometrocolpos is usually diagnosed prenatally as the cause of abdominal cystic mass (4). However, in the present case, prenatal evaluations were negative, and abdominal swelling started to progressively enlarge after the 4th day when the swelling was first noticed. The fact that the presence of imperforate hymen was overlooked was the pitfall in the evaluation of this case. It resulted in major surgery under general anesthesia while it could have been enough to make incision on the imperforate hymen to drain the fluid.

The present case had obstructive uropathy and moderate hydronephrosis bilaterally. Previous authors described the possibility of hydrometrocolpos causing urinary stasis and acute renal failure due to obstructive uropathy (9). Because of the delay in diagnosis of hydrometrocolpos as a cause of the obstructive uropathy, the renal function was deteriorating progressively and contributed to the death of the baby.

Hydrometrocolpos is a possible cause of huge abdominal swelling in a newborn. Imperforate hymen and obstructive uropathy could be associated conditions. Clinicians should consider the possibility of hydrometrocolpos as a cause of huge abdominal swelling in newborns. The genitals should be carefully examined to save patients from major surgeries and related complications.

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polydactyly, congenital heart defects and non-immune mediated hydrops fetalis, has also been described (1,3).

Among the obstructive genitourinary anomalies, imperforate hymen has been described as the most frequent cause of hydrometrocolpos. It is due to failure of partial resorption of the membrane during the embryonic development and also the hymen failing to rupture during the eighth

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