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Radical laparoscopic nephrectomy for a huge left renal mass in situs inversus totalis: A case report and video presentation

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ABSTRACT

This case report describes the successful laparoscopic radical nephrectomy in a 69-year-old woman with situs inversus totalis and a large, hypervascular left renal mass. Despite the presence of metastatic disease, the patient underwent neoadjuvant immunotherapy followed by surgery. The successful laparoscopic approach highlights the feasibility of minimally invasive surgery even in patients with complex anatomical variations. This report provides valuable insights into the surgical management of renal tumors in individuals with situs inversus totalis.

1. Introduction

Situs inversus totalis (SIT) is a congenital abnormality characterized by a mirror-image transposition of both the abdominal and thoracic organs. Various malformations have been associated with SIT, including cardiac, splenic, and gastrointestinal malformations.[1] The incidence of SIT is approximately one in every 5000 to 20,000 live births. SIT is typically an incidental finding and presents asymptomatically. Despite its historical documentation, this anomaly remains underrecognized within the medical community due to its rare occurrence.[2]

While the exact cause of situs inversus totalis is not yet understood, a mutation in the DNAH11 gene has been identified as a contributing factor in at least one form of the condition.[3] Additionally, a potential links between situs inversus and other genetic components, including lefty genes, nodal genes, ZIC3, ACVR2B, and Pitxz.[4] At the time of gastrulation, latality is established at an early stage of development during the formation of the primitive streak. A cascade of genes and signaling molecules is required.[5]

Renal cell carcinoma (RCC) is a prevalent form of malignancy that causes a considerable number of deaths worldwide, with 81,000 new cases and 15,000 fatalities occurring each year; and had increased by more than 30 % in the last two decades.[6] There has been a significant increase in RCC incidence in the sixth decade, with almost twice as many cases among men as among women. Most cases of renal cancer are detected incidentally; the classic triad of symptoms associated with renal cancer, i.e., hematuria, flank pain, and palpable mass, is an uncommon finding, usually associated with patients with advanced disease.[7] In this report, we describe a patient with situs inversus totalis and a large, hypervascular left renal mass, as well as a brief review of the relevant literature, the clinical presentation, radiological findings, laboratory findings, and surgical approach.

2. Case report

A 69-year-old postmenopausal woman with a history of diabetes mellitus and hypothyroidism presented to the emergency department with a two-month history of left flank pain. This pain was not associated with hematuria, fever, back pain, weight loss, or night sweats. Her surgical history was unremarkable, her social history was non-contributory, and her family history was devoid of any relevant conditions. On physical examination, the patient was alert, oriented and afebrile, and her vital signs were within normal limits. A local abdominal examination revealed a soft abdomen with no masses. The remainder of the physical examination was unremarkable.

Laboratory evaluation, including complete blood count, serum chemistry, renal function panel, and liver function tests, yielded normal results. An enhanced computed tomography (CT) scan of the abdomen and pelvis demonstrated features of situs inversus totalis. There is a large

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hypervascular locally advanced left-sided renal mass containing a central necrotic component and tiny calcifications, currently measures 10.5 x 10.2 × 12.6 cm compared to 11.7 x 10.3 × 18 cm with extension beyond the posterior pararenal fascia and abutting the left side lower intercostal muscle and left hepatic lobe with minimal invasion. The renal hila exhibit typical vascular anatomy, with a single renal artery and vein present. The left gonadal vein drains into the left renal vein, while the right gonadal vein drains into the inferior vena cava. (Fig. 1(A and B)). There is a 5.8 cm pelvic mass centered at the upper vagina and cervix, which currently no measurable mass could be identified. Stable endometrial thickening measures 1 cm. Biopsies from the kidney and vagina confirmed metastatic clear cell renal cell carcinoma (CCRCC) in both sites.

The patient underwent nivolumab-ipilimumab therapy followed by a left laparoscopic radical nephrectomy. Positioned in the right lateral decubitus position, four trocars were inserted: two 12mm ports for the camera and primary working instrument and two 5mm ports for assistance. Intraoperative findings revealed distorted anatomy with the liver situated on the left side and partial visualization of the left kidney. Following medial mobilization of the colon, the gonadal vessels and inferior vena cava were identified. A lower window was created after identifying the ureter and exposing the psoas muscle. The ureter was retracted, and dissection proceeded superiorly, identifying and controlling the lower pole renal vessels with Hem-o-lok clips prior to reaching the renal hilum. The upper pole dissection proved challenging due to hepatic displacement into the surgical field. Following the incision of the hepatorenal ligaments, an additional 5mm port was inserted for liver retraction. The upper pole renal vessels were then identified, clipped with Hem-o-lok clips, and dissected. The renal hilum was secured using a stapling device. The kidney was wholly mobilized, placed within a laparoscopic bag, and extracted. Total surgical time was approximately 60 minutes (skin-to-skin). No drain was placed. Fig. 2 shows the gross specimen of the left kidney following laparoscopic left radical nephrectomy. Visual documentation of the radiological imaging findings and surgical technique is available in the supplementary material (Video 1).

Postoperative recovery was uncomplicated, and the patient was discharged on postoperative day 3. Histopathological analysis confirmed CCRCC and TNM stage of T3a N0 M1. The patient's adjuvant therapy regimen consisted of first-line ipilimumab and nivolumab, followed by second-line cabozantinib.

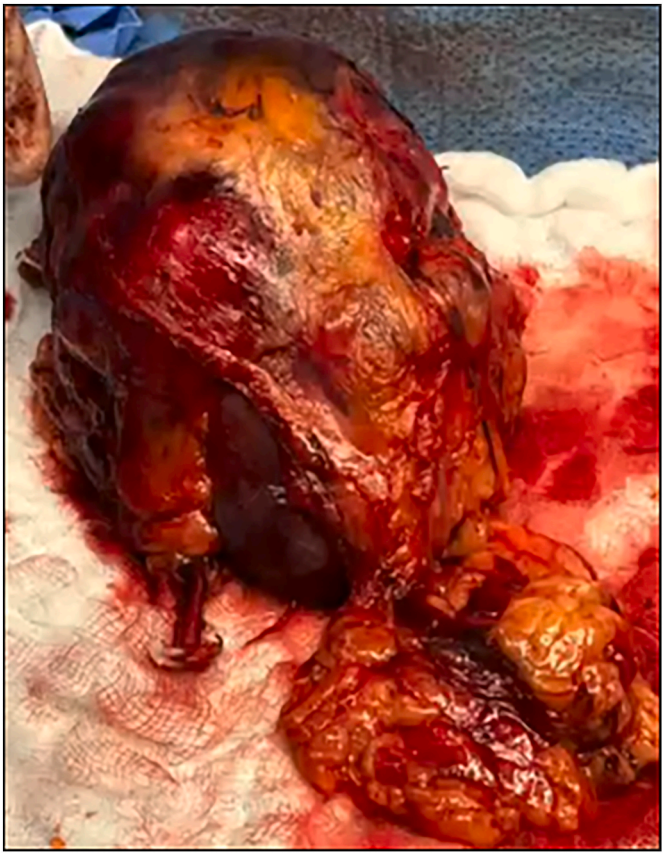

Fig. 2. Gross specimen of the left kidney following laparoscopic left radical nephrectomy.

3. Discussion

Situs inversus totalis (SIT) can present diagnostic and therapeutic challenges, as its presence may complicate the management of concurrent pathologies. Physicians must adopt standard approaches to account for the reversed anatomical orientation of organs, necessitating a

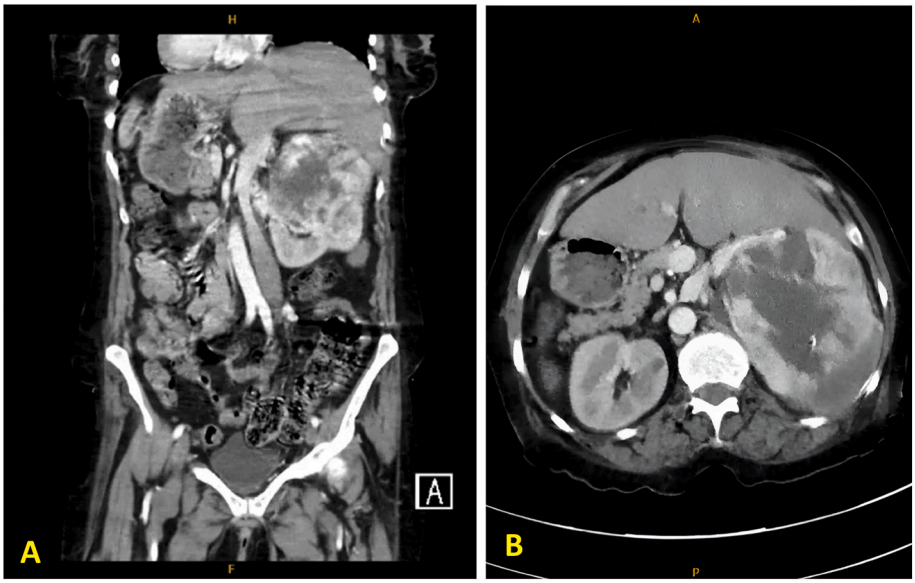

Fig. 1. (A–B). .Abdominopelvic CT scan, both axial and coronal cuts, demonstrates a left renal mass measuring 10.5 x 10.2 × 12.6 cm. Mirror imaging of intrabdominal organs which is a finding consistent with SIT.

thorough understanding of the altered spatial relationships and potential variations in vascular and lymphatic drainage.

The existing literature documents a paucity of cases involving renal cell carcinoma (RCC) in patients with SIT. This case report aims to contribute to the existing body of knowledge regarding this infrequent clinical entity by detailing a case of SIT in a patient presenting with renal cell carcinoma.

Situs inversus can be associated with other congenital anomalies. Renal anomalies, such as dysplasia, agenesis, polycystic kidneys, hypoplasia, ectopia, and horseshoe kidney, are observed with increased frequency in individuals with SIT. Cardiac malformations are another significant association, with reported incidence varying from 8 % to 40 % of SIT cases. Notably, there is no established association between situs inversus and an increased risk of malignancy.[8] This highlights the importance of comprehensive evaluation for associated conditions in patients diagnosed with SIT. Furthermore, approximately 25 % of individuals with SIT are reported to have bronchiectasis, a chronic respiratory condition characterized by abnormal dilation of the bronchi. Kartagener's syndrome, a rare autosomal recessive disorder characterized by the triad of SIT, chronic sinusitis, and bronchiectasis, affects approximately 10 % of individuals with SIT.[9,10]

The incidence of RCC in adults ranges from 2 to 3 % of all malignant diseases. Up to 25 % of patients with RCC exhibit tumor extension into the renal vein and inferior vena cava (IVC), with 1 % of cases demonstrating involvement of the right atrium.[11] This configuration is rarely of medical significance, but it is imperative to understand its unique anatomy when surgical intervention is required.

According to most reports, laparoscopic procedures in patients with SIT have been associated with technical difficulties and longer operating times owing to disorientation caused by reversed abdominal organs, as well as a modification of the surgeon's techniques as a result of the reversed organ position.[12] Chevli et al. employed a hand-assisted laparoscopic radical nephrectomy for RCC in a patient with SIT.[13] They reported that this approach facilitated excellent visualization of the patient's atypical abdominal anatomy, enabling efficient dissection. Their choice of the hand-assisted technique was influenced by surgeon preference, tumor size, and tumor location. While no significant vascular anomalies, aside from venous branching and neovascularization proximal to the kidney, were observed, the expected mirrored lateralization of the inferior vena cava and aorta resulted in a shortened left renal vein. This shortened renal vein, a critical anatomical consideration when accessing the renal hilum, potentially increases the risk of tumor thrombus extension to the inferior vena cava due to the reduced distance. However, no tumor thrombus was present in this particular case, and the procedure was completed without intraoperative or postoperative complications. Zou et al. described a 64-year-old male with localized RCC of the left kidney complicated by SIT and abdominal cocoon.[8] This patient underwent robot-assisted laparoscopic partial nephrectomy. Intraoperatively, significant adhesions were noted between the entire colon and the anterior abdominal wall, leading to the diagnosis of abdominal cocoon. The procedure was completed without incident, with successful resection of the tumor and preservation of the tumor capsule. No intraoperative or postoperative complications, including intestinal injury, were reported, and the patient's postoperative recovery was unremarkable. These cases further support the feasibility and potential benefits of minimally invasive surgical approaches in patients with SIT. The accumulated evidence suggests that, despite the altered anatomy, minimally invasive techniques, when performed by experienced surgeons, can offer outcomes comparable to those achieved in patients without SIT while potentially minimizing morbidity associated with more extensive open procedures. Further research, including larger comparative studies, is warranted to establish the optimal surgical approach in this unique patient population.

4. Conclusion

In conclusion, we reported a rare case of a large renal mass with total situs inversus. The infrequent occurrence of RCC in patients with SIT than managed laparoscopically with proper preoperative preparation and planning. Continued documentation of such cases will enhance understanding and improve surgical strategies for RCC in SIT patients.

CRediT authorship contribution statement

Abdullah Naser A. Alotibi: Writing – review & editing, Writing – original draft, Project administration, Investigation, Formal analysis, Data curation, Conceptualization. **Abdulrahman Aldayhani:** Visualization, Software, Formal analysis. **Ebtesam Almajed:** Writing – review & editing, Writing – original draft, Resources, Methodology, Data curation. **Abdulrahman F. Alruwaily:** Visualization, Validation, Supervision. **Ahmed Alzahrani:** Visualization, Validation, Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2025.103032.

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