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Uterus didelphys and obstructed right hemivagina associated with ipsilateral ovarian endometriosis and renal agenesis

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ABSTRACT

An unusual cause of pelvic pain in a 13 year old girl is presented.

1. Introduction

Müllerian duct anomalies (MDAs) are uncommon but pelvic pain in an adolescent female is a recognised presentation of an obstructed anomaly. We present a case of obstructed uterus didelphys with typical findings of haematocolpos and haematometra associated with both a longitudinal and a horizontal vaginal septum. In addition two associations of haemato/hydrosalpinx and an endometrioma not previously demonstrated in the imaging literature are described.

2. Case report

A 13-year-old post menarchial girl presented to the emergency department with a 12-h history of right lower abdominal pain. She had had similar pain previously, and though her menstrual cycle was not yet regular, the pain did seem cyclical and associated.

Of relevance in her past medical history was right renal agenesis. She was virgo intacta.

On examination she had a temperature of 38.3 °C and a tender lower abdomen. Urine microscopy was normal and haematological investigations revealed a white cell count of 14×10^9/l (normal $4–11 \times 10^9$/l) and C reactive protein of 30 mg/dl (normal <1 mg/dl). She was admitted under the paediatricians.

Ultrasound showed two separate uterine horns: The one on the right was distended and fluid filled (Fig. 1). An apparently single, distended fluid filled vagina was seen. The right ovary was deemed to be normal, the left ovary not visualized and the right kidney was absent.

On the basis of these findings the patient was discharged with outpatient appointments for a pelvic MRI and gynaecological review.

The MRI examination and the subsequent scan were performed on a Philips Intera 1.5 T MR scanner. Overall the following sequences were used:

Axial T1 turbo spin echo (TSE), 350 field of view (FOV), 5 mm slices, 1 mm gap, TE 10, TR 596, 3 NSA
Coronal T2 TSE, 200 FOV, 4 mm slices, 1 mm gap, TE 100, TR 4139, 6 NSA
Sagittal T2 TSE, 200 FOV, 4 mm slices, 1 mm gap, TE 100, TR 4972, 6 NSA
Coronal T1 TSE, 200 FOV, 4 mm slices, 1 mm gap, TE 10, TR 490, 3 NSA
Axial oblique T2 TSE, 200 FOV, 4 mm slices, 1 mm gap, TE 100, TR 2396, 6 NSA

The patient was fasted for 6 h prior to the scans to minimize small bowel peristalsis and hyoscine butylbromide 20 mg was administered intravenously immediately before each examination.

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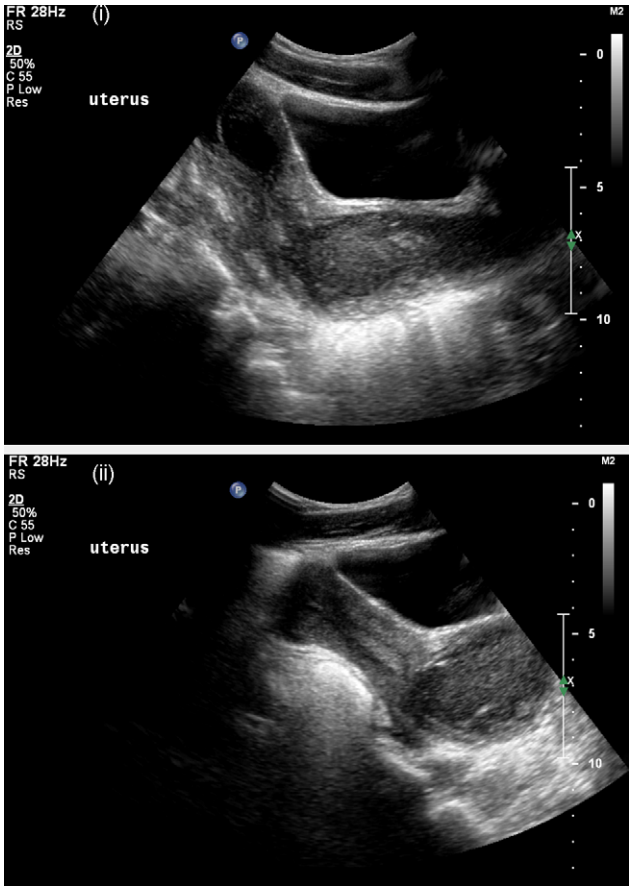

Fig. 1. Sagittal section ultrasound image demonstrates (i) distended, fluid filled right uterine horn and haematocolpos (ii) left uterine horn and haematocolpos. Key: A – right endometrioma; B – haematometracolpos complex.

On MRI, the anatomy was distorted by a right haematometrocolpos. Two uterine bodies and two cervices were confirmed consistent with uterus didelphys. A longitudinal vaginal septum was present with a distended right hemivagina due to a transverse septum across the lower third. The left hemivagina was empty. There was a right hydro/haematosalpinx and a cyst containing blood products arising from the right ovary. The left ovary was normal (Fig. 2). No ureter was identified on the images.

Subsequently, an examination under anaesthesia was performed and a hysteroscopy attempted. The right haematocolpos was confirmed and the transverse septum in the right hemivagina was incised. The hysteroscopy failed as no cervix could be found.

Three months after the acute presentation a second pelvic MRI examination was performed. This demonstrated resolution of the haematometra and haematocolpos with less distortion of the anatomy. There was a persisting right hydrosalpinx and blood containing cyst on the right ovary: the signal characteristics of this being diagnostic of an endometrioma (Fig. 3).

In view of the patient's young age no immediate further intervention was planned, and conservative management with continued review was deemed most appropriate.

3. Discussion

MDAs are congenital anomalies of the female genital tract that arise from nondevelopment or non-fusion of the Müllerian ducts or failed resorption of the uterine septum. Their reported incidence is 0.5–5.0% [1,2].

The uterus, tubes, and upper two thirds of the vagina originate from paired Müllerian (paramesonephric) ducts, whereas the lower third of the vagina arises from the urogenital sinus. At 6 weeks gestation the paired ducts invaginate, grow caudally crossing over the Wolffian (mesonephric) ducts to meet at the midline. Fusion and resorption of the midline septum takes place by 10.5 weeks. Cranially the uterine tubes remain paired and unfused. Caudally the urogenital sinus invaginates to form the lower third of the vagina.

Müllerian duct development is closely related to the development of the urinary system as both originate from a common embryonic mesoderm. This accounts for the frequent association of anomalies. The coexistence of unilateral renal agenesis with MDAs has a reported incidence of 55–75% [3], and uterus didelphys has a particular association with unilateral renal agenesis [3–5].

Renal development begins in the 5th week of gestation from two sources; 1. the ureteric bud, an outgrowth of the Wolffian duct near the cloaca, and 2. the metanephric cap. The ureteric bud develops a lumen becoming the ureter and renal pelvis. Adjacent cells within intermediate mesoderm form the metanephric cap and this surrounds the renal pelvis. The ureteric bud undergoes multiple divisions forming calyces and collecting tubules. This in turn induces the nephrons to develop from the metanephric cap tissue.

It is presumed that early degeneration of the ureteric bud results in renal agenesis due to failure of induction of metanephric cap tissue proliferation [6]. This would suggest that in the setting of renal agenesis, the ipsilateral ureter is also absent.

The patient under discussion was known to have right renal agenesis but it is inconclusive as to whether there was also absence of the right ureter as this structure was not identified on MRI. Indeed, the left ureter was not seen either, but it is not unusual not to visualize a normal ureter.

Uterus didelphys constitutes 5% of MDAs [7]. It results from nonfusion of the Müllerian ducts, with the development of two separate normal sized uteri and cervices. There is a longitudinal vaginal septum in 75% of cases and frequently a transverse septum across a hemivagina [3,4,8]. A common complication is haematometrocolpos secondary to the transverse septum. Moreover the syndrome of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis, as in the current case, has frequently been cited. This constellation of findings is most often right sided [3,8].

Obstructed uterus didelphys typically presents with acute or chronic pelvic pain shortly after the menarche but in spite of this, the diagnosis may be delayed. This may be because the condition is uncommon and therefore not considered. Also, adolescents presenting with dysmenorrhoea are often given anti-inflammatory drugs or oral contraceptives without recourse to imaging [8].

The differential diagnoses include acute appendicitis, ovarian cyst accident (torsion or haemorrhage), ectopic pregnancy and pelvic inflammatory disease. The latter two were immediately excluded in the current case as the patient was virgo intacta. Regardless, a pregnancy test would be mandatory. However, the diagnosis of an obstructed MDA was made promptly with the admission ultrasound examination (US). Its exact nature was determined later with MRI.

In the setting of an ovarian cyst haemorrhage MRI would have been a useful problem solving diagnostic tool. Nishino et al. [9] have described the MRI findings of acute ovarian haemorrhage; intermediate signal intensity on both T1 weighting (T1W) and T2 weighting (T2W). They state that the presence of haemorrhagic ascites is another useful sign and it has similar signal characteristics to the ovarian haematoma. Whilst an ovarian cyst containing blood products was demonstrated at MRI in the case under discussion, its signal characteristics were not of acute haemorrhage but of