Case Report

A case of uterus didelphys and unilateral renal agenesis

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حالة لإزدواج الرحم مع عدم تخلق الكلوي المماثل

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الخلاصة
يعتبر ازدواج الرحم مع ارتق عنق الرحم آحادي الجانب من تشوهات الرحم الخلقية النادرة نسبيا. ويرتبط ذلك مع عدم تخلق الكلوي المماثل. هذا التقرير يصف حالة العرض التقديمي، ملخص التحقيقات ذات الصلة، إدارة ومتابعة لحالة ازدواج الرحم.

Summary
Uterus didelphys with unilateral imperforate cervix is a relatively rare congenital anomaly. It is associated with ipsilateral renal agenesis. This is a case report of seventeen-year-old girl presenting with right iliac fossa pain and a mass arising from the pelvis. Transvaginal ultrasound revealed a large cystic mass (10 x 10 x 7 cm) filling up the pelvis. Abdominal ultrasound confirmed this with an incidental finding of unilateral renal agenesis. She underwent a laparotomy where it was found that she had uterine didelphys. The 'cyst' was a haematometra. This was aspirated vaginally. Postoperative magnetic resonance imaging confirmed uterus didelphys with two separate uterine cavities. This girl was referred to a consultant gynaecologist who has a special interest in this area.

Keywords: Müllerian duct anomalies, abdominal pain, haematometra

Introduction

Uterus didelphys with obliterated unilateral cervix can be diagnosed early and accurately. It is easily corrected, with diminished long-term morbidity and with preservation of reproductive function in most of cases. Diagnosis of the condition is best reached with the gold standard – magnetic resonance imaging (MRI) [1].

Case Report

The patient was a seventeen-year-old unmarried girl, para 0+0; her last menstrual period was 25 days previously. She had her menarche at age thirteen; initially, periods were very irregular, then they became moderate, usually lasting five days. Two weeks prior to her presentation, she had unprotected intercourse. Her pregnancy test was negative.

She was referred to the emergency department by her GP with a four-day history of intermittent 'crampy' right iliac fossa (RIF) pain. There were no identifiable, alleviating or aggravating factors. She reported no change in urinary/bowel habits. Her medical history was unremarkable and she was on no regular medication. Her mother died six years before with metastatic breast cancer.

Abdominal examination revealed a large ballotable smooth tender mass arising from the pelvis, extending up to one finger below the umbilicus. There was no rebound tenderness or guarding. The cervix was not felt on vaginal examination. Transvaginal ultrasound scan demonstrated a large cystic mass filling up the whole of the pelvis, measuring 10 x 10 x 7 cm. The uterus could

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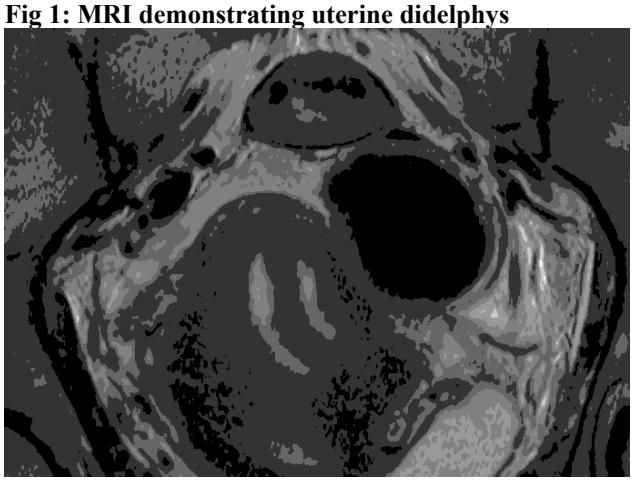
not be identified separately from it. The patient was subsequently admitted for a 'laparotomy and drainage of the cyst'.

Prior to the operative procedure, she had an abdominal ultrasound scan which did not detect the right kidney and had compensatory hypertrophy of left kidney. Once again, a cyst was demonstrated in the pelvis extending from the RIF to the pouch of Douglas. The radiologist queried that as an adnexial cyst compressing the cervix. The right ovary was unremarkable and left ovary was not seen. As the origin of the abdominal mass was not clearly identified pre-operatively, the patient underwent a laparotomy.

Examination under anaesthesia revealed normal looking vulva and vagina. Hymen was not intact as she was sexually active. The cervix was not identified and a large cystic mass was palpated. Laparotomy revealed a uterus didelphys with normal tubes and ovaries and a large cystic mass below the uterus replacing the cervix. The 'cyst', was a haematometra. This was aspirated vaginally and showed altered blood.

Postoperatively, the patient had an MRI pelvis as shown in Fig 1, to help ascertain a definitive diagnosis. This revealed a uterus didelphys with two separate uterine cavities with a septum dividing both of them) with two cervices; one being imperforate.

Fig 1: MRI demonstrating uterine didelphys



The condition was explained to the patient and she was referred to a tertiary centre with experience in management of such cases. She was started on gonadotropin-releasing hormone (GnRH) analogue to prevent any further menstruation until she had longer term management.

Discussion

Müllerian duct anomalies affect between 0.1% and 3% of women[2]. In the most extreme form of the Müllerian duct non-fusion, uterus didelphys results, with complete duplication of the uterus, cervix and vagina. This anomaly accounts for 11% of uterine malformations and is typically asymptomatic[3,4,5,6]. Uterus didelphys is most often recognised as part of a syndrome associated with an obstructed hemi-cervix and ipsilateral renal agenesis. The obstruction of one hemi-cervix will block outflow leading to the creation of a cystic mass. This may result in complications such as haematometra as demonstrated by this patient.

The triad of common symptoms seen in these patients is dysmenorrhoea - that begins shortly after menarche, increasing in severity of dysmenorrhoea-with each subsequent period and a unilateral pelvic mass[7,8]. Occasionally, patients may present with symptoms from the urinary tract, including acute urinary retention and dysuria or having gastrointestinal complaints such as rectal pain, constipation or signs of an acute abdomen. Fortunately, this patient's symptoms did not progress further to cause gastrointestinal/urinary disturbance.

This case highlights the importance of careful assessment of this group of young patients including the use of appropriate imaging modalities when a young woman presents with the above symptoms. Laparotomy could therefore have been avoided. Appropriate follow-up would include MRI, preferably at a tertiary centre with expertise in interpretation of Müllerian anatomy, as well as early consultation and referral to a centre with expertise in the management of such cases.

Follow-up was necessary because the affected cervix was not opened for future escape of the menses; accordingly the condition may recur. This girl was referred to a consultant gynaecologist who has a special interest in this area, feedback is awaited. She was also referred to a urologist for the incidental finding of renal agenesis.

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