Case 3309

Hydrometrocolpos in a neonate
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Section: Paediatric radiology
Imaging Technique: Ultrasound
Case Type: Clinical Cases
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Patient: 14 days, female

Clinical History:
An asymptomatic female neonate presented for a follow-up ultrasound, after an antenatal ultrasound revealed hydronephrosis and a cystic mass in the pelvis.

Imaging Findings:
Ultrasound of the pelvis was performed on an asymptomatic female neonate, to follow up an antenatal diagnosis of hydronephrosis with a cystic mass lesion seen in the pelvis. The postnatal ultrasound again showed hydronephrosis, which was caused by a compression of the distal ureter by the pelvic mass. The mass in the pelvis was identified as a dilated, fluid-filled uterus and vagina.

Discussion:
A wide spectrum of obstructed uterovaginal anomalies exist, and some knowledge of genitourinary embryology is useful in understanding them. In early embryonic development, both Wolffian (male) and Müllerian (female) ducts are present. The Müllerian ducts meet in the midline at their caudad end, fuse and join with the Müllerian tubercle on the posterior aspect of the urogenital (UG) sinus. The fused ducts form the uterus, the cervix and the upper vagina. The cranial Müllerian ducts remain unfused and become the fallopian tubes. The lower vagina is thought to be formed from the Müllerian tubercle and the sinovaginal bulbs of the UG sinus. The urethra, the vestibule and Bartholin’s glands also develop from the UG sinus. The hymen membrane represents the separation of the vaginal lumen from the urogenital sinus. In the female, the Wolffian duct system involutes. Uterovaginal obstructions are classified into three groups: (1) Müllerian agenesis, (2) defects of vertical fusion, and (3) defects of lateral fusion. The Mayer–Rokitansky–Kuster–Hauser syndrome is a partial or complete vaginal agenesis, in association with abnormalities of the uterus and cervix. The lower third of the vagina is usually present, due to its different embryological origins. Almost half of the patients with this syndrome will also have renal tract malformations, and a significant proportion will have skeletal abnormalities. Vertical fusion defects include a transverse vaginal septum, an imperforate cervix, cervical agenesis and vaginal agenesis with a normal uterus. An imperforate hymen may also be included in this group. Lateral fusion defects result in uterine duplication anomalies. A vaginal septum is most commonly associated with a uterus didelphus. Lateral fusion disorders are implicated in infertility and also in pregnancy-related problems. Most patients with obstructed uterovaginal anomalies are first seen at around the time of puberty, and present with a perineal or abdomen-pelvic mass due to a haematocolpos or haematometrocolpos, respectively. Abdominal pain, primary amenorrhea or voiding dysfunction may also be the presenting symptoms in this group of adolescent girls. A vaginal obstruction in the neonate is usually associated with a UG sinus or cloacal malformation. These patients may present with hydro- or mucocolpos (dilatation of the vagina with fluid or mucous secretions), with or without hydrometrocolpos (distension of the uterus and the vagina). It is thought that these neonates are either more sensitive to, or are exposed to higher levels of maternal oestrogen, and this leads to the accumulation of secretions in the uterus and vagina. An ultrasound scan is the most common initial imaging technique in the evaluation of these patients, and shows a “light-bulb” shaped, fluid-filled mass: the dilated uterus...
and vagina. In some cases, the uterus, which is less distensible, may be seen as a small cap-like structure superior to a markedly dilated vagina. A pelvic MRI is a helpful adjunctive examination to define the anatomy of the upper Müllerian structures. In our patient, the pelvic mass caused hydronephrosis due to the compression of the distal ureters. In cases of Müllerian agenesis, a spinal ultrasound scan in neonates, and an MR imaging of the spine, and spinal radiographs in older girls are required to rule out co-existent vertebral and spinal cord anomalies.

**Differential Diagnosis List:** Hydrometrocolpos.

**Final Diagnosis:** Hydrometrocolpos.

**References:**


Figure 1

Description: A parasagittal image of the right kidney, showing a dilatation of the pelvicalyceal system secondary to compression of the distal ureter by the pelvic mass. Origin:
Description: A longitudinal image, showing the fluid-filled uterine cavity and the vagina. Origin:
**Description:** A magnified view of the uterine body and the cervix. The contour of the distended uterus is easily recognisable. **Origin:**