CLINICAL CASE - TEST YOURSELF Abdominal Imaging

Uncommon cause of pelvic pain in adolescent female patient

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PART A

A 13-year-old female patient presented to the emergency department with a 3-month recurrent and progressive pelvic pain during the menstrual bleeding. Her menarche was about 6 months ago. No abnormal findings were found on physical examination. The laboratory tests revealed normal white blood cell count. Pelvic ultrasound was initially performed (**Fig.1**), followed by magnetic resonance imaging (MRI) (**Fig. 2**). The patient then underwent a vaginohysteroscopy and a post-surgery MRI.

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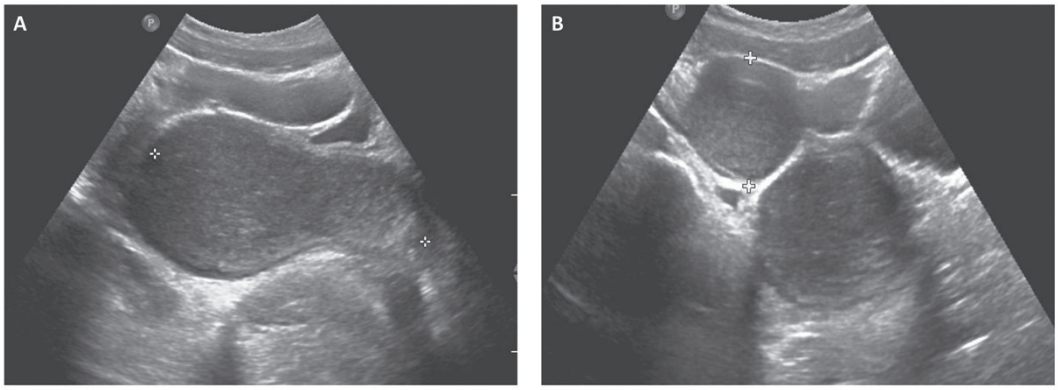

Fig. 1. *Pelvic Ultrasound in longitudinal* ***(A)*** *and axial* ***(B)*** *views.*

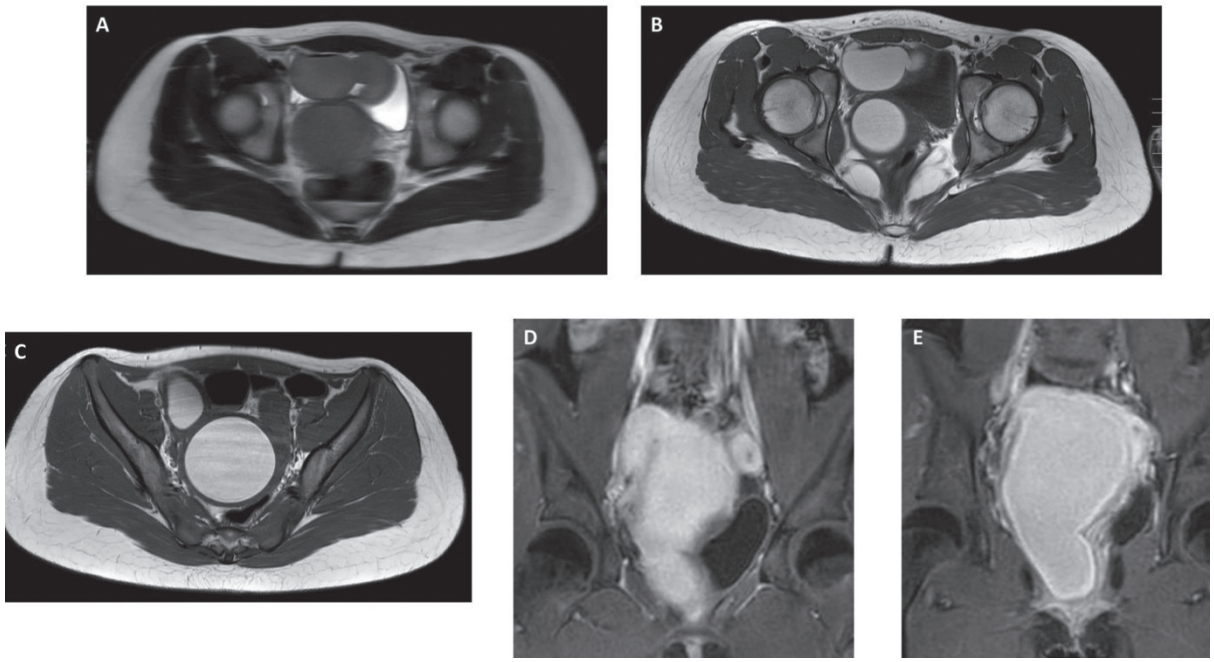

Fig. 2. *Magnetic Resonance Images.* ***(A)*** *Axial T2W images;* ***(B)****,* ***(C)*** *Axial T1W images;* ***(D)****,* ***(E)*** *Coronal T1W contrast-enhanced images.*

PART B

Diagnosis: Herlyn-Werner-Wunderlich syndrome (uterus didelphys, blind hemivagina and ipsilateral renal agenesis)

Sonographic findings included at least 3 cystic lesions in the anatomic region of the uterus and the ovaries, with maximum longitudinal diameter at 10 cm, filled with hyperechoic material, whereas both ovaries were normal (**Fig. 1**). Due to the size of the cystic lesions, it wasn't easy to determine their origin The differential diagnosis included haematometra, pyometra, hydrosalpinx, haematosalpinx or pyosalpinx combined with haematocolpos and endometrioma. Further investigation with pelvic MRI demonstrated uterus didelphys with two separate uteri and two separate cervices (**Fig.2**). The right endometrial cavity and cervix were dilated. The right fallopian tube was also dilated (haematosalpinx), filled with inhomogeneous high-signal content (**Fig.2**). The left uterus and cervix were displaced, but not dilated. No free fluid was shown. The absence of the right kidney raised the suspicion of Herlyn-Werner-Wunderlich syndrome.

A vaginohysteroscopy was performed for further investigation and for relief of symptoms. Vaginoscopy confirmed the presence of right-sided haematocolpos, with blood draining from the right haematometra and a longitudinal vaginal septum. Incision and resection of the vaginal septum produced about 300 ml of old unclotted blood.

A post-surgery MRI scan performed 2 days after surgical intervention demonstrated a less dilated right-sided hemiuterus and cervix (**Fig.3**). The patient was discharged 4 days after surgery.

Herlyn-Werner-Wunderlich syndrome is a rare congenital malformation of the urogenital track, also known as OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrome with an estimated incidence of 1/2,000 to 1/28,000 births [1,2].

In 1971, Herlyn and Werner reported a case of ipsilateral renal agenesis with blind hemivagina and a Gartner duct cyst and later, in 1976, Wunderlich described the association of ipsilateral renal agenesis, bicornuate uterus with simple vagina, and isolated haematocervix [3].

It is typically characterized by the presence of uterus didelphys with blind hemivagina and ipsilateral renal agenesis [1]; however, several anatomical variants, i.e. fenestrated vaginal septum, bicornuate uterus and other renal anomalies including dysplastic, multicystic and ectopic kidney have also been reported [3]. The pathogenesis of this syndrome is associated with abnormal development of both Mullerian and mesonephric ducts, but the exact aetiology remains unknown [4].

This syndrome is mostly discovered during adolescence, shortly after menarche [5]. The main clinical manifestations include non-specific symptoms such as progressive and recurrent pelvic pain after menarche, dysmenorrhea, and vaginal discharge of smelly, clotted blood (in case of communication between the two haemovaginas) while a palpable mass due to the associated haematocolpos/haematometra can be revealed on physical examination [1]. The syndrome is usually under-recognized at first, as the menstrual flow from the normal (i.e. the unobstructed) system may be regular [1]. Long-term complications include pelvic inflammatory disease, endometriosis, pelvic adhesions and increased risk of infertility [6].

Diagnosis is based on imaging studies; ultrasound is a costless, non-invasive and very useful initial diagnostic tool, but MRI is the diagnosis and classification of urogenital track abnormalities [5]. Ultrasonography can detect the presence of haematocolpos, haematometra and renal abnormalities, but cannot identify the vaginal septum [6]. MRI provides more details about the uterine and cervix morphology, the presence of two hemivaginas (obstructed or not) and the fluid nature [5]. Laparoscopy is mostly used for identification of complications, including endometriosis and pelvic infection and adhesions [6].

The treatment is surgical; administration of ovarian suppressive agents (i.e. oral contraceptives, GnRH agonists) is commonly used as a bridging therapy until the surgery [7]. Single-stage vaginoplasty and vaginal septum resection is the treatment of choice for the majority of the patients [6], providing relief of the symptoms and prevention of disease-related complications. Hemihysterectomy is considered when vaginoplasty is

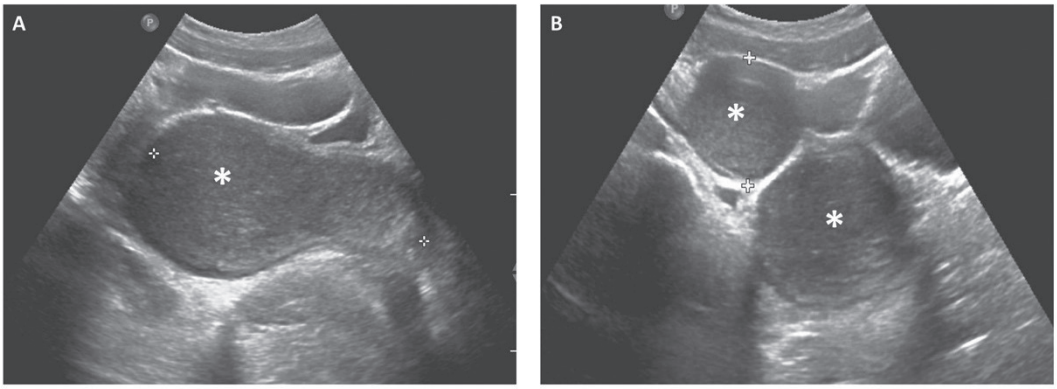

Fig. 1. *Ultrasonographic images in longitudinal (**A**) and axial (**B**) views showing the uterus filled with haemorrhagic material (**A**) and the fallopian tube filled with haemorrhagic material (**B**).*

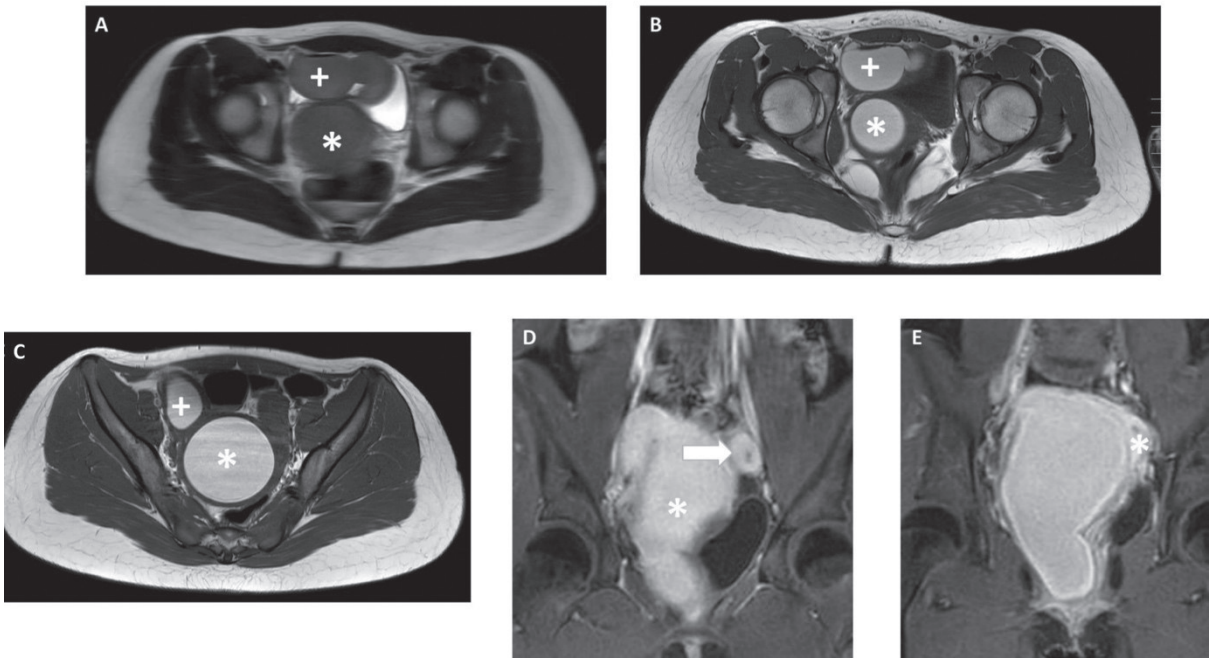

Fig. 2. *Magnetic Resonance Images. (**A**) Axial T2W images showing the endometrial cavity (asterisk) and the right fallopian tube filled with hypointense fluid (plus sign). (**B**), (**C**) Axial T1W images non contrast showing the uterus (asterisk) and the right fallopian tube filled with haemorrhagic material (plus sign). (**D**), (**E**) Coronal T1W contrast enhanced images demonstrate duplication of the endometrial cavities, normal left cavity (arrow) and right cavity full of blood (asterisk).*

impossible (i.e. proximal vaginal septum) or in case of sepsis [6].

In conclusion, Herlyn-Werner-Wunderlich syndrome is a rare congenital malformation of the genitourinary track should be suspected in cases with recurrent pelvic pain started shortly after menarche and in neonates with renal abnormalities. The evaluation of both renal and genital system is every important and can easily lead to a diagnosis. Prompt diagnosis is essential for symptomatic treatment and prevention of future complications. Although abdomen ultrasound is useful diagnostic tool, MRI is considered the gold-standard imaging modality. **R**

Conflict of interest

The authors declared no conflicts of interest.

KEY WORDS

Herlyn-Werner-Wunderlich syndrome, Renal agenesis, Mullerian duct, OHVIRA syndrome, pelvic MRI

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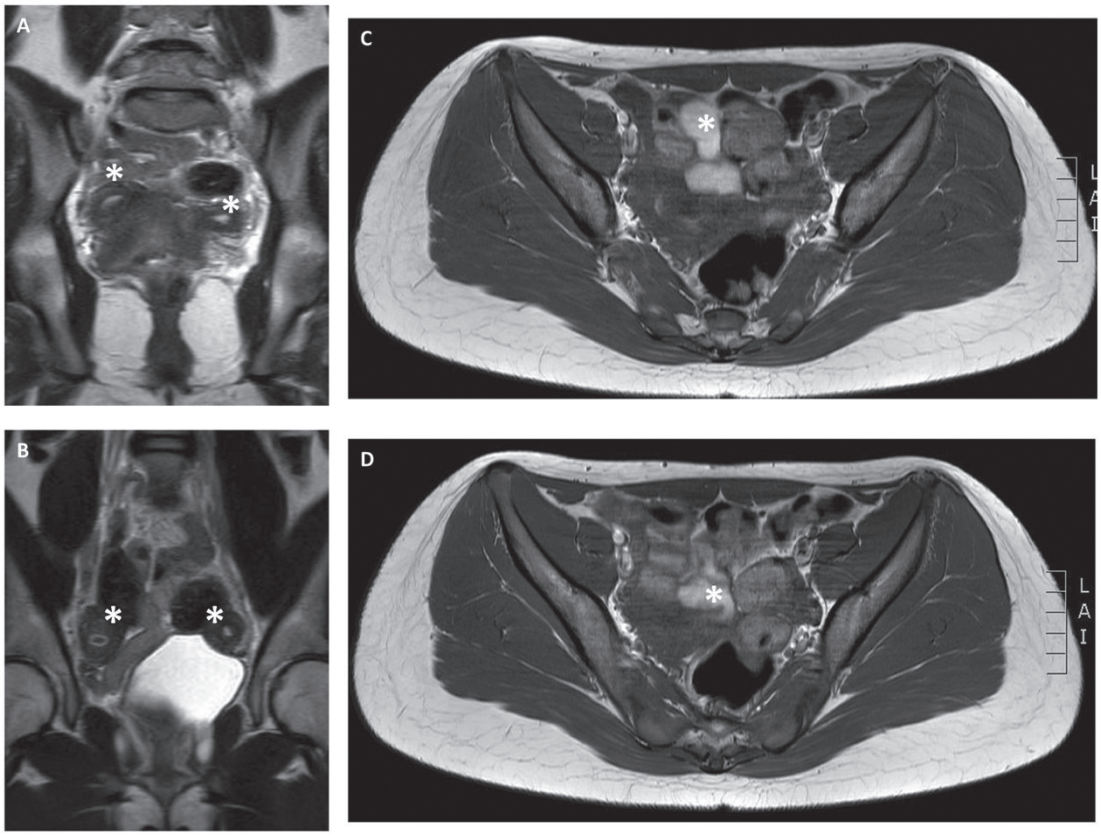

Fig. 3. *Post-surgery Magnetic Resonance Images. **(A)**, **(B)** Coronal T2W images showing duplication of the endometrial cavities, both cavities are normal (asterisks). **(C)**, **(D)** Axial T2W images showing small amount of haemorrhagic material in the right adnexa.*