

Congenital vaginal obstruction with intrauterine hydrometrocolpos: A case report

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Abstract

Intrauterine hydrometrocolpos is a rare condition caused by congenital vaginal obstruction. Imperforate hymen is the most common cause and manifests as a huge abdominopelvic mass. Ultrasonography is an invaluable tool in screening and evaluation of fetal intra-abdominal mass but even with serial ultrasonography it may be very difficult to delineate the exact site of origin and come to a diagnosis. MRI is the most reliable and sensitive modality to evaluate complex malformations/ masses. In a female child born with cystic abdominopelvic mass a thorough neonatal examination after birth focusing on the urogenital system and the extremities will give a significant clue in diagnosis and subsequent management of hydrometrocolpos. Here we present a case of intrauterine hydrometrocolpos which remained in diagnostic dilemma until the postnatal period.

Key words: complex malformations, congenital vaginal obstruction, hydrometrocolpos

Introduction

Failure of development of vagina (agenesis) affects one in 4 to 5 thousand female births.¹ Imperforate vagina is caused by the failure of cells to degenerate in the center of the vaginal plate and by the failure of the vaginal canal to open into the urogenital sinus while an imperforate hymen is caused by the failure of cells to degenerate in the lower part of the vaginal plate and the wall of the urogenital sinus.²

The three main types of congenital vaginal occlusions that lead to hydrometrocolpos include imperforate hymen, transverse vaginal septum and segmental vaginal atresia. In the rare segmental atresia, there is a thick fibrous tissue obliterating a segment of vagina while in transverse vaginal septum, a fibroepithelial membrane is present most often in the mid-portion of the vagina. We describe a case report.

Case report

Twenty-one years old (Gravida 1, Parity 0) underwent routine obstetrical ultrasound at 34 weeks of gestation. Sonogram revealed a large cystic mass of approximately 5.7 x 5.2 cm in dimension occupying the fetal pelvis and the lower abdomen, displacing the urinary bladder anterosuperiorly. The mass showed low-level internal echoes and peripheral, lobulated and echogenic component. Both kidneys were normal and there was no hydro-ureteronephrosis. Provisional diagnosis of dermoid cyst was made ultrasonographically (Fig.1). Review of prior routine screening scan performed at 18th weeks of gestation showed normal anatomy of the fetus.

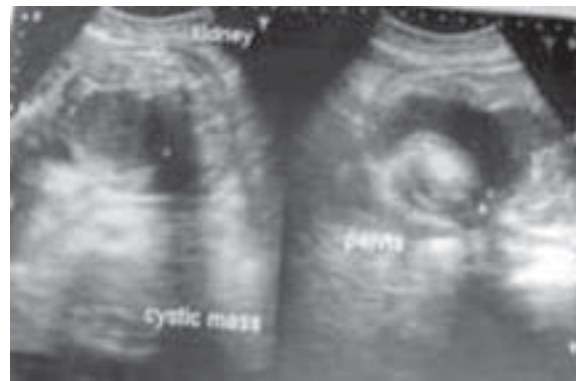


Fig.1 Transverse and sagittal antenatal ultrasound scans at 34 weeks of gestation showing a complex cystic mass in fetal pelvis. A diagnosis of dermoid cyst was given.

A repeat sonogram was performed a week later to reevaluate the fetus and to review the diagnosis. Though a detailed evaluation of the urinary bladder was difficult it appeared to be flattened and displaced anteriorly. A large smooth tubular/ cystic mass extending from pelvis to the upper abdomen was noted with mild wall thickening, suggesting bowel origin of the mass. Thus a diagnosis of dilated rectum with co-existent ano-rectal malformation was made. Scanning of the rest of the fetal parts revealed no abnormality .

Labor was induced at 35+2 days of gestation and a normal vaginal delivery with episiotomy was possible with undesired mild intrapartum eclampsia that complicated the event. A live, female baby of 2 kgs, with an Apgar score of 2/10, 8/10 was delivered. CNS reflexes were intact and normal. Local

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examination revealed a large and compressible, cystic mass in the perineal region increasing in size during bouts of cry. Anal opening was intact. Laboratory parameters were within normal range except for mild hyperbilirubinemia.

Ultrasound scanning of the neonatal abdomen was carried out immediately after delivery. A large cystic abdominopelvic mass without clear delineation of its origin was re-noted. MRI was then considered for further evaluation. A large, pear shaped, cystic abdominopelvic mass with low signal intensity in T1WI and high signal intensity in T2WI was seen to be extending inferiorly into the perineal region. Urinary bladder was separately identified anterior to the cystic mass. Lumbar and sacral spines were normal and rectum was identified between the cystic mass and sacrum. Diagnosis of hydrometrocolpos was suggested (Fig. 2). A detailed physical examination was then carried out by the neonatologist, which clarified the abnormality.



Fig.2 T1 and T2 weighted Sagittal MRI Scan of the neonate showing a large fluid filled uterus and vagina.

The baby became febrile after few days of conservative observation. Staphylococcus aureus was isolated in blood culture. Pediatric surgeons were then called in for surgical intervention. Approximately, 1500 ml thin fluid, mixed with pus, gushed out in pressure after a small incision was placed in the hymen. A drain tube of 7 Fr was then kept in situ for the subsequent drainage.

Cytology report from the fluid sample revealed plenty of inflammatory cells ruling out the possibility of dermoid cyst. Culture and sensitivity of the same fluid didn't show any growth within 24 hours. A week post- drainage, rescanning was done which revealed slightly bulky but normal uterus with the drain tube inside the uterine cavity. The cystic mass had completely disappeared (Fig 3).

Comment

Imperforate hymen is the simplest of these lesions.² It may manifest as a huge abdominopelvic mass due to retained fluid and mucus. Vaginal obstructions are usually diagnosed in the new born as intrapelvic mass lesions, hydrocolpos in the newborn and haematocolpos at puberty.¹

In some cases, fetal urination into the uterus and vagina occurs which causes their dilatation. This is because of a common urogenital sinus. The result may be proximal urinary tract dilatation due to pressure effect. Sometimes, there may be a rectovaginal communication on the dorsal wall of the lower vagina above the urogenital sinus, allowing meconium to mix with urine. If there is communication between rectum and uterus/vagina, the malformation is then referred to as the cloacal form of imperforated anus.²

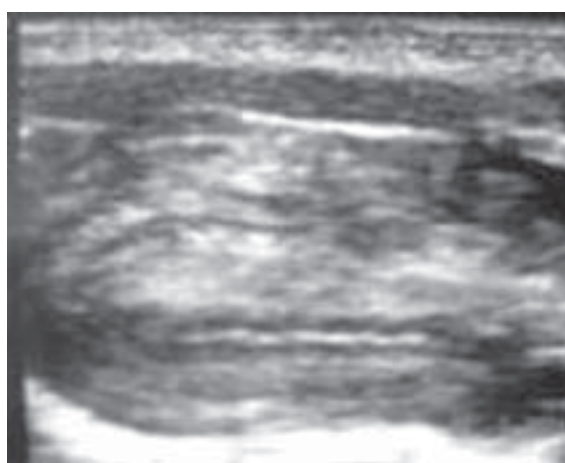


Fig. 3 Sagittal ultrasound scan of the neonatal uterus after drainage

When a large cystic abdominopelvic mass is noted in the fetus during ultrasonography, one should not be confused with uterine – vaginal swelling, a self-limiting condition receding rapidly after birth attributed to maternal estrogen stimulation. *McKusick – Kaufman syndrome* (MKKS) and *Bardet Biedl syndrome* (BBS) are associated with congenital hydrometrocolpos. Some affected infants may have finger and toe anomalies as part of a genetically determined syndrome. *MKKS* is a rare, recessively inherited condition, reported mainly in young children characterized by vaginal atresia with hydrometrocolpos, postaxial polydactyly and congenital heart defects.³ *MKKS* was first delineated by McKusick et al in 1964 in two Amish siblings and rapidly confirmed over 60 cases have now been reported and autosomal recessive inheritance is clearly established.⁴ Cardinal features of *MKKS* are hydrometrocolpos and polydactyly and it is often reported as the “hydro-metrocolpos polydactyly syndrome”.^{5,6}

BBS is another recognized but heterogeneous group of autosomal recessive disorder characterized by retinal dystrophy or retinitis pigmentosa (appearing usually between 10 and 20 years age), post axial polydactyly, obesity, nephropathy, mental disturbances or occasionally mental retardation.^{7,8}

In our case, there was no evidence of upper urinary tract dilation, anal opening was patent and the extremities including the finger and toes were normal. This ruled out the possibility of syndromes associated with hydrometrocolpos.

Conclusion

Ultrasonography is an invaluable tool in screening and evaluation of fetal intra-abdominal mass, however; exact delineation of its origin may be difficult even in the postnatal period. Currently, MRI is the most reliable and sensitive modality to evaluate complex neonatal intra-abdominal malformations/ masses and determine their site of origin. Suspicion of hydrometrocolpos should be made when a large cystic abdominopelvic mass is noted in the fetus during ultrasonography and the scan should be directed to detect any associated fetal congenital anomaly, especially of the genitourinary system, extremities, and the heart. Appropriate antenatal delivery and intervention protocols should then be formulated for a desired fetal outcome.

In a developing nation like Nepal where antenatal ultrasound is not widely available, a thorough neonatal examination after birth focusing on the external genitalia may provide a significant clue in diagnosing and subsequent management

of hydrometrocolpos in any female child born with abdominopelvic lump.

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