



Robotic surgery in patients with achondroplastic dwarfism: evaluation of risks and issues in an anatomical challenging bilateral partial nephrectomy

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Received: 25 September 2018 / Accepted: 2 December 2018 / Published online: 23 January 2019
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Abstract

The reports on the performance of robotic surgery in patients with dwarfism are anecdotal; anesthesiological issues and a challenging anatomy are the main factors that lead most of surgeons to prefer a more traditional approach. We present a case of bilateral robotic partial nephrectomy in a patient affected by achondroplastic dwarfism and aim to evaluate risks and issues in this type of surgery.

Keywords Achondroplasia · Robotic partial nephrectomy · RAPN · Robotic surgery · Dwarfism

Introduction

Robotic surgery is increasingly a widespread practice in many disciplines and for many types of operations and in particular in urology, becoming more and more the technique of choice for prostatectomy and partial nephrectomy [1–3].

The increasing number of procedures annually performed naturally translates in the complexity of patients managed with a minimally invasive robotic approach, such as patients with unfavorable anatomic conditions and complex diseases.

The reports on the performance of robotic surgery in patients with dwarfism are anecdotal [4], both due to the low prevalence of this anatomical condition, and for anatomical features that can lead surgeons to prefer a more traditional approach [5].

Therefore, we aim to present the clinical case of a patient affected by achondroplastic dwarfism submitted to a robotic bilateral partial nephrectomy for bilateral renal tumor to discuss the possible factors that might make this surgical approach challenging.

Case presentation

A 59-year-old achondroplastic dwarf was admitted in June 2018 for bilateral renal mass diagnosed with an abdominal sonography after an episode of macroscopic haematuria. He denied pain, other symptoms or previous significant medical history. He was 129 cm tall and 60 kg heavy with a resulting BMI of 36. Urine cytology was negative and pre-operative routine laboratory tests showed a Hb of 14.9 g/dl and a creatinine of 0.63 mg/dl. The patient underwent an abdominal CT scan in June 2018 which showed the presence of a 35 × 25 × 26 mm renal mass with high contrast enhancement in the anterior margin of the right kidney and another mass in the left kidney with the dimensions of 20 × 19 × 17 mm, both highly suspicious for malignancy (Fig. 1). The PADUA score was 12 for the right mass and 6 for the left. The patient was candidate to robotic-assisted surgery as first choice to take advantage of the minimally invasive approach. However, the patient was fully instructed about the possibility to change to open surgery if needed for both the operations.

The patient underwent a right Robot Assisted Laparoscopic Partial Nephrectomy (RAPN) with the use of the DaVinci Si. The decision to treat first the right kidney mass was based on dimensional criteria as well as on the proximity of the renal hilum.

Under general anesthesia, the patient was placed in a flexed, full flank position. A Hasson technique by transperitoneal approach was used to establish a pneumoperitoneum

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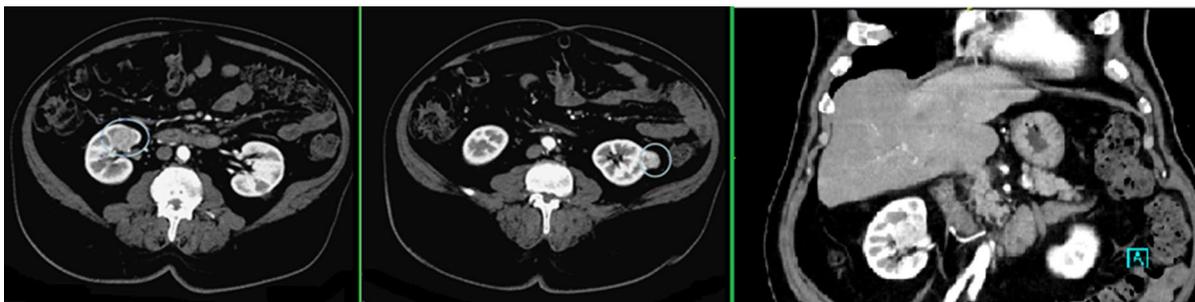


Fig. 1 CT scan showing bilateral renal tumors in achondroplastic patient

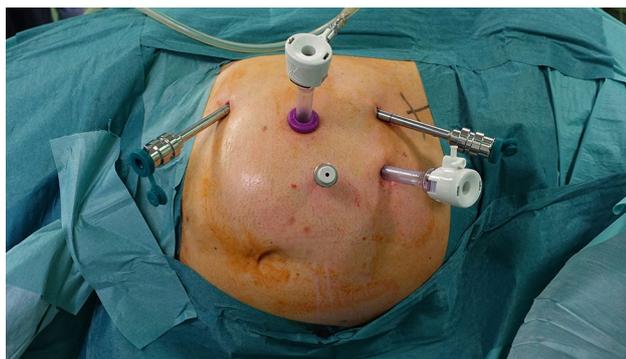


Fig. 2 Trocar placement for the first of the two operations performed

of 12 mm Hg. A 12-mm camera port with 0° lens was placed approximately 12–15 cm laterally to the umbilicus. We decided to place the optical access trocar more sideways compared to our standard placement, to gain space to guarantee an adequate extent of movements to the robotic arms. Under direct vision, the 8 mm robotic trocars were placed: one subcostal and one in the lower quadrant. A 12-mm assistant port was placed in between the robotic trocar sites, and about 1 cm below the plane of the camera port (Figs. 2, 3). Despite the right-sided mass, due to the limited space, we decided not to place the 5 mm subxiphoid trocar usually used to lift up the liver and better expose the kidney. Monopolar scissors, Maryland bipolar forceps and robotic needle driver were the instruments of choice.

The kidney was exposed by incising the peritoneum sharply along the right parietocolic gutter and reflecting the colon medially to provide optimal exposure of the retroperitoneal space. Subsequently, the ureter, the gonadal vessels, and the lower pole of the kidney were identified and retracted laterally. The renal hilum was identified. An intra-operative real-time ultrasound was performed to identify the mass and to define its dimensions. Vascular clamping was performed with bulldog clamps placed by the assistant. The excision of the tumor was then performed following standard oncological principles, preferentially with monopolar scissors and

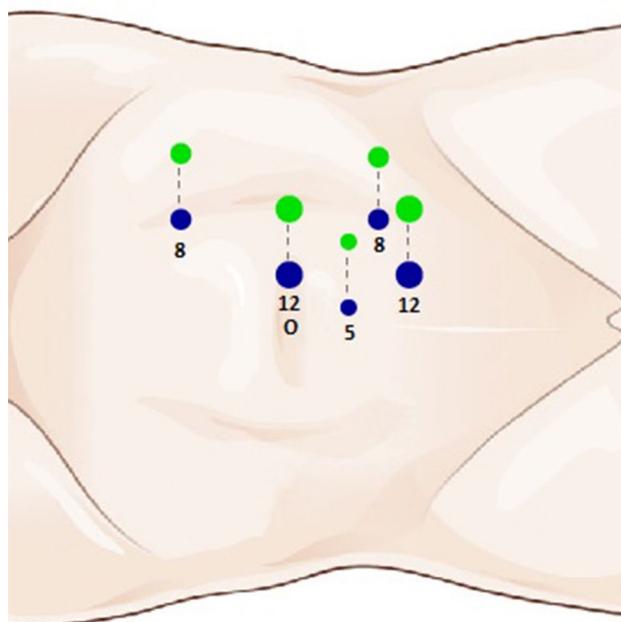


Fig. 3 Modified position of the trocars compared to a standard Robot Assisted Partial Nephrectomy (RAPN) at our Institution. In blue is showed the normal position of trocars for RAPN while in green the modified position for this case

the aid of the assistant's suction to provide opposite traction and to maintain the surgical field dry. The renorrhaphy was performed using a sliding clip technique. A 2.0-braided absorbable suture with 27 mm 5/8 needle was used for the medullar layer. A 0-braided absorbable suture with a 37-mm 1/2 needle was used for the cortical layer. No hemostatic sealant agents were used. Then, the specimen was placed in a retrieval Endo-bag for extraction through one of the trocars. The perirenal fat was repositioned over the kidney and secured by placing a Hem-o-Lock clip, a standard drain was also placed and secured. Finally, wounds were closed with subcuticular sutures. The operative time was about 3 h with an ischemia time of 27 min, and blood loss less than 150 ml. The early postoperative course was regular, without the

development of any complications. Both drain and vesical catheter were removed in the first postoperative day (POD). The patient was discharged in good conditions in the third POD with an Hb of 13.7 g/dl and a creatinine of 0.62 mg/dl. The pathological evaluation showed the presence of a Clear Cell Carcinoma p T1a p Nx R0 Fuhrman grade 2 without necrosis and lymphovascular infiltration.

Due to the success of the previous surgery, a specular operation (Left Robotic Partial Nephrectomy) but with a clamp-less technique was performed 2 months later to remove the left renal mass. The operative time was of 1 h and 20 min with blood loss of 100 ml. The patient was discharged in good conditions in the fourth POD with an Hb of 12.7 g/dl and a creatinine of 0.65 mg/dl. The pathological evaluation showed the presence of Clear Cell Carcinoma p T1a p Nx R0 Fuhrman grade 2 without necrosis and lymphovascular infiltration.

Discussion

Achondroplasia is the most frequent among the over 100 types of dwarfism. It is a genetic disorder caused by mutations in the FGFR3 gene which is responsible for lengthening bones. This mutation leads to a shorter stature, abnormally shaped bones and macrocephalia. While contraindications for minimally invasive abdominal surgery are decreasing, surgery in patients with anatomic deformities such as achondroplastic dwarfs still represents a challenge for most surgeons and anesthesiologists. Very few are reports of laparoscopic or robotic abdominal surgery in these patients [6].

From a surgical point of view, a shorter surgical field might represent an issue, especially for robotic's arms that should have the necessary space to move. However, this is not an absolute contraindication, being slowly increasing the number of robotic partial nephrectomies performed in children and teenagers affected by Wilms' tumors [7, 8]. The obesity, that is often associated in dwarf patients, further complicates the robotic approach for several reasons [9]. These include the potential for restricted instrument range of motion and reach due to a thicker abdominal wall as well as decreased intra-abdominal working space. The accentuated hyperlordosis and hyperkyphosis might also affect the intra-abdominal anatomy and must be considered during trocars insertion. In dwarfs, the thoracic cage is often tilted anteriorly, with a low-lying costal margins that can divide the operative field and is reported to be a substantial risk factor for conversion to a laparotomy when performing laparoscopic surgery [10].

Moreover, a great attention is required also by positioning the patient on the surgical bed, to minimize spine damages and joint contractures that are frequent in these patients.

Achondroplastic dwarfs might be a challenge also for the anesthesiological management. In fact, a reduced size chest and upper airway abnormalities can lead to cervico-medullary compression, restrictive lung disease, difficult mask ventilation and intubation. These patients have also a tendency to hypersalivation, sleep apnea and a tenfold increased cardiovascular risk [11]. For these reasons is usually recommended to schedule an extended stay in the anesthetic recovery room.

Our case, like a previous one described in the literature [3], showed the feasibility of robotic partial nephrectomy in patients affected by achondroplastic dwarfism. The two operations were performed with the Si system. It is reasonable to speculate that the use of the Xi system could facilitate the adoption of the daVinci robot in such cases with limited working space. A detailed study of the clinical case and of the surgical anatomy through CT imaging, a planned and modified positioning of the trocars and a multidisciplinary pre- and post-operative care are mandatory to perform a successful and safe surgery in such challenging cases.

Compliance with ethical standards

Conflict of interest Authors declare that they have no competing interest.

Informed consent Written informed consent was obtained from the patient for publication of this case report/any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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