Case	(305) A case of ohvira syndrome:
Authors	M. Espinosa De Los Monteros Fernandez, J. Jurado Serrano, E. Merchante Garcia, F. Galvan Banqueri, P. Gomez-millan Ruiz, T. Rocha Iñigo.
Centre	Hospital Virgen Del Rocio.

CASE PRESENTATION

A 20-years-old woman presented with diffuse abdominal pain, dysuria, pollakiuria, and tenesmus for the last 10 days. She had no fever, no vomiting, and no diarrhoea. Last menstrual period was two weeks ago. Personal background:

- Solitary left kidney since born.

- Appendectomized.

The initial diagnosis was urinary infection and the patient was treated with antibiotics with no improvement. Blood tests results were unremarkable. An ultrasound examination was performed (Figure 1), during which left urinary tract dilation and a cystic pelvic mass, with hypoechoic content and no Doppler signal, were observed. These findings were interpreted as a fully distended bladder with hydronephrosis. There was no urine flow after inserting a urinary catheter.

A computed tomography scan (CT) was carried out (Figures 2 and 3), which revealed two uterus (white arrows) next to the cystic pelvis mass, which was located between the bladder and the rectus, with a density of around 50 Hounsfield Units. The right ovary showed a nonspecific calcification (orange arrow), while the left ovary was normal. The right kidney was absent and the left kidney presented a dilated excretory system.

Speculoscopy revealed that the mass was pushing back the anterior wall of the vagina, making impossible the visualization of the cervix. During culdocentesis, three litres of bloody thickened fluid were drained from the cystic mass.

For further evaluation, a magnetic resonance imaging (MRI) was performed. On T2weighted imaging (Figure 4), two vaginas can be seen; one is a blind vagina ending in the right uterus, and the other one connects the left uterus to the vulva.

DISCUSSION

The radiological findings suggest a uterine duplicity disorder known as OHVIRA Syndrome (Obstructed Hemivagina Ipsilateral Renal Agenesis), associating hematocolpos and ureterohydronephrosis secondary to mass effect.

This syndrome, also known as Herlyn-Werner-Wünderlich syndrome, is a rare mullerian anomaly. It consists of uterus didelphys (two uterus and two vaginas), unilateral vaginal obstruction and ipsilateral renal anomaly, most commonly agenesis.

These patients usually present cyclical dysmenorrhea, which later evolves into persistent pelvic pain.

CONCLUSION

Although MRI plays a major role in the diagnosis of congenital uterine anomalies, CT scan usually hints this diagnosis in emergency cases. Also, it is important to remember the strong association between uterine duplicity and renal anomalies.

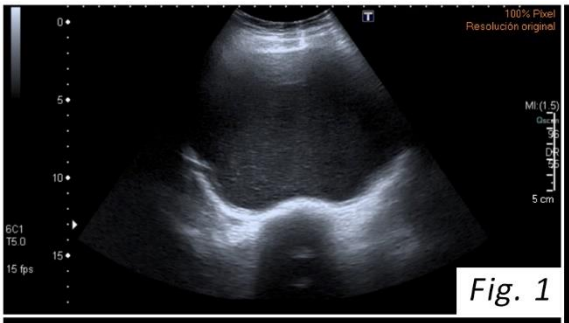

Fig. 1

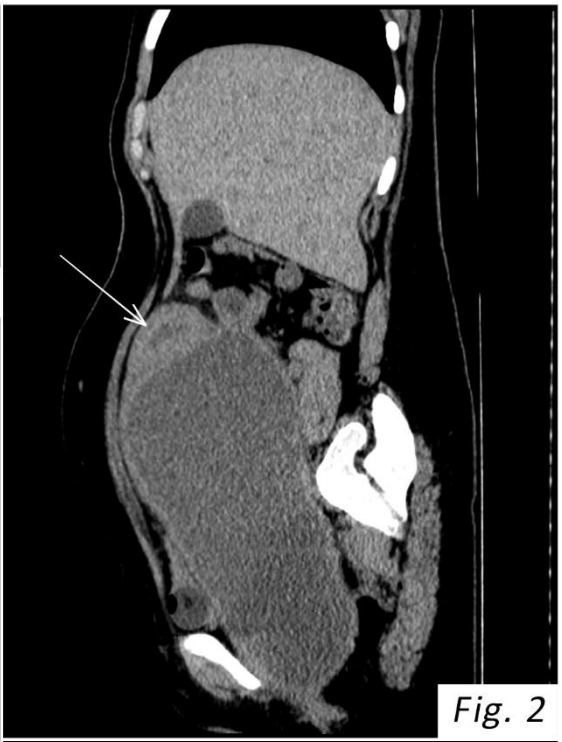

Fig. 2

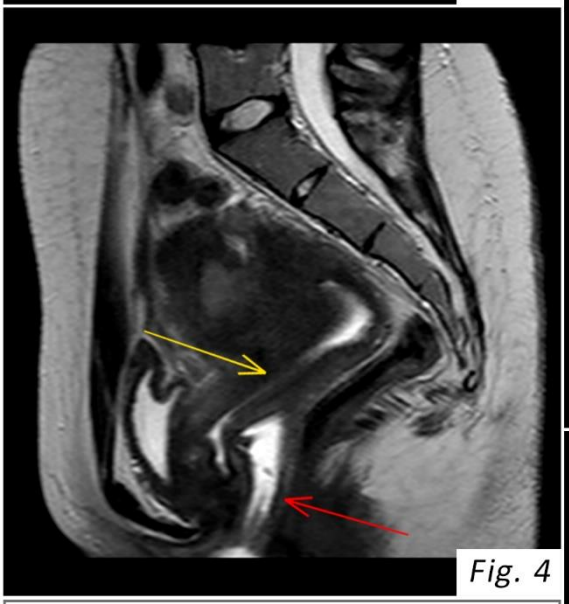

Fig. 4

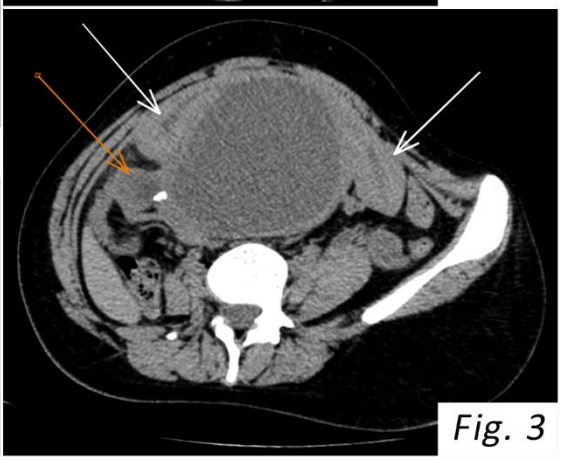

Fig. 3

Figure 1. Sonography, cystic pelvic mass.
Figures 2 and 3. CT scan. Uterus (white arrows) and right ovary (orange arrow).
Figure 4. MRI, T2 imaging. Blind vagina (yellow arrow) and non-obstructed vagina (red arrow).

BIBLIOGRAPHY

- Bajaj S, Misra R, Thukral B, et al. OHVIRA: Uterus didelphys, blind hemivagina and ipsilateral renal agenesis: Advantage MRI. J Hum Reprod Sci. 2012; 5(1): 67–70.

- Huneeus A, Sanz A, Pino V, et al. Sindrome de Hemivagina Obstruida, Utero Didelfo y Agenesia Renal Ipsilateral (OVHIRA), Drenado Parcialmente por Fistula Utero-Uterina Espontanea. REV CHIL OBSTET GINECOL; 2017; 82(6): 595-602.