

Case Report

Early Diagnosis and Successful Treatment of Congenital Huge Hydrometrocolpos Secondary to Low Transverse Vaginal Septum with Obstructive Symptoms

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Abstract: Obstructive congenital anomalies of the female reproductive tract are rare and usually noticed during adolescent period for failure to see menses with cyclic abdominal pain, abdominal mass and local compressive symptoms. It is very rare for such cases to be symptomatic during early childhood from mucous collection. Congenital hydrometrocolpos (an accumulation of watery fluid in the uterus and vagina) that occurs during fetal period is a very rare condition, only with some case reports. The diagnosis is challenging and usually made late which delays the management resulting poor outcome from local compressive symptoms. We present a case diagnosed with huge congenital hydrometrocolpos secondary to low transverse vaginal septum using ultrasound by experienced radiologist and meticulous genital examination in a 5 day old neonate who had abdominal distention and difficult to pass urine since birth where incision of the septum transvaginally and drainage of the fluid was done to relieve symptoms with successful outcome.

Keywords: Congenital (Hydrometrocolpos), Transverse Vaginal Septum, Fetal Cystic Abdominal Mass, Neonatal Abdominal Mass with Obstructive Uropathy

1. Introduction and Literature Review

Congenital anomalies of the female reproductive system are rare conditions which arise from failure of fusion of the mullerian duct or failure of resorption. Obstructive genital tract anomalies usually manifest after age of menstruation with cyclic abdominal pain, lower abdominal swelling and local urinary or rectal compressive symptoms from the hematocolpometra. [1] Rarely they can be manifested in childhood from collection of mucous in the vagina and present with compressive symptoms. Hyperstimulation of the cervical glands by maternal hormones during fetal and early neonatal period could result in production of excessive cervical discharge and accumulation fluid in the vagina and/ or uterus giving a condition called hydrometrocolpos. Congenital

hydrometrocolpos which usually occurs from imperforate hymen and rarely from distal vaginal atresia is a very rare condition with significant diagnostic challenge resulting in late diagnosis which delays management and increases mortality. [2] Antenatal diagnosis needs high index of suspicion by experienced radiologist using an ultrasound or MRI. Meticulous neonatal evaluation could help early diagnosis and treatment of such cases. The aim of treatment is distal vaginal drainage, which can be achieved by a perineal procedure in most cases. Laparotomy is indicated only in cases of high vaginal atresia, which require a vaginal pull-through procedure. [3]

2. Case Report

2.1. History

A 5 day old 3.5 kg female neonate born from a 28 years old primipara lady at gestational age of 38weeks and 7days delivered spontaneously through vagina after 12 hours of labor with an Apgar score of 8&9 in Gondar university hospital, North west Ethiopia, was admitted to NICU with abdominal distention. She had failure to pass urine for 3 days and difficulty of defecation for which she was catheterized. But despite passing urine through the catheter there was no decrement in the abdominal distention. An abdominal ultrasound was done by an experienced radiologist and reported to have hydrometrocolpos for which gynecologic consultation was made. In the last ANC visit after an obstetric ultrasound was done the mother was told to have a fetus with distended bladder due to possible outlet obstruction with no further evaluation.

2.2. Physical Examination

She is healthy looking with no gross anomaly or dysmorphic feature. The Vital signs are normal. She has grossly distended abdomen with visible veins draining upwards, and a mobile and non tender cystic abdominopelvic mass reaching to the level of umbilicus (figure 1). The anus is patent with the mass felt down to the perineum through it.



Figure 1. Photograph of the abdomen of the patient showing distended abdomen with distended superficial veins.

She has a normal female type external genitalia, annular hymen. A low transverse vaginal septum is seen bulged during straining for defecation.



Figure 2. External Genital appearance.



Figure 3. Annular hymen seen.

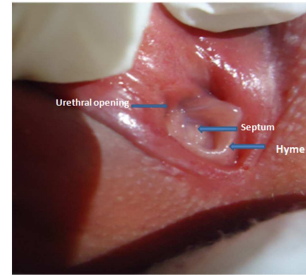


Figure 4. Low transverse vaginal septum seen just behind the hymen bulged during straining.

2.3. Investigations

U/S = Both kidneys are have normal size and echo pattern with no hydronephrosis. Empty urinary bladder, hugely distended vaginal vault with echodebrinous fluid extending to the endometrial uterine cavity effacing the cervix to 6.2cm. (10.4cm by 5.8cm) with an ultrasound diagnosis of hydrometrocolpos 2o to imperforate hymen ddx distal vaginal agenesis.

Trans perineal ultrasound = distance from perineum to the mass is 0.5cm.



Figure 5. Abdominal ultrasound image showing hydrometrocolpos with the vagina on the right side.



Figure 6. Transperineal ultrasound showing the distance from the perineum to the fluid.

2.4. Diagnosis

Abdominopelvic mass secondary to congenital hydrometrocolpos secondary to low transverse vaginal septum with compressive urinary and bowel symptoms.

2.5. Treatment

Consent was taken from the mother, under anesthesia urethra was catheterized, the transverse septum grasped at two sites with artery forceps and opened. About 250 ml of thin fluid was drained, #12 Foley catheter was inserted for continuous drainage and to prevent closure for three days.

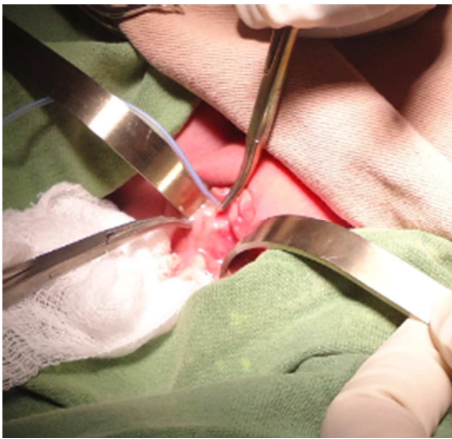


Figure 7. Incision procedure with a pediatric NG tube in the urethra.



Figure 8. #12 Foley catheter inserted through the incised septum.



Figure 9. Abdominal photograph after drainage.

3. Outcome and Follow up

She started passing urine and stool without difficulty. On third post procedure day transabdominal ultrasound was done showing collapsed thin and long uterus with no fluid in it. Patient discharged with advices for possible evaluation when adolescent. Analysis of the fluid showed no organism.



Figure 10. Transabdominal ultrasound showing empty and elongated uterus (5 x 2.1cm) on third post procedure day.

4. Discussion

Hydrometrocolpos though a rare condition, the incidence is increasing from time to time because of increased rate of diagnosis. [4] The etiology of hydrometrocolpos is as simple as imperforate hymen to the most severe cloacal malformations. [5] It is a serious condition which can be complicated with compressive symptoms, bilateral hydronephrosis and renal failure and can be superinfected resulting sepsis and death. [6-9] It can be associated with peritonitis from bowel perforation due to compression or fetal ascites. [10, 11] Diagnosis is challenging which delays management. [12, 13] Ultrasound and MRI are useful studies in the diagnosis. [14, 15] The presence of fetal cystic abdominal mass on routine ultrasound scanning should raise the possibility of the problem and MRI should be done. [16, 17] In neonates presenting with abdominal masses and compressive urinary or bowel symptoms, hydrometrocolpos should be suspected and radiologic evaluations should be done timely. [18] Difficulty of diagnosis resulted in unnecessary laparotomy and some were identified only after autopsy. [7, 19] Gentle evaluation of the genital tract could easily identify the obstructing membrane especially when it is caused by imperforate hymen, and low transverse vaginal septum. Careful incision and drainage of the fluid is a good option in the management of such cases.

5. Conclusion

Though congenital hydrometrocolpos is a very rare condition with fatal complications, in female fetuses with cystic abdominal mass and in neonates with abdominal mass and obstructive features, it should be strongly considered. An ultrasound by experienced radiologist with meticulous genital evaluation could be enough in resource limited areas to early diagnose the problem. Timely identification and surgical treatment with incision of the obstructing septum transvaginally is a simple and life saving procedure.

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