ACTA RADIOLÓGICA PORTUGUESA
January-April 2025 | Vol 37 nº 1 | 27-28 https://doi.org/10.25748/arp.34637

Images of Interest / Imagens de Interesse

Abdominal Pain in a Female Adolescent: A Diagnosis to Consider

Dor Abdominal numa Adolescente: Um Diagnóstico a Considerar

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Received: 11/02/2024
Accepted: 05/04/2024
Published: 30/04/2025

Abstract

OHVIRA syndrome is a rare congenital female anomaly, characterized by the triad: uterus didelphys, unilaterally obstructed hemivagina and ipsilateral renal agenesis. The authors present a case of a 12-year-old girl with cyclic abdominal pain, scarce catamenia and intense dysmenorrhea refractory to NSAIDs. An abdominal and pelvic computized tomography was carried out which revealed renal agenesis, duplication of the uterus, cervix, and vagina, and hematocolpos, suggesting vaginal obstruction. The prompt treatment allows a clear improvement in the clinical and reproductive prognosis, so the authors emphasize the importance of the knowledge and suspicion of this condition, allowing the diagnosis, referral, and early approach.

Keywords

Congenital defects; Diagnosis; Female genitourinary diseases; OHVIRA syndrome.

Resumo

A síndrome de OHVIRA é uma rara anomalia congénita feminina, caracterizada pela tríade: útero didelfo, hemivagina unilateral obstruída e agenesia renal unilateral. Os autores apresentam um caso de uma menina de 12 anos com dor abdominal cíclica, cataménio escasso e intensa dismenorreia refratária aos AINEs. Foi realizada uma tomografia computorizada que revelou uma agenesia renal, duplicação do útero, colo uterino e vagina, e hematocolpos a sugerir obstrução vaginal. O tratamento precoce permite uma melhoria significativa do prognóstico clínico e reprodutivo, por isso os autores realçam a importância do conhecimento e suspeita desta condição, permitindo um diagnóstico, referenciação e tratamento precoce.

Palavras-chave

Defeito congénito; Diagnóstico; Doenças genitourinárias femininas; Síndrome de OHVIRA.

Case

A 12-year-old girl, with an irrelevant medical history, was admitted to the pediatric emergency department with pollakiuria and abdominal and genital pain for two weeks, with worsening in the day before admission. There was a reference to menarche in the six months before, with scarce catamenia and intense dysmenorrhea refractory to NSAIDs. Last catamenia had been two weeks prior to the admission. On physical examination, the lower abdominal quadrants were rigid and painful on palpation and decompression, mainly in the right iliac fossa. An analytical study, urinalysis and pregnancy test were performed, all negative. Therefore, she did an abdominal ultrasound which suggested an hematocolpos (Fig. 1), but could not correctly evaluate the uterus. To better characterization, an abdominal and pelvic computized tomography was carried out which revealed the absence of the right kidney and retroaortic left renal vein, duplication of the uterus, cervix, and vagina, and marked distension of the right hemivagina and endometrial cavity, with hematocolpos, suggesting vaginal obstruction (Fig. 2). Afterwards, she did a renal scintigraphy for a renal morphofunctional assessment, which confirmed a right renal agenesis (Fig. 3). These findings allowed the diagnosis of an OHVIRA syndrome.

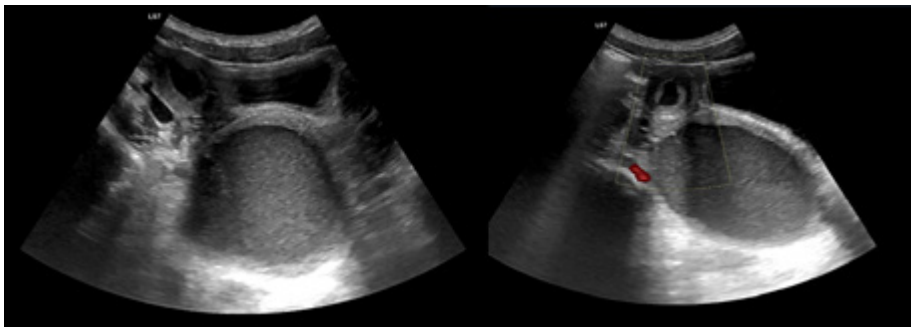

Figure 1 – Abdominal ultrassound: hematocolpos.

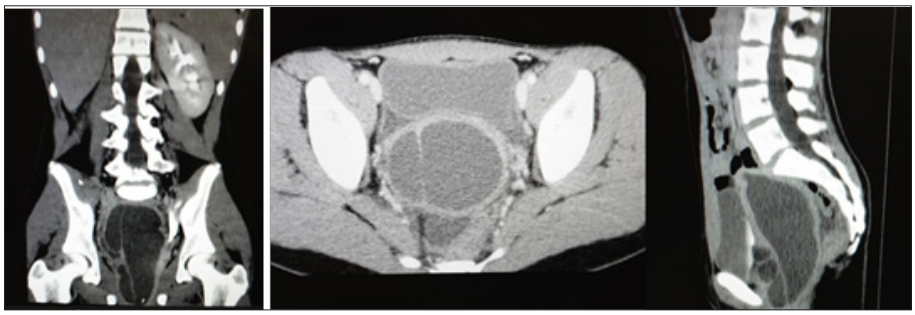

Figure 2 – Abdominal and pelvic computed tomography: right kidney and retroaortic left renal vein agenesis; duplication of the uterus, cervix, and vagina; hematocolpos.

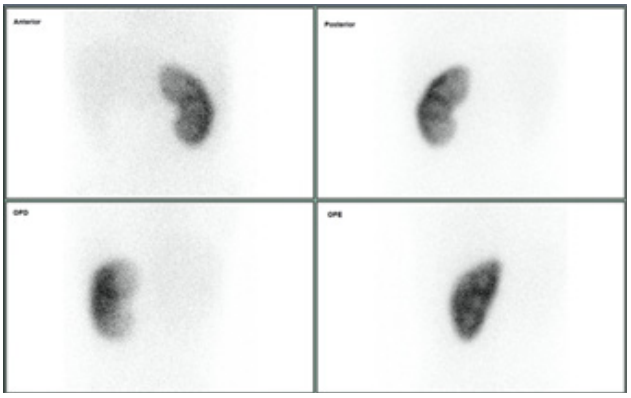

Figure 3 – Renal scintigraphy: right renal agenesis.

Discussion

OHVIRA syndrome is a rare congenital female anomaly, that results from an abnormal embryological development of the Mullerian and Wolffian ducts.[1,2,3] It is characterized by the triad: uterus didelphys, unilaterally obstructed hemivagina and ipsilateral renal agenesis.[1,2,3] This syndrome should be considered as a differential diagnosis in female adolescents with dysmenorrhea, abdominal pain, pelvic mass and/or inpatient retention, since it is the typical presention.[1,2,3] Also, in female patients diagnosed with renal agenesis, these anomalies should be investigated, even in pre-pubertal stages.[3] The prompt diagnosis and surgical treatment allow a clear improvement in the clinical and reproductive prognosis, avoiding late complications, such as endometriosis, pelvic adhesions, and infertility.[1,2,3] Thus, we emphasize the importance of the knowledge and suspicion of this condition, allowing the diagnosis, referral, and early approach.

Acknowledgments

We thank the parents of the patient for giving us permission to publish the data.

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

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

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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