Laparoscopic nephroureterectomy as treatment in obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome

Maria Medina-González 1, Jorge Panach-Navarrete 1, Lorena Valls-González 1, Ana Castelló-Porcar 1, Jose María Martínez-Jabaloyas 1

1 Department of Urology. University Clinic Hospital of Valencia, Facultat de Medicina i Odontologia, Universitat de València, Spain

ABSTRACT

Introduction: OHVIRA syndrome is a rare entity characterized by renal and Mullerian anomalies. The objective of the video is, through a clinical case, to discuss the importance of diagnosis, management and treatment, to avoid the complications that this syndrome entails, and to improve the long-term prognosis.

Materials and Methods: We report the case of a 10-year-old girl who consulted for abdominal pain, being diagnosed with OHVIRA syndrome. We describe the diagnosis and the surgical technique. In addition, we perform a systematic review in PubMed to report the published literature of this topic and we show the optimal management of this pathology.

Results: This syndrome is characterized by a bicornuate uterus with obstructed hemivagina, renal agenesis or dysplastic and atrophic kidney with ectopic ureter, which drains to the obstructed hemivagina. It causes clinical symptoms such as persistent vaginal drainage after resection of the vaginal septum and cyclic abdominal pain. The diagnosis was completed with a URO-CT scan and magnetic resonance imaging, to identify the anatomical alterations and to plan a correct surgery.

In the video we show a laparoscopic left nephroureterectomy of an atrophic and ectopic kidney without incident as an effective treatment to avoid complications.

Conclusions: In OHVIRA syndrome, if an atrophic kidney exists surgical treatment is recommended, in order to avoid complications. For this, a suitable diagnosis is necessary for a good preoperative plan, as well as the knowledge of the spectrum of renal anomalies for an early diagnosis. We show the indicated procedure, by a laparoscopic approach.

CONFLICT OF INTEREST

None declared.