ACTA RADIOLÓGICA PORTUGUESA
May-August 2020 | Vol 32 nº 2 | 35-36

Images of Interest / Imagens de Interesse

OHVIRA Syndrome with a Blind-Ended Ureteral Remnant

Síndrome OHVIRA com Remanescente Uretérico

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Abstract

OHVIRA syndrome is characterized by a didelphys uterus with an obstructed/blind hemi-vagina and ipsilateral renal agenesis. We presented a case of a female child with a pre-natal diagnosis of left renal agenesis whose post-natal imaging findings were consistent with OHVIRA syndrome.

Keywords

OHVIRA syndrome; Mullerian malformations; Congenital female genital tract anomalies.

Resumo

O Síndrome OHVIRA é caracterizado pela presença de útero didelfos com uma hemi-vagina obstruída e agenesia renal ipsilateral. Apresentamos o caso de uma criança com diagnóstico pré-natal de agenesia renal esquerda cuja investigação imagiológica pós-natal revelou um Síndrome OHVIRA.

Palavras-chave

Síndrome OHVIRA; Malformações mullerianas; Anomalias congénitas genitais femininas.

Ten-day-old female referred to a pediatric urology consultation due to prenatal diagnosis of left renal agenesis and a pelvic cystic image, which raised the suspicion of a nephro-urologic malformation.
At birth, analytically with normal values of Urea (34 mg/dl) and Creatinine (1.38 mg/dl). Ultrasound in the first week of life confirmed a hydrohematocolpos, showing a pelvic, non-pure, cystic image (Fig. 1), identified two hemi-uterus suggesting a didelphys uterus and a tubular image with bladder insertion adjacent to the left uterus, interpreted as a ureteral remnant.

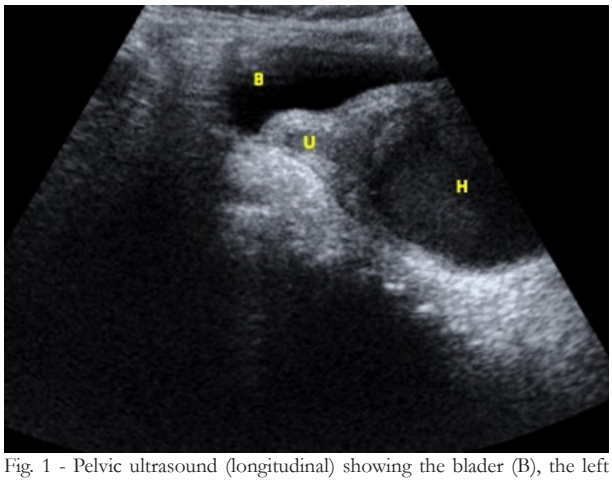

Fig. 1 - Pelvic ultrasound (longitudinal) showing the blader (B), the left hemi-uterus (U) and a hematocolpos of heterogenous content in the obstructed hemi-vagina (H).

Hydrohematocolpos was drained to prevent infection.
Magnetic resonance imaging was performed at 7 months of age, confirming left renal agenesis, a left ureteral remnant with ureterocele (Fig. 2) and a didelphys uterus (Fig. 3).

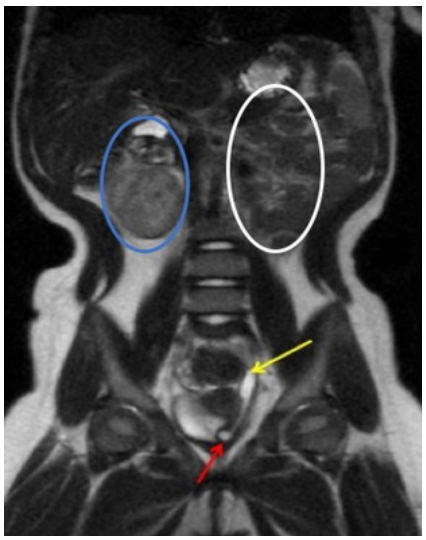

Fig. 2 – Coronal T2-weighted image demonstrating the presence of a right kidney (blue circle), absence of the left kidney (white circle), a left ureteral remnant (yellow arrow) and an ureterocele (red arrow).

Currently at 2 years-old, she remains asymptomatic and maintaining regular follow-ups with urologic pediatricians.
OHVIRA (Obstructed hemivagina and ipsilateral renal anomaly) syndrome, also known by Herlyn-Werner-Wunderlich syndrome, is a congenital anomaly of the female urogenital tract resulting from an anomaly of the Mullerian and mesonephric ducts. It is characterized by

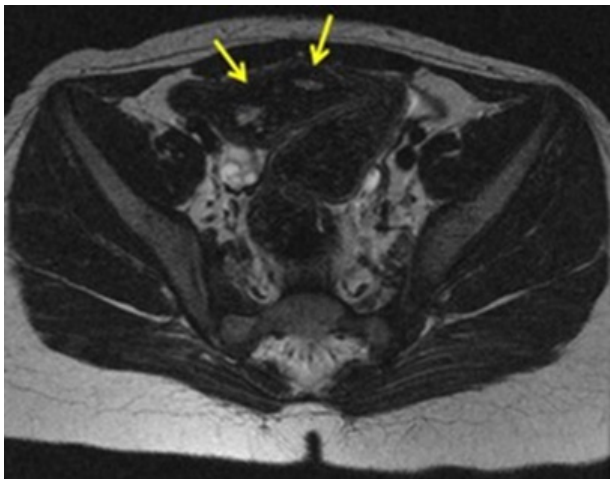

Fig. 3 – Axial T2-weighted image showing two hemi-uterus (arrows).

a didelphys uterus with an obstructed/blind hemi-vagina and ipsilateral renal agenesis,[1] accounting for 0.16-10% of Mullerian duct malformations (MDM).

Patients are usually asymptomatic until puberty, when they begin to have dysmenorrhea and cyclic pelvic pain due to hematometrocolpos, as a consequence of obstructed hemi-vagina. In the postnatal period and early infancy, the same symptoms may occur due to the influence of maternal hormones.

Acute complications include pyohematocolpos and pyosalpinx, and long-term complications include endometriosis, pelvic inflammatory disease, and infertility.[1]

Differential diagnosis includes others MDM (bicornuate uterus, septate uterus), imperforate hymen and transverse vaginal septum.

Treatment is symptomatic, with vaginal septotomy for hematocolpos drainage. Hemi-hysterectomy is not indicated.[2,3]

As there is a strong association of female genital tract and renal anomalies, when the patient presents with one of these, a screening for associated anomalies should be performed.

Received / **Recebido** 04/04/2020
Acceptance / **Aceite** 12/05/2020

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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