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Fetal hydrometrocolpos

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ABSTRACT

This report describes a case of hydrometrocolpos presenting antenatally as a cystic mass arising from the pelvis of a female fetus. The baby was born at 33 weeks and died within an hour due to pulmonary hypoplasia. Postmortem examination demonstrated hydrometrocolpos, dense adhesions in the abdomen and pulmonary hypoplasia.

INTRODUCTION

Hydrometrocolpos is distension of the uterus and cervix by non-hemorrhagic fluid. The post-natal diagnosis and treatment have been well documented[1–3]. We describe the appearances on prenatal ultrasound scanning in a case of hydrometrocolpos.

CASE REPORT

A 39-year-old Caucasian mother who had three previous normal pregnancies and delivered healthy babies had an uneventful antenatal course during her fourth pregnancy until 32 weeks' gestation. Although an ultrasound scan at 19 weeks had been reported as normal, a follow-up scan at 32 weeks demonstrated right renal agenesis, moderate left hydronephrosis and a cyst arising from the pelvis and extending to the level of the left kidney. The cyst was separate from and posterior to the bladder, and funnelling was seen at the lower end (Figure 1), similar to that found in male fetuses with posterior urethral valves; however, this fetus was female. There was hyperechogenic sediment in the cyst, and a possible cloacal abnormality was suggested. The amniotic fluid volume was normal.

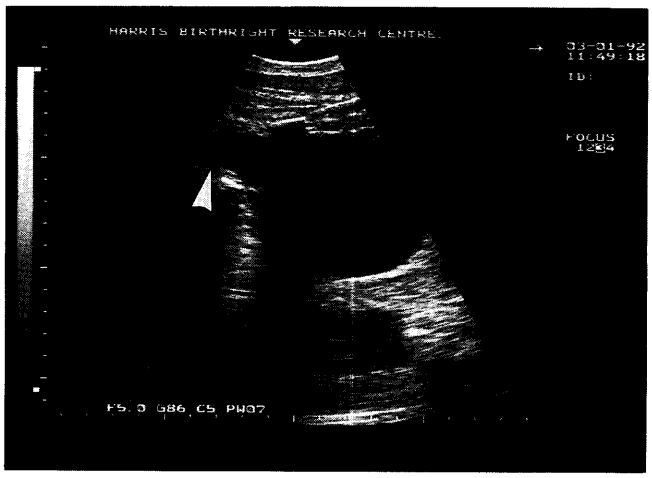

Figure 1 Ultrasound image of the lower abdomen in a female fetus showing a cyst with characteristic funnelling (arrow) arising from the pelvis

Spontaneous premature labor at 33 weeks led to an uncomplicated vaginal delivery of a normal-looking girl weighing 2300 g. The Apgar scores were 1 and 7 at 1 and 5 min, respectively. The neonate had severe respiratory distress and at 14 min following an apneic episode, assisted ventilation was instituted. Ventilation proved difficult and at 40 min the infant had a cardiac arrest, from which she was resuscitated. External examination revealed a distended abdomen containing a cystic mass arising from the pelvis. A urethral catheter was introduced, but despite passage of urine and some relaxation of the abdominal wall, the mass persisted. At 50 min of age there was irreversible cardiac arrest.

At autopsy, external examination revealed normal facial features, limbs and digits. The perineum showed a short median raphe, perforate anus and a single opening anterior to the anus. The genitalia were otherwise normal. There were dense adhesions in the abdomen between the viscera and peritoneum. The abdominal contents were displaced by a cystically dilated vagina that contained cloudy, white fluid, which on chemical analysis proved to be urine diluted with a serous fluid. The uterus was bicornuate, the myometrium was thickened and the cervix opened into the dilated vagina. No obstructing membrane or valve could be identified at the introitus but urethral catheterization had been attempted which may have ruptured a membrane that had been present. The right kidney and ureter were absent and there was hydronephrosis, hydroureter and subcapsular hemorrhage on the left side. The urethra stretched over the cystic vagina and opened into its anterior wall (Figure 2). The hydrometrocolpos was of the urinary type. The other abdominal viscera were normal. The trachea and larynx

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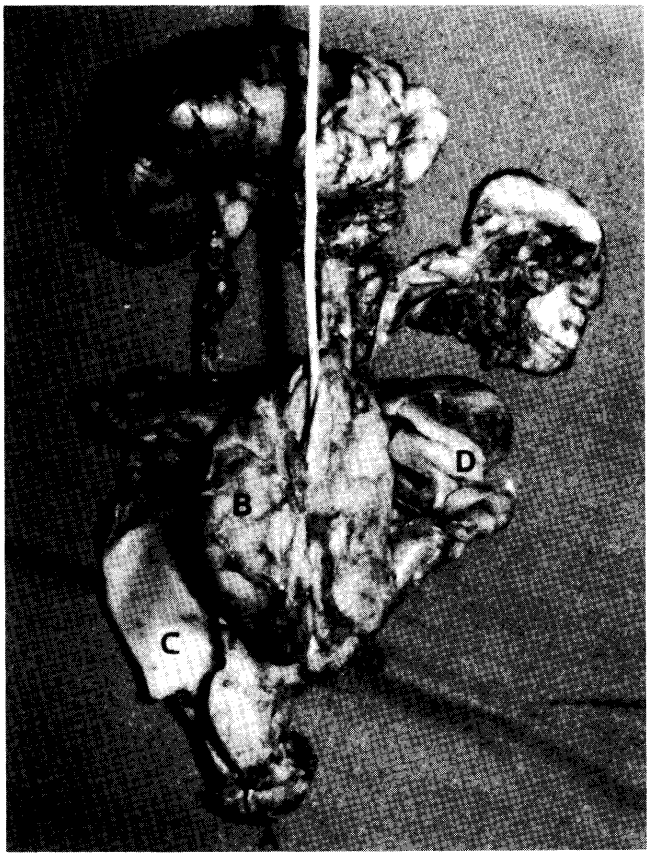

Figure 2 Autopsy dissection demonstrating absent left kidney. The tip of a probe (A) inserted into the bladder (B) is seen in the cystically dilated vagina (C); the uterus is bicornuate (D). The external visceral surfaces appear dull and irregular due to peritoneal adhesions

were normal. The lungs were solid, airless and hypoplastic. The infant had a normal female karyotype.

DISCUSSION

Hydrometrocolpos – distension of the uterus and cervix with non-hemorrhagic fluid – is of two types. The urinary type relates to an urogenital/cloacal abnormality, and the secretory type results from obstruction of the vaginal outlet and increased endocervical gland secretion. Of 62 cases presenting postnatally[1], 42 were due to imperforate hymen, 13 were due to vaginal atresia and seven had a vaginal septum. Obstruction can also be caused by the narrow and angular course of the urogenital sinus and its junction with the vagina[2]. A case of cloacal valve in a persistent urogenital sinus has been reported as a cause of hydrometrocolpos[4].

Postnatally, diagnosis is dependent on the presence of a lower abdominal mass in a female infant that does not disappear on catheterization. Careful examination of the external genitalia reveals a bulging membrane between the labia majora which may be exaggerated when the baby cries. If there is vaginal atresia, the hymen may be retracted upwards as the cystic mass escapes into the abdomen. Hydrometrocolpos may cause compression of the urinary tract, intestinal tract, or thoracic cage, or obstruct venous return. Other congenital abnormalities, which are the usual cause of mortality, are found in 17–81% of cases[1,3] and commonly relate to urogenital and hindgut anomalies, including unilateral or bilateral renal agenesis, bicornuate uterus, double vagina, bifid clitoris, tracheal stenosis or atresia[1–6]. Hydrometrocolpos is part of the triad of the McKusick–Kaufman syndrome which includes congenital heart disease and polydactyly and has an autosomal recessive inheritance[7]. Tracheal atresia, pulmonary hypoplasia, craniofacial anomalies, unilateral renal agenesis, ovarian dysgenesis and hydrometrocolpos have been reported in an infant with 47,XXX karyotype[5].

Prenatal diagnosis of hydrometrocolpos is made on ultrasonography by the presence of a cystic mass arising posterior to the bladder[7–10]. The amniotic fluid volume may be normal, increased or decreased[7,8]. The differential diagnosis includes ovarian and mesenteric cysts, dilated ureter and loops of bowel[7,8,10]. Rarely, the lesion may appear solid due to high mucus content of the cyst and mimic a neoplasm[9]. In our case, suspicion was raised by the presence of a cystic structure arising from the pelvis, similar to that seen in male fetuses with posterior urethral valves. In the absence of oligohydramnios and tracheal stenosis, as in this case, pulmonary hypoplasia is secondary to pressure from the abdominal cyst[8]. The backflow of retained fluid through patent Fallopian tubes caused peritoneal fibrosis, a condition recognized as plastic peritonitis[6].

REFERENCES

1. Spencer, R. and Levy, D. M. (1962). Hydrometrocolpos: report of three cases and review of the literature. *Ann. Surg.*, **155**, 558–71
2. Williams, D. I. and Bloomsberg, S. (1976). Urogenital sinus in the female child. *J. Pediatr. Surg.*, **11**, 51–61
3. Hahn-Pedersen, J., Kvist, N. and Neilsen, O. H. (1984). Hydrometrocolpos: current views on pathogenesis and management. *J. Urol.*, **132**, 537–40
4. Yu, S. F., Murugesu, J. J. and Chan, K. Y. (1967). An unusual case of neonatal hydrometrocolpos. *Am. J. Roentgen.*, **100**, 186–9
5. Hood, O. J., Hartwell, E. A., Shattuck, K. E. and Rosenberg, H. S. (1990). Multiple congenital anomalies associated with a 47XXX chromosome constitution. *Am. J. Med. Genet.*, **36**, 73–5
6. Ceballos, R. and Hicks, G. M. (1970). Plastic peritonitis due to neonatal hydrometrocolpos: radiologic and pathologic observations. *J. Pediatr. Surg.*, **5**, 63–9
7. Hill, S. J. and Hirsch, J. H. (1985). Sonographic detection of fetal hydrometrocolpos. *J. Ultrasound Med.*, **4**, 323–5
8. Russ, P. D., Zavitz, W. R., Pretorius, D. H., Manco-Johnson, M. L., Rumack, C. M., Pfister, R. R. and Greenholz, S. K. (1986). Hydrometrocolpos, uterus didelphys and septate vagina: an antenatal sonographic diagnosis. *J. Ultrasound Med.*, **5**, 211–13
9. Boulot, P., Deschamps, F., Hedon, B., Laffargue, F. and Viala, J. L. (1991). Prenatal diagnosis of an abdomino-pelvic hydrometrocolpos: a case report. *Eur. J. Obstet. Gynecol. Reprod. Biol.*, **40**, 233–6
10. Davis, G. H., Wapner, R. J., Kurtz, A. B., Chibber, G., FitzSimmons, J. and Blocklinger, A. J. (1984). Antenatal diagnosis of hydrometrocolpos by ultrasound examination. *J. Ultrasound Med.*, **3**, 371–4