Congenital Imperforate Hymen with Hydrocolpos Diagnosed Using Prenatal MRI

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Imperforate hymen is a rare genital anomaly in which a layer of epithelized connective tissue that forms the hymen has no opening and completely obstructs the vaginal introitus. Hydrocolpos and hydrometrocolpos may occur secondary to this condition. Imperforate hymen usually does not cause symptoms until puberty [1]. This anomaly manifests as an abdominal mass that is detectable during the prenatal period only rarely [2].

We present a case of bilateral hydroureteronephrosis caused by hydrocolpos in a female fetus with an imperforate hymen that was diagnosed using prenatal MRI.

Case Report

A 35-year-old woman, gravida 2, para 1, underwent prenatal sonography examination because the results of a serum triple-marker screening test were positive (1/130). For Down syndrome screening, a risk of more than one in 270, the midtrimester risk of a woman who is 35 years old, is used as the cutoff for performing prenatal sonography. The result of amniocentesis came back as 46, XX, and no chromosomal abnormalities were detected.

Prenatal sonography of a female fetus was performed at both 18 and 22 weeks of gestation. Sonography showed bilateral hydronephrosis, megacystis, and polyhydramnios. The initial interpretation of the perinatologist was megacystis-microcolon-intestinal hypoperistalsis syndrome.

For these findings to be evaluated further and to be validated, prenatal MRI was performed at 22 weeks of gestation. The mother was in a supine position for the examination, and the images were acquired during maximal inspiration. The fetal urogenital system was examined in sagittal, transverse, and coronal orientations using a T2-weighted sequence (HASTE; TR/TE, 1,380/103; field of view, 250 mm; slice thickness, 4 mm; matrix, 256 × 256; number of excitations, 1). MRI showed bilateral hydronephrosis, megaureters, and a huge cystic mass extending from the mid abdomen to the perineum (Figs. 1A and 1B). The bladder was visualized anterior to the cystic mass (Fig. 1C).

Visualization of the small bladder ruled out megacystis-microcolon-intestinal hypoperistalsis syndrome. MRI findings of the cystic pelvic mass suggested imperforated hymen with hydrocolpos. There were no accompanying congenital anomalies. The differential diagnosis of a pelvic cystic mass in a fetus includes sacrococcygeal teratoma and anterior meningomyelocele, both of which were ruled out on the basis of the characteristic MRI appearance of imperforate hymen with hydrocolpos and the extension of the lesion.

The infant was born at 38 weeks' gestation by cesarean delivery. The Apgar scores was normal, and a thin protruding vaginal membrane and abdominal distention were noted on physical examination. Postnatal sonography showed bilateral hydroureteronephrosis and a huge cystic mass with heterogeneous interior echo in the pelvis and lower abdomen (Fig. 1D).

Voiding cystourethrography revealed a small bladder displaced to the anterior abdominal wall due to the mass effect of the hydrocolpos. Neither reflux nor ureteroceles were seen. The patient was referred to pediatric surgery. Hymenotomy was performed and caused discharge of 200 mL of clear serous and mucoid fluid (Fig. 1E). The infant died on the fifth postnatal day due to urosepsis.

Discussion

Congenital imperforate hymen is an external urogenital anomaly that usually presents in infancy and early childhood. Although it is a congenital anomaly, imperforate hymen diagnosed during the antenatal period is very rare because it seldom causes hydrocolpos or

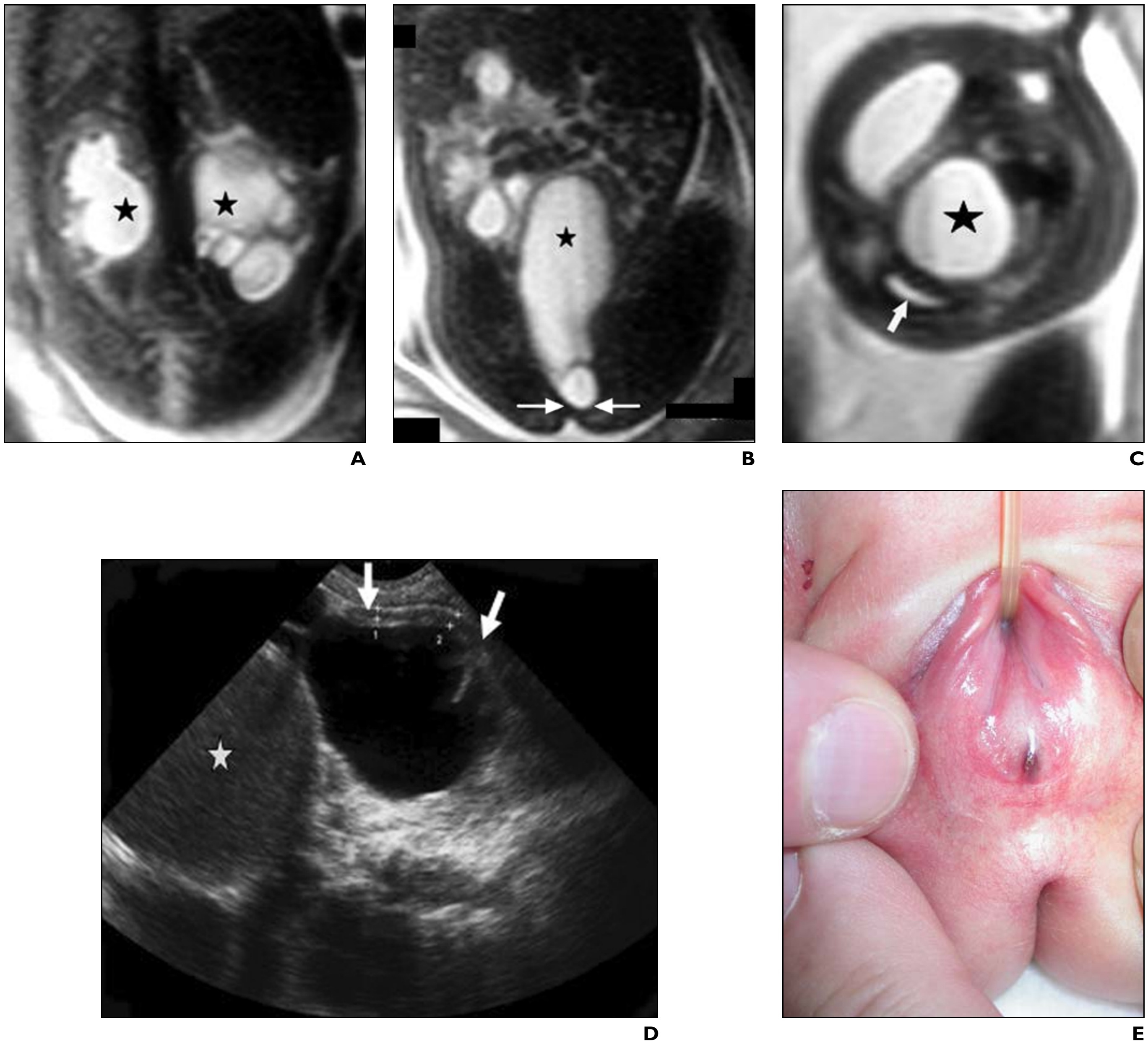

Fig. 1—Prenatal T2-weighted sequence (HASTE; TR/TE, 1,100/120; field of view, 250 mm; slice thickness, 4 mm; matrix, 256 × 256; number of excitations, 1) and postnatal sonography and physical examination findings.
A, Coronal image shows bilateral hydroureteronephrosis (*stars*).
B, Sagittal T2-weighted image reveals huge cystic mass (*star*) that extends from mid abdomen to perineum (*arrows*).
C, Axial T2-weighted image shows bladder (*arrow*) anterior to cystic mass (*star*).
D, Postnatal sonogram shows huge cystic mass (*star*) with internal echoes and thin cortex of left kidney (*arrows*) with dilated collecting system.
E, Photograph obtained during postnatal urogenital physical examination shows thin protruding membrane.

hydrometrocolpos in utero. In the literature, sonography has been the imaging technique of choice for the prenatal diagnosis of imperforate hymen with hydrocolpos [3].

The association of an imperforate hymen with other genitourinary anomalies is well known, and the investigation for associated anomalies is almost always performed by sonography. Although sonography is useful for evaluating fetal anatomy and showing abnormalities, its capability in detecting fetal abnormalities decreases if the mother is obese or has oligohydramnios and when the fetus is in certain positions [4].

MRI has become an alternative and complementary method for some of the equivocal prenatal cases owing to the recent developments in MRI technology. It provides excellent anatomic detail and soft-tissue contrast with multiple reconstruction planes and a large field of view. In the past, fetal motion

and longer acquisition times limited the role of MRI in fetal imaging. Ultrafast MRI has overcome these problems, so MRI has become more valuable in this setting and its role is still expanding [5].

In the present case, prenatal sonography failed to yield findings for a diagnosis, so MRI was ordered. MRI showed the exact location and extension of the cystic mass from the mid abdomen to the perineum in addition to the findings seen on sonography. The bladder was clearly shown on MRI as well. The cystic mass was diagnosed as hydrocolpos because it extended to the perineum and because a bladder abnormality could be excluded. The differential diagnosis of a pelvic cystic mass includes sacrococcygeal teratoma and anterior meningomyelocele. The sacrococcygeal teratoma mostly contains internal septations and solid components and has an extrafetal extension. Anterior meningomyelocele characteristically extends to the spinal canal and usually has dysraphism findings [6, 7].

The prenatal MRI diagnosis was also confirmed at physical examination, which revealed a thin protruding vaginal membrane, and by postnatal sonography. In retrospective analysis, we thought that the bladder could not be visualized on sonography because of the compressive effect of the huge cystic mass.

In conclusion, we suggest that MRI should be used as an alternative method to sonography in equivocal prenatal cases of imperforate hymen with hydrocolpos and that MRI is a more useful tool than sonography for the investigation of associated anomalies.

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