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A non-classic presentation of OHVIRA syndrome

Uma apresentação não-clássica do síndrome OHVIRA

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Keypoints

What is known

- OHVIRA syndrome is a rare urogenital malformation, characterized by uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis.
- It typically presents with progressive and severe dysmenorrhea, but it can also be diagnosed due to acute abdominal pain.

What is added

- OHVIRA syndrome should be a differential diagnosis in acute abdominal pain in female patients, especially adolescents.
- Patients with absent or dysplastic kidney should be routinely investigated for uterovaginal abnormalities, in order to prevent potentially severe complications.

Description

A 13-year-old female adolescent was admitted to the Pediatric Emergency Department with abdominal pain and occasional vomiting, for three days. Her past medical history included a dysplastic right kidney. She had her menarche at the age of 12 years and she had regular menses, without dysmenorrhea. She was haemodynamically stable and physical examination revealed a tender and guarding abdomen in the right iliac region, without any other findings. An initial investigation was performed to exclude acute appendicitis. Laboratory evaluation showed mild leucocytosis ($12.8x10^9$/uL), elevated C-protein reactive (11 mg/dL, for a normal value of < 0.2 mg/dL) and haemoglobin of 12.0 g/dL. Abdominal and pelvic ultrasound revealed two uterine cavities and a tubular image of 15.8 x 8.7 x 8.3 cm, located in the vaginal canal, filled with what seemed to be hematic content. Magnetic resonance image (MRI, Figs. 1 and 2) with contrast was performed and it reported uterus didelphys and two probable hemivaginas, the right one obstructed, with a resultant hematometrocolpos. A dysplastic right kidney and an ectopic right ureter was also evident, with a distal implantation in the right hemivagina. These findings suggested the diagnosis of OHVIRA (obstructed hemivagina and ipsilateral renal agenesis/anomalies) syndrome. The patient underwent an incision in the vaginal septum, with drainage of the accumulated blood and partial resection of the septum, to prevent obstruction of the hemivagina in the future. The procedure went without complications, and she was discharged home the day after. Since then, she has been pain free. After one year of the procedure, the hemivagina is still unobstructed.

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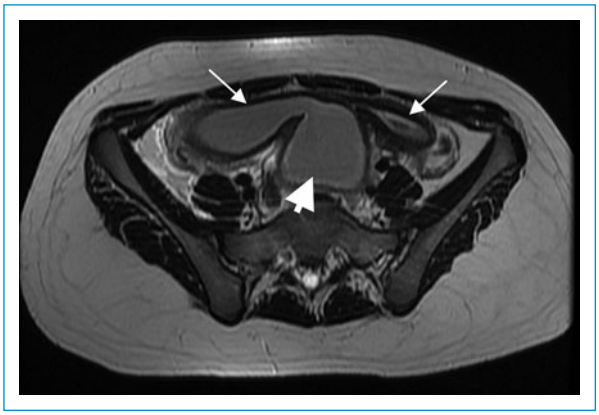

Figure 1. Axial section of magnetic resonance image of the pelvis with contrast showing uterus didelphys, with two uterine cavities (arrows) and what was assumed as the right obstructed hemivagina (large arrow), with an hematometrocolpos.

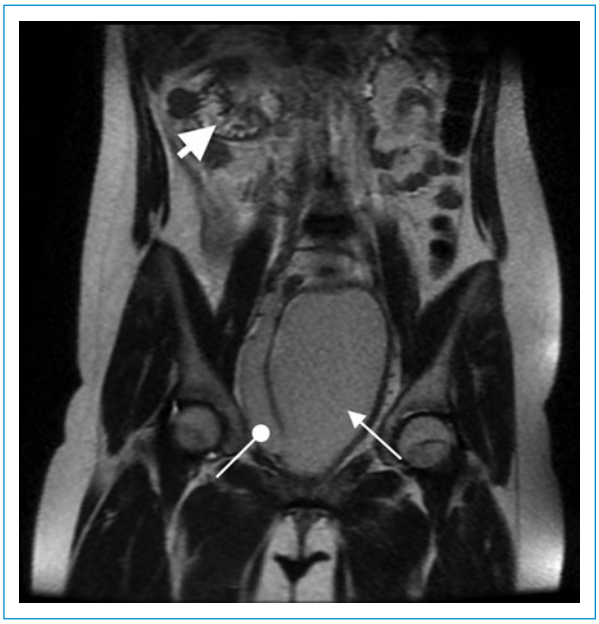

Figure 2. Coronal section of magnetic resonance image of the pelvis with contrast showing what was assumed as the right obstructed hemivagina (arrow), with a distal implantation of an ectopic right ureter (round arrow). Dysplastic right kidney is also shown (large arrow).

OHVIRA syndrome (or Herlyn-Werner-Wunderlich syndrome) is a rare urogenital malformation, occurring in only 5% of all Müllerian duct abnormalities[1]. It is typically characterized by uterus didelphys, obstructed hemivagina, due to a vaginal septum, and ipsilateral renal agenesis, although other renal findings, like dysplastic kidneys and ectopic ureters, are possible[1-4].

The diagnosis, in most of the cases, is delayed until puberty, and it is usually made due to progressive and severe dysmenorrhea, with or without a pelvic mass, which happens because the obstructed hemivagina causes an increasing hematometrocolpos[2-4]. Less frequently, some cases present with urinary retention or acute abdominal pain, like in our patient[2,3]. MRI is the gold standard for anatomic characterization and final diagnosis, although ultrasonography may have an important initial role[1-4].

The treatment of choice is surgical, most of the cases with a resection of the vaginal septum, and it is extremely important for symptom relieve and prevention of long-term complications. An obstructed hemivagina leads to a retrograde flow of the blood into the uterine cavity, which can lead to endometriosis and pelvic adhesions. Other complications, like impaired fertility, pregnancy complications and pelvic infections can also occur[1-3].

This case is important to remind that OHVIRA syndrome should be a differential diagnosis in acute abdominal pain, although it is not the classic presentation. Furthermore, it is also a diagnosis to bear in mind when facing patients with absent or dysplastic kidney, and some authors recommend investigation of uterovaginal abnormalities in such cases to allow early recognition and treatment[2].

Awards and presentations

This case was presented at the 21th National Congress of Pediatrics that took place between 27th and 29th October 2021, Braga, Portugal.

Funding

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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