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Step-by-step robotic technique to manage an extensive retroperitoneal fibrosis

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Rosai-Dorfman disease (RDD) is a rare histiocytic disorder most frequently presenting as bilateral cervical lymphadenopathy in children and young adults. However, extranodal disease may occur, with the retroperitoneum being one of the possible affected sites. Herein, we show an uncommon case of localized retroperitoneal RDD in a 33-year-old female. MRI revealed an extensive amount of thickened retroperitoneal tissue, without contrast enhancement, determining ureteral compression and bilateral hydronephrosis. Assuming it was a case of idiopathic retroperitoneal fibrosis, after placing bilateral ureteral stents, a systemic therapy with rituximab and tocilizumab was administered, but without benefit. A retroperitoneal biopsy was therefore performed, revealing a localized RDD. After multidisciplinary discussion, it was decided to proceed with robot-assisted ureterolysis, preferring not to treat both sides simultaneously. Surgery was carried out with the Da Vinci Si Surgical System, with a 4-arm configuration. The first step of the procedure mainly consists of the medialization of the colon, until anatomical landmarks are recognised, such as the psoas muscle or the gonadal vein. The fourth arm equipped with a Prograsp forceps is fundamental in such cases, to provide good

traction and a better exposure of the surgical field. Isolation of the ureter starts distally from the iliac cross, where fibrosis is less represented, and thus a delicate ureterolysis is performed while proceeding proximally. The presence of the ureteral stent may help in correctly identifying the ureter, while the use of a vessel loop reduces direct ureteral manipulation, thus lowering the risk of iatrogenic damage. Also, in this phase near-infrared fluorescence may be useful to further delineate the course of the ureter even when it is completely embedded in tissue. As the ureter isolation and ureterolysis proceed upwards, the grade of fibrosis may significantly increase. In this case, the proximal ureter and renal pelvis appeared completely embedded in the fibrotic reaction. When the affected segment is correctly identified, careful dissection of the ureter is carried out by splitting the fibrous capsule anteriorly. With the anterior ureter identified, the remaining ureter is freed circumferentially from the fibrous reaction. Intraoperative ultrasonography allows timely recognition of the kidney, ureteropelvic junction, and proximal ureter. Once a delicate excision of adherent tissue embedding the ureter and the kidney has been carried out, the last step consists of robotic omentoplasty.

After proper mobilization, an omental wrap is used to surround the isolated ureter. The importance of the wrap is not clearly demonstrated; in particular, whether it may simply act as a barrier to avoid further compression by fibrosis or play a role in feeding and revascularizing the lysed ureter. From our point of view, this procedure is of utmost importance to provide a new ureteral vascularization and to prevent de novo extrinsic compression.

The total operative time was 240 minutes. No surgical-related complications were reported. The abdominal drainage and catheter were removed on the 3rd postoperative day, and the patient was discharged on the 5th postoperative day with bilateral indwelling ureteral stents. At 3-month

follow-up, serum creatinine was 0.60 mg/dL and the left ureteral stent was removed.

DECLARATION

Informed consent was obtained from the patient. All procedures performed in this study involving human participants were in accordance with the ethical standards of the Institutional and National Research Committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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