Partial nephrectomy in a patient with dwarfism

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We describe the case of a 50-year-old male with achondroplastic dwarfism who presents with a renal mass in his left kidney concerning for renal cell carcinoma. The patient successfully underwent a robotic partial nephrectomy, which revealed a T1a renal cell carcinoma. The tumor was excised successfully without any intraoperative complications demonstrating that a robotic partial nephrectomy is technically both safe and effective in patients with achondroplastic dwarfism.

Key Words: renal cell carcinoma, dwarfism, achondroplasia, kidney tumor, partial nephrectomy

Introduction

The proper evaluation and treatment for renal cell carcinoma (RCC) is imperative for today's practicing urologist. Improved imaging technologies have provided patients with earlier detection of renal masses, often lending to more nephron-sparing surgical procedures with partial nephrectomy as the gold standard for small renal masses. Despite the rising incidence of RCC and subsequent partial nephrectomies, there have been no documented cases of patients with dwarfism treated via robotic partial nephrectomy. Dwarfism is not a single entity, but a condition of short stature with over 200 known causes, most commonly due to achondroplasia.[1] With greater than 35,000 people with dwarfism living in the United States and achondroplasia seen in 1.5 out of 10,000 births, dwarfism, particularly from achrondoplasia, is a patient condition that most physicians have and will work with during their career.[1]

Surgery on this patient population is well documented in other surgical fields, including oophorectomy, and bone lengthening.[2,3] However, the urologic literature on patients with dwarfism is sparse. Gyomber et al described a radical retropubic prostatectomy procedure in an achondroplastic dwarf, but, to our knowledge, partial nephrectomy has not been described in this patient population.[4] Moreover, there are no documented robotic-assisted surgeries of any type in an achondroplastic patient group. Herein, we describe a robotic partial nephrectomy in a patient with dwarfism.

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Case report

A 50-year-old male was referred to our clinic for evaluation of a left renal mass. The patient's pertinent past medical history included achondroplastic dwarfism which resulted in an adult height of 124.5 cm, weight of 53.1 kg, 1 pack per day tobacco use, body mass index (BMI) of 34.26 and a past surgical history of cholecystectomy in 2007. His renal mass was discovered following a computed tomography (CT) scan of the abdomen/pelvis for a complaint of left flank pain. His initial CT scan showed an enhancing, 2.6 cm significantly endophytic lower pole left kidney mass with a RENAL nephrometry score of 7a, suspicious for clinical T1a renal cell carcinoma. The patient elected to undergo surgical excision of the mass via robotic partial nephrectomy. Preoperative vital signs, basic metabolic panel, and complete blood count were within normal limits with a hemoglobin count of 15.8, hematocrit count of 46.5 and creatinine of 0.6. Other metastatic work up including a chest x-ray was negative. Imaging did not suggest any significant scoliosis or kyphosis.

The patient was then taken to the operating room. Extra measures were taken to ensure that adequate padding and securing of the patient's pressure points and extremities were achieved in a safe manner. He was placed in the standard left lateral decubitus position with slight table flexion, Figure 1. Trocar placement was uncomplicated and achieved using a medial arrangement with the standard robotic partial nephrectomy template with one 12 mm camera port, three robotic arm ports and one 12 mm assistant port, Figure 2. Using the da Vinci Si surgical system (Intuitive Surgical Inc., Sunnyvale, CA, USA), the left lower pole renal mass was then successfully excised via a transperitoneal approach and renorrhaphy was performed. A frozen section of the deep margin was negative. Total warm ischemia time was 32 minutes. There were no intraoperative complications, and estimated blood loss was 300 cc.

Figure 1. Patient is placed in the standard left lateral decubitus position with slight table flexion. Adequate padding is placed and the patient's pressure points and extremities are secured.

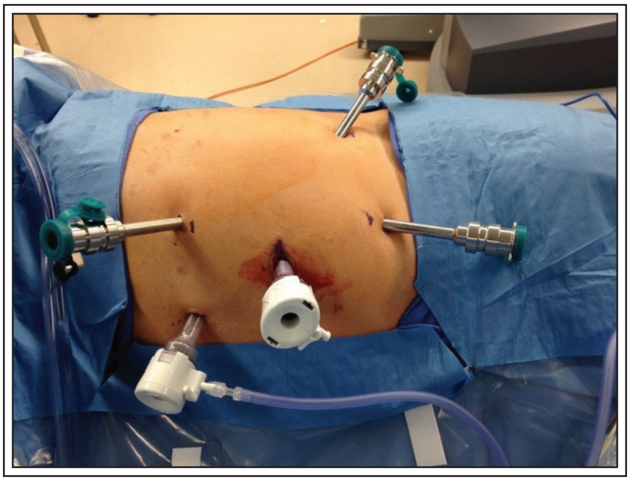

Figure 2. Trocars were placed in a medial arrangement using the standard robotic partial nephrectomy template with one 12 mm camera port, three robotic arm ports and one 12 mm assistant port.

Postoperatively, there were no neuromuscular injuries. Pain was well controlled and his diet was advanced in the usual fashion. He did, however, have a prolonged course of gross hematuria per Foley catheter, for which a CT urogram of the abdomen/pelvis with unenhanced phase with contrast enhanced nephrogram and excretory phase was obtained. The CT scan ruled out any evidence of arteriovenous malformations, extravasation, or fistulas. His hematuria spontaneously resolved with medical management and his hemoglobin remained stable throughout the episode. The patient's catheter was removed at the resolution of his hematuria, and he subsequently voided without difficulty. His postoperative creatinine peaked at 0.95 with a nadir of 0.66, which was his baseline preoperative creatinine.

Pathology revealed clear cell renal cell carcinoma with Fuhrman grade 1 and negative margins. The tumor measured 3.0 cm x 2.5 cm x 2.0 cm and was confined to the kidney, consistent with pathologic T1a staging. At his 3 month follow up in clinic the patient had returned to a normal state of health. He had well-healed incisions and denied flank pain, hematuria, and gastrointestinal complications. The patient will be monitored in accordance with AUA and National Comprehensive Cancer Network guidelines.

Discussion

In evaluating the safety of a partial nephrectomy in a patient with dwarfism there are several factors to consider. Tolerance of insufflation and pneumoperitoneum during the procedure is important in the achondroplastic population, as the presence of pneumoperitoneum induces pathophysiologic changes in several ways.[5] In normal physiologic state, carbon dioxide absorption causes hypercapnia and acidosis, whereas elevated intra-abdominal pressure causes attendant changes in respiratory parameters including elevated peak airway pressures, decreased vital capacity, and decreased compliance.[5] In patients with healthy lungs, the physiological effects of this process are benign, however, patients with lung disease are at risk for significant hypoxemia.[5] The complex anatomy of patients with achondroplastic dwarfism exacerbates these hemodynamic stresses. For example, rib hypoplasia leading to thoracic cage dystrophy, progressive kyphosis, scoliosis and thoracic lordosis are all common pathologic findings experienced by the achondroplastic dwarf phenotype that lead to restrictive lung disease.[1] Moreover, anatomic upper airway obstruction in these patients is common, owing to craniofacial bone and soft tissue abnormalities.[1,6] Although our patient tolerated pneumoperitoneum without complication, the strong association of anchondroplastic dwarfism with restrictive and obstructive lung disease must prompt adequate evaluation of pulmonary function prior to surgery.

The cardiovascular effects of pneumoperitoneum also play an important role when assessing the feasibility of robotic surgery in a patient with dwarfism. Pneumoperitoneum increases mean arterial pressure, systemic and pulmonary vascular resistances, right atrial pressure, and pulmonary capillary wedge pressure, while decreasing cardiac output.[5] These hemodynamic changes are especially relevant as pulmonary hypertension, and subsequent right-sided heart dysfunction, is the most common cardiovascular pathology in this population.[1]

Overall, however, achondroplastic dwarfism should not be considered an absolute contraindication to robotic-assisted procedures, as evidenced by the total absence of intraoperative cardiopulmonary complications during the patient's robot-assisted partial nephrectomy.

Another consideration is the feasibility of the surgical approach of the robotic partial nephrectomy in patients with dwarfism. In evaluation of our patient, there was concern of sufficient room for proper port placement for the four trocars and assistant port that are required to perform a robotic partial nephrectomy. The two most common trocar approaches described in the literatures are the lateral and medial camera approaches.[7] The medial approach, which we selected to use on our patient, provides the surgeon with maximized visualization and tracking of instruments passed by the assistant. The lateral approach, in comparison, has a potential advantage of having a closer view of the kidney at the expense of a more limited views of the surgical field and instruments as the assistant passes them.[7] The total operative times had been reported to be comparable between the two camera approaches.[7] In an achondroplastic patient with an already complex anatomy, we felt that the medial approach was the safest and most effective means of performing the surgery. There was also a concern regarding adequate working space during the surgery. In the case report by Gyomber et al, the pelvis of the achondroplastic patient was too small to perform a laparoscopic or robotic radical prostatectomy.[4] Unlike the pelvis, however, the abdominal space was sufficient for proper robotic arm placement and the standard robotic surgical approach was feasible.

When considering the feasibility of robotic-assisted laparoscopy in patients with dwarfism, the pediatric literature provides an excellent surrogate for the achondroplasia population with respect to size and weight limitations and smaller pelvic and abdominal spaces. Ballouhey et al demonstrated that pediatric patients weighing less than 15 kg had slightly higher risk of conversion to open (5%) compared to heavier pediatric patients who on average weighed 30.2 kg (2%). Nonetheless, they still concluded that robotic-assisted laparoscopy was feasible in even the smaller of patients.[8] Another study based feasibility of robotic surgery on distances between the anterior superior iliac spines (ASIS) and the puboxyphoid (PXD) distance. The study demonstrated that pediatric patients with ASIS distances > 13 cm or PXD distances > 15 cm had significantly less robotic collisions as well as shorter operative times.[9] Our patient met all of these criteria and by pediatric surgery standards was eligible for a robotic approach to his partial nephrectomy.

Positioning was also a concern, as there have been multiple reported complications in patients in the lateral decubitus position undergoing urologic laparoscopic procedures. Complications from positioning include rhabdomyolysis, abdominal wall neuralgias, and sensory and motor deficits.[10] Moreover, neuromuscular injuries are two times more common for urologic patients undergoing upper retroperitoneal laparoscopy than pelvic, with all injuries other than abdominal wall neuralgias most likely due to prolonged positioning.[10] Achondroplastic patients are prone to developing thoracic lordosis, lumbar kyphoscoliosis, and cervical scoliosis, putting them at an especially increased risk for neuromuscular injuries.[1] Our patient, however, safely underwent laparoscopic surgery in the lateral decubitus position and avoided any neuromuscular injuries. This demonstrates that attention to standard safety strategies such as meticulous padding of pressure points, proper positioning, and minimal table flexion can mitigate this risk. For example, partial table flexion versus full table flexion has been associated with lower interface pressures, and consequently less position-related complications.[11] Also of note, with a BMI of 34.26, our patient is categorized as obese. While obesity may be a relative contraindication to robotic surgery, recent literature suggests obesity does not affect robotic partial nephrectomy outcomes (operation duration, warm ischemia time, and postoperative complication rates) up to a BMI of 60. We found this to be true in our patient, as his BMI did not significantly impact patient positioning or affect the operation.[12]

Although this case was a technical success, there was a postoperative complication of gross hematuria. Imaging failed to identify the etiology and his hematuria eventually resolved spontaneously with expectant management, thus classifying this incident as a grade I surgical complication on the Clavien-Dindo scale. In addition, the patient lost 300 mL of blood. Though our patient maintained hemodynamic stability throughout the procedure and did not require any blood transfusions, in smaller patients with achondroplasia a blood loss of that magnitude might represent a larger fraction of total blood volume and may require a transfusion. Also of note, the warm ischemia time of 32 minutes was lengthy, however this can be explained by the significantly endophytic and complex nature of the renal mass.

In conclusion, this is a case report of both a robotic procedure and a renal procedure in a patient with achondroplastic dwarfism. With no technique alterations or major complications, we demonstrated that robotic partial nephrectomies are both effective and safe within the dwarf population. It must be noted that due to associated cardiopulmonary comorbidities associated with achondroplasia a thorough preoperative evaluation of these patients is imperative, but that robotic surgery is not absolutely contraindicated and in select cases may be performed safely with satisfactory oncologic outcome. ☐

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