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Renal hilar tumor with situs inversus- a case report and systematic review

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A B S T R A C T

Total situs inversus is a rare congenital condition (1 in 25,000) with mirror-image organ positioning, complicating nephrectomies. A 68-year-old female with situs inversus presented with a right renal hilum mass incidentally discovered. Imaging revealed a tumor difficult to distinguish from the renal artery and an elongated right renal vein mimicking left-sided anatomy. Radical nephrectomy achieved favorable recovery and preserved renal function. Reviewing 14 cases (1987–2024), focus has shifted to anatomical variations, especially vascular anomalies, affecting surgical complexity. Preoperative imaging to assess adjacent structures is crucial for optimizing surgical approaches and ensuring patient safety.

1. Introduction

Situs inversus is a congenital autosomal recessive variation,[1] characterized by a mirror-image reversal of internal organs compared to normal anatomical positioning, involving the heart, stomach, liver, spleen, and other organs[2]。 The incidence of total situs inversus is estimated to range from 1 in 6500[3] to 1 in 25,000.[4] Most patients are asymptomatic.[5] However, situs inversus can present significant challenges in surgical procedures, especially when dealing with complex anatomical structures.[6] Since the kidneys are paired organs located within the abdominal cavity, the impact of situs inversus on renal function is generally minimal,[7] partial nephrectomy typically yields favorable outcomes for T1b renal cell carcinoma.[8] However, hilar tumor, located near critical vascular structures such as the renal artery and vein, pose a higher degree of surgical difficulty.[9,10] Additionally, hilar tumors are an independent prognostic factor for recurrence following partial nephrectomy[11] This paper presents a case of a patient with total situs inversus who was diagnosed with a hilar renal tumor and successfully underwent laparoscopic nephrectomy. Through a detailed account of this case, we explore the impact of situs inversus on surgical techniques and share clinical insights into the use of laparoscopic surgery for managing complex hilar renal tumors. We also reviewed all the published articles about patient underwent nephrectomy of renal cell carcinoma and *situs inversus* to understand the prognosis postoperatively.

2. Case report

A 68-year-old female patient was admitted to the hospital due to an incidental finding of a right renal mass during a hospital stay in the orthopedic department 10 days ago. The patient reports no discomfort, fever, pain, or hematuria. She has well-controlled hypertension for 20 years with metoprolol. No history of chronic illnesses or elevated blood glucose treatment.

Enhanced CT showed a right renal mass with associated hydronephrosis, located at the renal hilum, measuring approximately 63.30 × 52.07 mm, with clear margins and heterogeneous enhancement during the contrast phase. Imaging findings were consistent with a malignant renal tumor, staged as T1bN0M0. Multiple small cysts were observed in the left kidney, without any obvious masses. Angiography revealed that the inferior vena cava (IVC) was located to the left of the aorta, and the right renal vein was significantly longer than the left renal vein. The right ovarian vein drained into the right renal vein and then into the IVC (Fig. 1). Preoperative renal function was good, with an eGFR-epi of 94.27 ml/min/1.73 m^2, right kidney GFR of 16.81 ml/min/1.73 m^2, and serum creatinine of 51.01 μmol/L. Cardiopulmonary function was

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normal, and after evaluation, a right nephrectomy was planned.

During operation, the right renal mass was found at the renal hilum, with dense adhesions to the surrounding fat and tissues. The IVC was located to the left, and the right renal vein was notably longer. The right ovarian vein traveled alongside the ureter and drained into the right renal vein before flowing into the IVC. Intraoperative blood loss was estimated at 220 ml, and the surgery lasted approximately 100 minutes. The resected specimen measured 10 x 6 × 5 cm, and the tumor was approximately 5 cm in diameter, located at the renal hilum. On post-operative day 5, the retroperitoneal drain was removed without any incidents.

On postoperative day 7, routine blood tests were normal, with an eGFR-epi of 83.93 ml/min/1.73 m^2 and serum creatinine of 64.98 μmol/L, the patient has been discharged following a successful course of treatment. After the three-months follow-up, both eGFR-epi and serum creatinine were within normal ranges.

3. Discussions

Total situs inversus is a rare anatomical variation with an incidence of approximately 1 in 25,000. In nephrectomy procedures, its primary impact is on the identification and exposure of anatomical structures during surgery. In this case, we observed that the right renal vein was significantly elongated and received drainage from the right gonadal vein, a configuration typically seen in the left kidney of individuals with normal anatomy, which was also reported in other cases of situs inverse.[12,13] The tumor, located at the renal hilum, almost completely occupied this region, and imaging revealed that the tumor was difficult to distinguish from the renal artery. Under these circumstances, the risks of partial nephrectomy were high, and preoperative renal function assessment showed that the GFR of the right kidney was only 16.81 ml/min/1.73 m^2. Considering all factors, radical nephrectomy was deemed to provide the greatest benefit for the patient, as evidenced by postoperative recovery of renal function and changes in drainage output.

Despite the *situs inversus* causing the vascular distribution of the right kidney to resemble that of a normal left kidney, the anatomical structure is quite different from the normal mirrored arrangement, Anatomically, the right renal artery originates from the anterolateral aspect of the aorta, coursing to the right kidney posterior to the inferior vena cava, while the left renal artery arises from a slightly higher and more lateral position on the aorta, running almost horizontally to the left kidney.[14] In our case of total situs inversus, despite the left renal artery originating from the anterolateral surface of the abdominal aorta and passing postero-inferior aspect of the inferior vena cava and renal vein towards the left kidney, and the right renal artery arising from a more lateral position on the aorta and running horizontally to the right kidney, the left renal artery arises at a higher level than the right, which is consistent with the typical anatomical pattern in normal individuals (Fig. 2). However, a case also reported[15] that the left renal artery being positioned lower than the right renal artery.

The heights of the renal hila and the upper poles of both kidneys were nearly identical, although the tumor may have caused the lower pole of the right kidney to be slightly lower than that of the left kidney. This finding contradicts the traditional anatomical view that the right kidney is generally positioned lower due to pressure from the liver. Further basic research may be needed to explore this phenomenon. Additionally, the longer right renal vein and its greater distance from the diaphragm likely made surgical mobilization and resection of the kidney easier.

According to our literature review, from 1987 to 2024, 14 cases of situs inversus patients with renal cell carcinoma undergoing nephrectomy have been reported previously (Table 1). Of these, 5 involved the right kidney, and 9 the left; 6 were laparoscopic, while 8 were open surgeries. Blood loss was reported in only 3 cases, showing considerable variation. One report described a robot-assisted partial nephrectomy for a T1b renal hilar tumor,[16] but long-term follow-up data is lacking, further studies are needed to assess the long-term safety of this

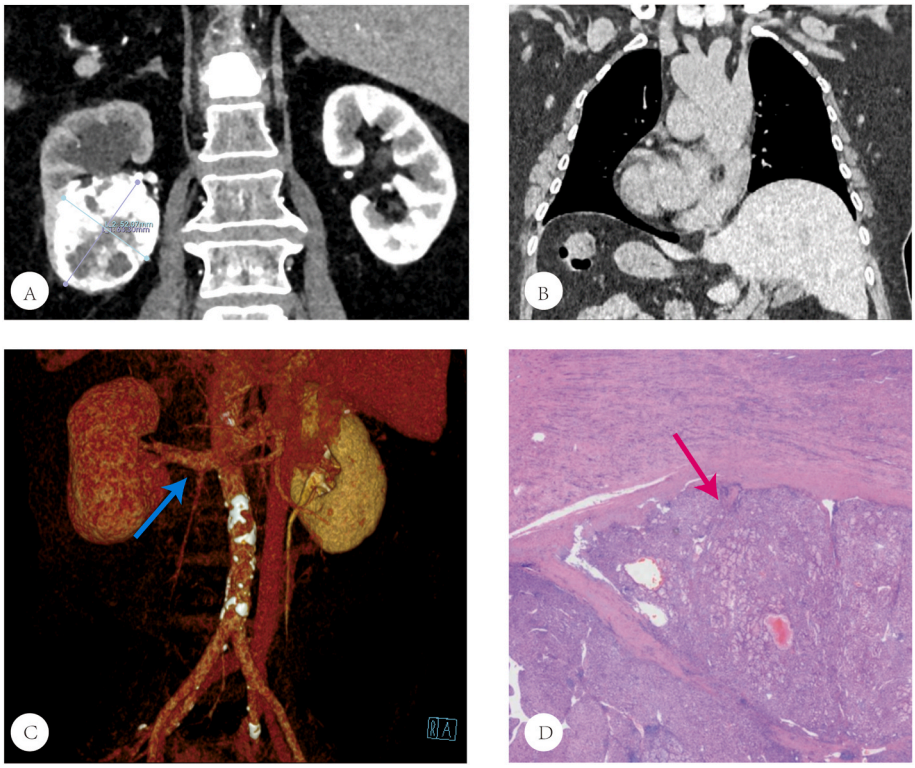

Fig. 1. A. Tumor at the right renal hilum with hydronephrosis; liver on the left. B. Dextrocardia. C. Right gonadal vein (blue arrow) drains into the IVC via the renal vein. D. Clear cell renal carcinoma (red arrow) on pathology.

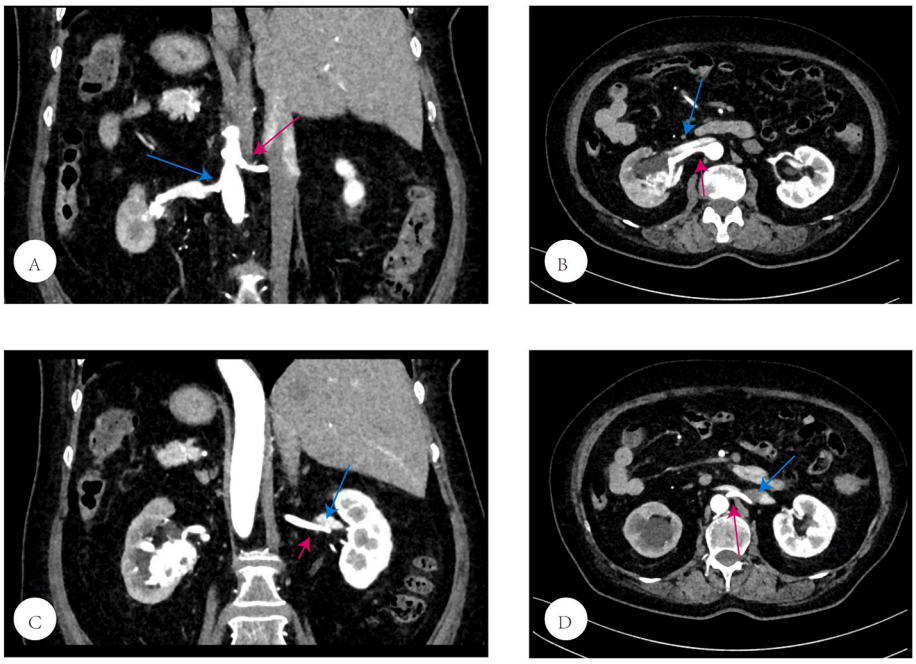

Fig. 2. A. Left renal artery (red arrow) higher than right (blue arrow). B. Right renal artery (red arrow) behind vein. C. Left renal artery (red arrow) below left vein (blue arrow). D. Left renal artery (red arrow) behind IVC (blue arrow).

Table 1
Cases with situs inversus undergoing nephrectomy for renal cell carcinoma.

Case	Age/ Gender	Tumor Side	RENAL Score	Pathological Diagnosis	Surgical Method	Surgical Approach	Renal Vascular Variation	Blood Loss (ml)	Postoperative Prognosis
Bertini et al., 1987	82/F	right	–	–	Radical	Open	–	–	2 months metasis
Treiger et al., 1993	82/F	left	–	–	Radical	Open	lower origin of left renal vein	–	10 weeks dead
Adler et al., 1998	58/M	left	–	–	Radical	Open	–	–	–
Jimenez et al., 2000	43/M	right	–	adenocarcinoma	Radical	Open	–	–	2 months 55Gy radical radiotherapy
Jewell et al., 2001	43/M	left	–	Clear-cell	Radical	Open	–		9 days bilateral lower limbs dry gangrene
Takagi et al., 2008	50/M	left	–	Clear-cell	Radical	Open	–	–	–
Terakawa et al., 2013	81/M	left	–	Clear-cell	Radical	Lap	–	10	discharged uneventfully
Oake et al., 2017	65/M	left	–	clear-cell	Radical	lap	2 renal veins	–	no recurrence
Itoet al, 2019	80/M	right	–	Clear-cell	Radical	Lap	right gonadal and adrenal veins to right renal vein.	–	9+ months no recurrence
Chevli et al., 2019	70/F	left	–	clear cell	Radical	Lap	neovascularization near the kidney, and mirror-image lateralization of IVC and aorta.	–	no issues during routine postoperative follow-up visits
Dergamounet al, 202011	50/M	right	6a	–	partial	open	–	–	1+ years without abnormalities
Wu et al., 2020	48/F	right	–	clear cell	Radical	open	renal vein dilation, cancer thrombi, and mirror-image organ placement	–	6 months dead
Zou et al., 20231	64/M	left	7	clear-cell	partial	lap	Three arteries: one from aorta to lower pole, two from renal artery to middle/lower pole.	200	10 months without abnormalities
Yazaki H et al., 2023	59/M	left	9	papillary renal cell carcinoma type1	partial	lap	IVC and renal artery fully mirrored	40	6 months without abnormalities
our study	68/F	right	10p	clear-cell	Radical	lap	right gonadal and adrenal vein to right renal vein. the origin of renal artery quite different form mirrored normal people	100	3 months without abnormalities

Footnotes M: male, F: female, "-" indicates not available.

approach.

Patients with situs inversus frequently exhibit vascular anomalies such as multiple renal arteries, mirrored placement of the IVC and aorta, and abnormal veins return (e.g., gonadal or adrenal veins), complicating nephrectomy procedures. Interestingly, as time has passed, reports of these vascular variations have become more frequent, likely due to advancements in imaging and surgical techniques that allow for better identification. This highlights the growing need to prioritize vascular considerations during surgery, as these anomalies can present greater challenges than the tumor itself, particularly in terms of ensuring surgical safety and complete tumor removal. Some cases also involved renal vein tumor thrombi, necessitating thrombectomy for complete tumor removal. The vascular structure of patients with situs inversus has undergone significant changes.

A complete preoperative examination can help select a more suitable treatment plan for the patient. Preoperative imaging studies provide a comprehensive understanding of the lesion's adjacent structures, particularly the blood supply around the tumor. This information aids in selecting the most appropriate surgical route in advance, reducing the risk of bleeding and trauma, ultimately benefiting patients postoperatively. In retrospect of published articles, both open and minimally invasive nephrectomies were performed, with blood loss ranging from 10 to 200 ml (Table 1). Despite these complexities, successful outcomes were achieved through meticulous preoperative planning and intraoperative vascular management. All patients with renal cell carcinoma and situs inversus require preoperative imaging to assess the anatomical variations of their mirrored structures. However, these variations have minimal impact on the choice of surgical approach and the associated risks and complications, and they are not independent risk factors for determining the surgical method.

CRediT authorship contribution statement

Chong Yan: Conceptualization, Formal analysis, Investigation, Writing – original draft. **Gang Li:** Methodology, Resources, Validation. **Zi-Hao Li:** Formal analysis. **Ze-Hua Wei:** Visualization. **Yu-Xin Peng:** Data curation, Visualization. **Yi-Fang Tao:** Data curation. **Tie Chong:** Project administration, Resources. **Zhen-Long Wang:** Conceptualization, Writing – review & editing.

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