Giant fetal hydrometrocolpos associated with cloacal anomaly causing postnatal respiratory distress

Full Title: Giant fetal hydrometrocolpos associated with cloacal anomaly caused postnatal respiratory distress

Running Title: Cloacal anomaly with respiratory distress

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Abstract

Persistent cloaca is a rare presentation wherein the urethra, vagina, and rectum converge into a common channel with a single perineal opening. Fetal hydrometrocolpos can result if fluid accumulates behind an obstruction of this common channel. A 29-year-old woman (G4P1021) was referred to us at 36 2/7 weeks of gestation for evaluation of a fetal abdominal cystic mass. Detailed ultrasonography and magnetic resonance imaging revealed 2 symmetric cystic masses, bilateral hydronephrosis, and oligohydramnios. Elective cesarean delivery was performed at 37 0/7 weeks; the baby weighed 4,043 g with Apgar scores of 5 and 6 at 1 and 5 minutes. Intubation was performed for respiratory distress, and the infant was noted to have an imperforate anus; persistent cloaca was diagnosed. Drainage of the hydrometrocolpos improved the infant’s breathing remarkably, and extubation was achieved. This child's imaging findings are among the largest ever reported and resulted in neonatal respiratory distress.

Key words: cloaca, cloacal malformation, persistent cloaca, hydrometrocolpos, prenatal diagnosis

Introduction

Persistent cloaca is a rare condition that only occurs in female patients. The urethra, vagina, and rectum converge into a common channel that has a single perineal opening.[1] Fetal hydrometrocolpos may result from a persistent cloaca if fluid accumulates behind an obstruction of this common channel during the prenatal period.[2] Here we report a patient with a giant hydrometrocolpos and severe oligohyramnios secondary to a cloacal anomaly; the patient experienced postnatal respiratory distress.

Case

A 29-year-old woman, gravida 4 para 1 abortus 2, was referred to our hospital at 36 2/7 weeks of gestation for evaluation of a fetal abdominal cystic mass. This mass was first detected at 35 6/7 weeks of gestation during routine fetal ultrasound examination; 2 weeks prior, there had been no such mass visible on ultrasonography. The patient was healthy with an unremarkable medical history. Detailed ultrasonography revealed 2 symmetric cystic masses (both masses together measuring 106 × 94 × 70 mm), bilateral hydronephrosis, and oligohydramnios (Figure 1a). The external genitalia were consistent with female sex, and there were no apparent morphologic abnormalities at other sites. Magnetic resonance imaging (MRI) showed that each cystic mass had an attached tubular structure, and the urinary bladder was located anterior to both masses (Figure 1b). Although the fetal status was reassuring on cardiotocography, severe urinary obstruction was assumed based on the bilateral hydroureteronephrosis and oligohydramnios. According to the ultrasonography and MRI imaging, we made the antenatal assessment of ureterovesical

junction obstruction. To preserve renal function, elective cesarean delivery was performed at 37 0/7 weeks of gestation. The infant weighed 4,043 g, and abdominal swelling was observed (Figure 1c). The Apgar scores were 5 and 6 at 1 and 5 minutes. Intubation was performed for respiratory distress. The neonatologist diagnosed an imperforate anus and suspected a persistent cloaca. Neonatal computed tomography findings were almost identical to those of the antenatal MRI, revealing a dilated vagina and a duplicated hydrometrocolpos. Drainage of the hydrometrocolpos and a transverse colostomy for the imperforate anus was performed. At laparotomy, the infant was confirmed to have a duplicate vagina and uterus, and yellow transparent ascites suspicious for urine. Genitography revealed a dilated vagina and rectum communicating with a single common channel. The infant's breathing was remarkably improved after drainage of the hydrometrocolpos, and extubation was achieved on the second postoperative day. The infant's subsequent course was favorable.

Discussion

In this patient, we observe the typical imaging findings of persistent cloaca with vaginal duplex after the onset of common-channel obstruction. The cloaca is formed when the embryo is at 4-5 weeks of development and divides into the urogenital sinus and anorectal sinus.[1] If something prevents this stage of development, the division is not successful, resulting in the fusion of these channels.[3] Fetal persistent cloaca is generally diagnosed by the presence of hydrometrocolpos and hydronephrosis resulting from obstruction of the common channel.[4-6] The diagnosis is usually made in the early-30 weeks of gestation.

Our patient exhibited typical findings of persistent cloaca: duplicate cystic masses in the pelvis. Extremely distended abdomen caused by cloaca anomalies raises an alert over acute respiratory distress for a new born baby.

A symmetric mass with a tubular structure is an important finding that indicates vaginal duplex; antenatal MRI can be extremely helpful to evaluate this further.[7-10] Although it is difficult to detect the anatomical structure of the anus on ultrasonography in the setting of oligohydramnios, imperforate anus is a key finding of persistent cloaca.[11] If the fetus is female and an abdominal cystic mass is observed, it is useful to check the anal sphincter and mucosa by ultrasonography.[12] Hayashi et al. noted that the prognosis of patients with persistent cloaca is favorable unless there are associated deficits and hypoplastic lungs.[7]

In our patient, we assumed that the oligohydramnios occurred at 33 weeks of gestation or later, given the history of a normal prenatal ultrasound at that time, and the fact that no hypoplastic lung was seen on antenatal MRI. We speculated that the fetal lung had already maturated and expected a favorable neonatal condition; however, the infant exhibited respiratory distress. Although there is a possibility that oligohydramnios could have affected the neonatal respiratory condition, compression of the lungs by hydrometrocolpos was deemed the most likely cause. A large size cystic mass (> 10 cm) with oligohydramnios might lead to neonatal respiratory distress. Compared to oligohydramnios developing in the second trimester, which induces pulmonary hypoplasia, oligohydramnios developing in the third trimester less affects pulmonary hypoplasia. In contrast, oligohydroamnios in the third trimester may indicate severe obstruction of the common channel with the development of a larger cystic mass and

severe hydronephrosis, which may cause postnatal respiratory distress due to compression of the chest (Table 1).

This hypothesis was verified when the respiratory distress improved rapidly after drainage of the dilated hydrometrocolpos. Respiratory distress secondary to a marked abdominal distension can complicate the postnatal course of cloacal anomaly, precise prenatal diagnosis is extremely important for the appropriate managements.

Disclosure

The authors declare that there is no conflict of interest regarding the publication of this paper.

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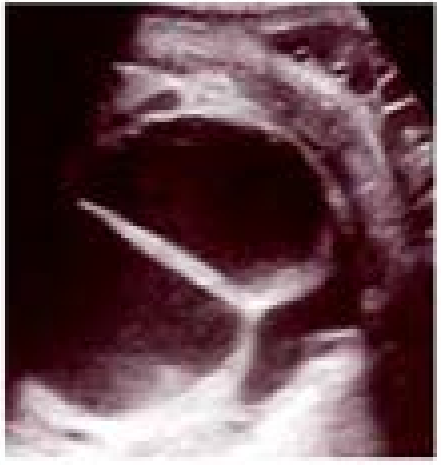
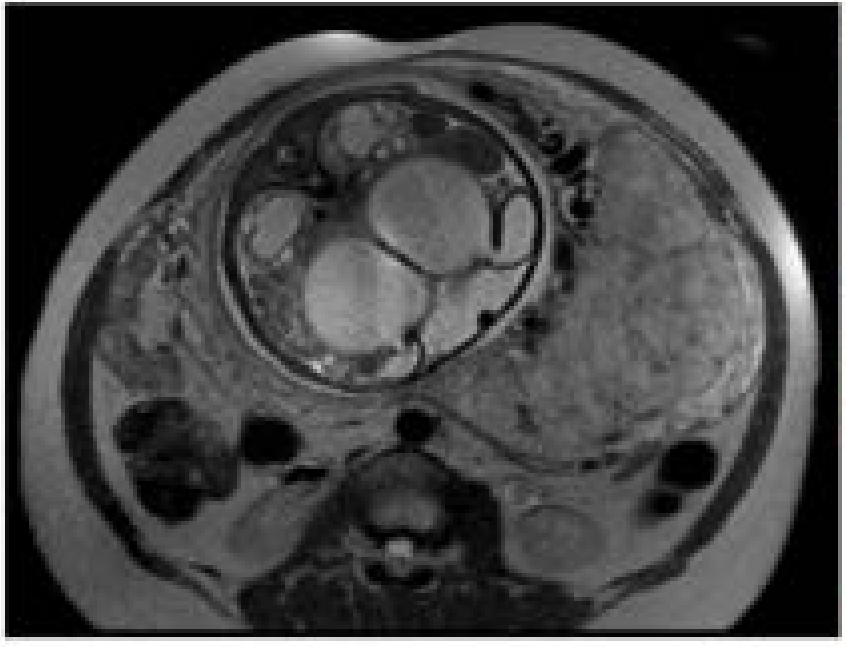
Figure legends

Figure 1. (a) Ultrasonography of the fetus at 36 week's gestation revealed 2 symmetric cystic masses (both masses together measuring 106 × 94 × 70 mm), bilateral hydronephrosis, and oligohydramnios. (b) MRI at 36 weeks' gestation revealed bilateral cystic masses in the pelvis. (c) Neonatal appearance with distended abdomen.

Table 1. Five cases of cloacal anomalies with or without respiratory distress



a)



b)

c)

Case	Time of diagnosis	Size of cyst	Other prenatal abnormality	Neonatal complications and prognosis	References
1	33 weeks gestation	**10** x 7.3 x 8.7 cm Double vagina	Bilateral hydronephrosis **Oligohydramnios** Ambiguous genitalia Single umbilical artery	Laparotomy performed on the 2nd day of life Died on the 27th day of life due to infectious complications	9
2	33 weeks gestation	**10 x 10 cm** Double vagina	**Oligohydramnios** Hydronephrosis Single umbilical artery Left ear deformity	Delivered by elective caesarean section at 36 weeks gestation **Intubated due to respiratory distress** Died on the 2nd day of life during surgery	6
3	32 weeks gestation	**10.1** x 6.3 cm Single vagina	Bilateral hydronephrosis	Delivered by elective caesarean section at 36 weeks gestation Laparotomy performed on the 3rd day of life Favorable prognosis	10
4	35 weeks gestation	7.7 x 7.6 x 7.3 cm Double vagina	Bilateral hydronephrosis	Delivered by emergency caesarean section at 38 weeks gestation due to arrest of labor Laparotomy performed on the 1st day of life Favorable prognosis	7
5	35 weeks gestation	**10.6** x 9.4 x 7 cm Double vagina	Bilateral hydronephrosis Single umbilical artery Ascites **Oligohydramnios**	Delivered by caesarean section at 37 weeks gestation **Intubated due to respiratory distress** Laparotomy performed on the 1st day of life Favorable prognosis	Current case

Table 1. Five cases of cloacal anomalies with or without respiratory distress