

Laparoscopic Nephrectomy in a Patient With Situs Inversus Totalis: First Reported Case

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ABSTRACT

Situs inversus totalis is a rare congenital condition. It is characterized by left-right transposition of the internal viscera, including the great vessels, liver, and spleen. Previous reports of radical nephrectomy for suspicious renal masses in these patients have all been performed with an open surgical technique. The present case is the first known report of laparoscopic nephrectomy for treatment of solid renal mass in an individual with situs inversus. The surgery was performed safely following proper preparation and careful dissection of the altered anatomy.

INTRODUCTION

Situs inversus is a rare congenital anomaly in which the thoracic and abdominal viscera are inverted along the midline sagittal plane, thus giving a mirror image of the normal, situs solitus position of the internal organs. First described in the late 1700's by Scottish physician and pathologist Matthew Baillie, situs inversus remains an uncommon phenomenon. It has an incidence of less than 1 in 10,000 people, which is approximately 0.01% of the United States population [1,2]. The totalis form, also known as situs inversus with dextrocardia, refers to the position and apical direction of the heart within the right chest. The totalis form is more common than the incompletus form, in which the heart is located conventionally on the left side [3]. Situs variants have been associated with cardiac, pulmonary, and renal anomalies, including an autosomal recessive form of situs inversus totalis and polycystic disease of the kidney and pancreas. Although renal anomalies including a horseshoe kidney and renal dysplasia are associated with situs inversus

and are known risk factors for renal cell carcinoma (RCC), no connection between situs inversus and RCC have been elucidated and there does not appear to be any increased incidence of malignancy [4].

Primary renal malignancies affect approximately 55,000 new individuals annually in the United States [5]. In the 4 published reports of renal masses in individuals with situs abnormalities, the patients were all managed with an open surgical approach [6-9]. However, with the advent of laparoscopy by Clayman in 1991 as a novel approach to treatment of renal malignancy, laparoscopic nephrectomy has become the standard of care for most localized RCCs. This surgical technique provides equivalent oncological outcomes with less morbidity (eg, decreased blood loss, shorter hospitalization, less analgesia) when compared with the open approach [10-12]. Although there are reports of laparoscopy and hand-assisted donor nephrectomy in patients with situs inversus in both general surgery and transplant surgery literature, no known cases using laparoscopy for renal malignancy have been published to date.

KEYWORDS: Laparoscopic nephrectomy; Surgical technique; Situs inversus

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Abbreviations and Acronyms

COPD = chronic obstructive pulmonary disease

RCC = renal cell carcinoma

CASE REPORT

The patient was a 70-year-old obese female with a past medical history of hypertension, coronary artery disease, type-2 diabetes mellitus, and chronic obstructive pulmonary disorder (COPD). She initially presented to her primary physician with complaints of left flank pain.

Evaluation

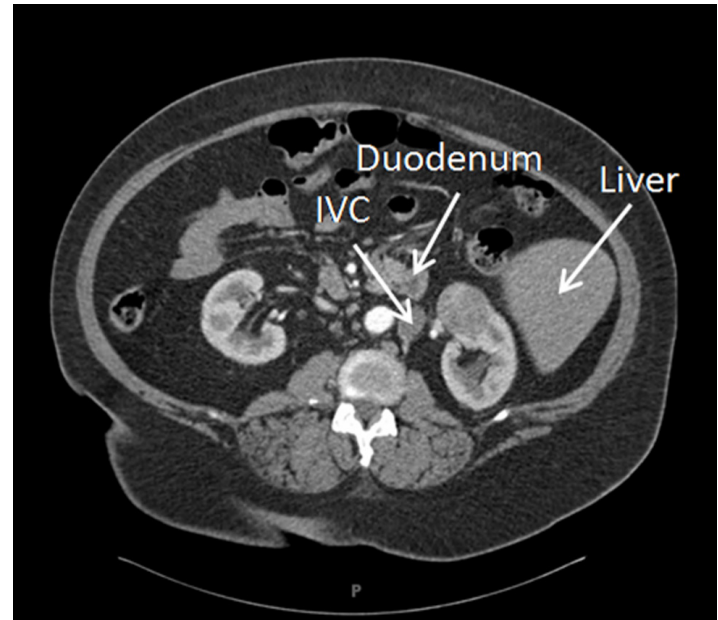
Ultrasound imaging revealed a solid renal mass, and subsequent 3-phase, renal protocol computed tomography (CT) imaging demonstrated a 4 cm x 3.4 cm left-side, anteromedial upper pole, solid enhancing mass abutting the renal hilum (Figure 1). No local invasion or renal vein involvement was identified; however, incidental finding of situs inversus totalis was found. Metastatic workup (including chest x-ray, bone scan, and serum alkaline phosphatase) was negative for distant disease.

Management

After a discussion of the risks and benefits of multiple treatment options, the patient elected to undergo a laparoscopic radical nephrectomy. Surgery was conducted with the patient in the left lateral decubitus position. Initial insufflation of the abdomen was performed using the Veress needle technique. The needle was placed in left lower quadrant to avoid inadvertent injury to the liver in the left upper quadrant. After gaining an adequate pneumoperitoneum with 15 mm Hg, 3 nonbladed 10 mm trocars (Ethicon Inc; Somerville, NJ, USA) were placed in a typical triangular pattern. The medial camera port was placed 6 cm lateral to the umbilicus due to the patient's morbid obesity. The anatomy was disorienting; a mirror image of the right hemiabdomen was on the left side. Essentially, the procedure progressed in the same manner as it would for a "right" nephrectomy, despite being located on the left side. The liver overlaid the anterocephalad aspect of the kidney. To facilitate dissection, an additional 5 mm trocar was placed in the upper midline for liver retraction. The ascending colon was mobilized medially and the duodenum was reflected using the Kocher maneuver, which was similar to a right-sided procedure. A Harmonic scalpel was used for dissection. A left-sided inferior vena cava and a left gonadal vein that inserted into the anterior cava were identified after tracing the ureter cephalad (Figure 2). The renal vein and artery were then exposed and divided separately with a laparoscopic stapler. The renal vein was longer than would be expected for this "right" nephrectomy and eased exposure. After the hilum was divided, the kidney was completely mobilized in a standard fashion, sparing the adrenal gland. The kidney was removed via a muscle-splitting 6 cm, left lower quadrant, diagonal incision, which extended the left lower quadrant port-site incision. This location was selected for extraction due to the patient's obesity.

Figure 1. Contrast Computed Tomography of the Abdomen Revealing a Left Enhancing Renal Mass and a State of Situs Inversus.

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The liver and inferior vena cava (IVC) can both be seen on the patient's left.

The extraction incision was closed in 3 layers with 0-Maxon followed by 3-0 and 4-0 Vicryl sutures.

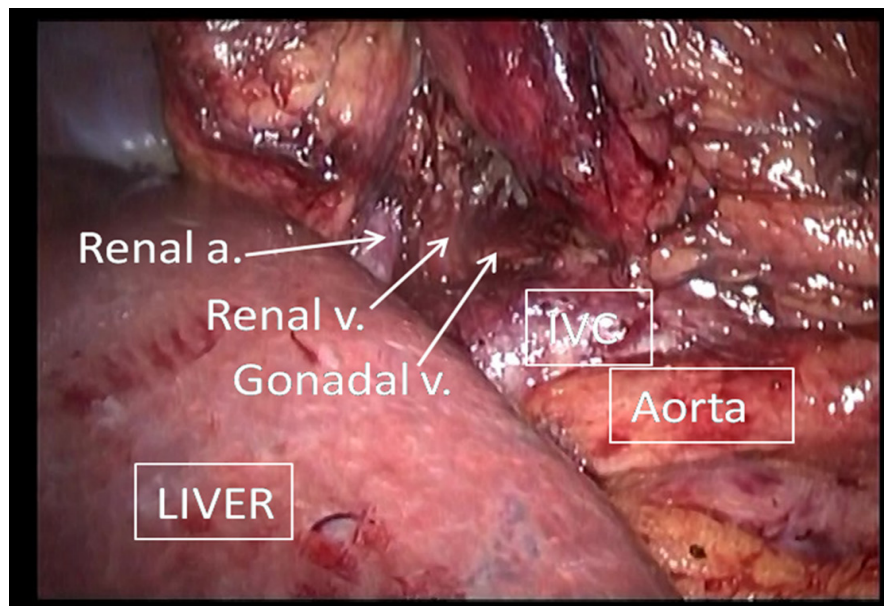
Total operative time was 150 minutes. The estimated blood loss was < 100 mL, and a total of 1000 mL of IV fluids were given intraoperatively. The postoperative course was uneventful and the patient was discharged to her home on postoperative day 2. Final pathology revealed pT1aNxMx, Fuhrman grade 2, RCC, chromophobe type.

DISCUSSION

The present case is the first known use of laparoscopy without hand-assistance to remove a renal mass in a patient with situs inversus. In general, laparoscopy remains a technically challenging procedure that requires the surgeon to operate in 3 dimensions while viewing the operative field in only 2 dimensions. Additional technical demands are placed on the surgeon with the disorienting mirrored view of situs inversus. The misplaced structures result in an increased risk of iatrogenic injury. A previously reported donor nephrectomy case [13] was performed on a right (ie, true left) kidney using a hand-assisted approach. Although that case had the inversus on the opposite side from the present case, similar difficulties were faced due to

Figure 2. Intraoperative Appearance of the Reversed Vasculature With the Vena Cava Positioned to the Left of the Aorta

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The liver is also present on the left side, which creates a confusing image.
Abbreviations: a, artery; v, vein; IVC, inferior vena cava.

the disorienting and mirrored anatomy. No operative times or specific challenges were stated in the donor nephrectomy case, so it remains unclear whether the anatomy lengthened the surgery as it did in the present experience.

A laparoscopic nephrectomy was selected for the present patient because the surgeons felt that the potential for complications from a large flank incision in an obese patient with COPD would be too high and overly morbid. By performing a laparoscopic extraction of the kidney and mass, the surgeons avoided a large, painful, muscle-splitting incision that would have caused splinting and potential postoperative side effects such as pneumonia and atelectasis. Furthermore, the authors have extensive experience with successful laparoscopic surgeries on obese patients with many comorbid conditions. Therefore, a laparoscopic nephrectomy was selected as the procedure with the lowest potential for pulmonary side-effects. It also provided the fastest recovery, as evidenced by the patient's discharge from the hospital on the second postoperative day without any complications.

As with any oncologic procedure, prior preparation with interpretation of axial imaging facilitates planning of the surgical approach. Although the approach is seemingly routine,

the reverse anatomy of situs inversus requires modification of the surgical approach and port-site placements. In other words, for a left-sided tumor, port placement and surgical dissection should be carried out in manner similar to that which is done for a right nephrectomy. This was most apparent during the mobilization of the left kidney. The correct approach is the key to performing successful and expeditious renal surgery in the patient with situs inversus.

Clear cell variant is by far the most common type of renal neoplasm, responsible for 65% to 75% of all cases. However, the present patient's pathology revealed chromophobe RCC, a less frequent (approximately 5%) variant arising from the collecting duct. This variant has a lower malignant potential relative to the clear cell subtype. It remains unclear whether patients with situs inversus are more likely to develop low-grade and low-stage kidney disease.

CONCLUSION

Despite the anomalous anatomy and increased technical challenges, laparoscopic nephrectomy is feasible and effective in patients with a suspicious renal mass and situs inversus. Although past reports have been managed with an open surgery technique, laparoscopy can be performed safely with

proper preparation and careful dissection of the altered anatomy. The take-home message is to modify the surgical approach so that the left-sided kidney tumors are treated like right-sided tumors, and vice versa.

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