

Robot-Assisted Complex Hilar Tumor Resection: A Case of Ectopic Adrenocortical Adenoma

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Key Clinical Message:

Ectopic adrenal adenomas causing Cushing's syndrome are rare. Precise diagnosis and tailored surgery are vital, especially for tumors near the renal hilum with contralateral kidney dysfunction. Robot-assisted laparoscopy enables successful resection with nephron preservation and excellent outcomes.

KEYWORDS

Ectopic adrenal, Case report, Cushing's syndrome, Robot-assisted laparoscopy

1.INTRODUCTION

Cushing's syndrome is a collection of metabolic disorders and signs resulting from long-term excessive cortisol secretion by the adrenal cortex. These include moon face, centripetal obesity, skin striae, acne, hypertension, osteoporosis, and others. It is also known as hypercortisolism[1], [2]. When the condition is caused by pituitary tumor leading to excessive ACTH secretion, it is referred to as Cushing's disease. If ACTH-like substances secreted by malignant tumors outside the pituitary and adrenal glands stimulate the adrenal cortex to secrete glucocorticoids, it is called ectopic ACTH syndrome[1]. In non-ACTH-dependent Cushing's syndrome, cortisol can be secreted not only by the adrenal glands autonomously but also by ectopic adrenal tissue or ectopic adrenal adenomas with secretory functions[1], [3]. In the case reported in this article, cortisol was secreted by an ectopic adrenal adenoma located at the renal hilum, leading to typical Cushing's syndrome, which is rare in clinical practice.

During embryonic development, the adrenal cortex arises from a group of mesothelial cells between the root of the mesentery of the abdominal wall and the urogenital ridge, while the adrenal medulla originates from the sympathetic nervous system. During the migration of medullary cells to the fetal adrenal cortex region, fragments of the adrenal cortex may be split, and some fragments closely related to the urogenital ridge may

become ectopic with the migration of the developing gonads[4], [5]. Ectopic adrenal tissue typically occurs in the following locations: abdominal axis (32%), broad ligament (23%), epididymis (7.5%), and spermatic cord (3.8%-9.3%)[6]. Case reports have shown that 0.1%-6% of ectopic adrenal tissue occurs in the kidney, primarily located beneath the capsule at the upper pole of the kidney[6]. Most of the adrenal tumors arising from ectopic tissue are non-functional[7], [8]. Interestingly, among reported cases of functional ectopic adrenal adenomas, most tumors are located at the renal hilum[3], [9]. Given the rarity of this condition and the complex anatomical structure of the renal hilum, when typical clinical manifestations of Cushing's syndrome are present, but imaging studies show no corresponding adrenal enlargement or nodular changes, and hormone tests indicate non-ACTH-dependent hypercortisolism, the possibility of ectopic adrenal cortisol-producing adenomas, particularly at the abdominal axis and gonadal region, especially at the renal hilum, should be considered. Additionally, the feasibility of tumor resection in these complex anatomical regions should also be evaluated[7], [8].

This report presents a case of an ectopic adrenocortical adenoma located in the renal hilum that caused Cushing's syndrome. In addition to exhibiting typical signs of hypercortisolism and corroborative hormonal test results, the ectopic adrenal adenoma was situated at the renal hilum, in close association with the renal vein, making resection challenging. Furthermore, the contralateral kidney was nonfunctional, necessitating maximum preservation of nephron units during surgery. We successfully performed complete tumor resection using robot-assisted laparoscopy. A 4-month postoperative follow-up showed satisfactory therapeutic outcomes. By sharing the diagnostic and therapeutic process, along with follow-up results of this case, we aim to provide a reference for the management of rare types of ectopic adrenal glands and to explore the advantages of robot-assisted laparoscopic techniques in complex tumor resections.

2. CASE HISTORY/EXAMINATION

The patient, a 20-year-old female, was admitted to the Department of Urology on June 24, 2024, due to "significant weight gain accompanied by hypertension for 4 years and detection of a left renal mass 15 days prior to hospital admission". Four years ago, the patient experienced significant weight gain without obvious triggers, accompanied by elevated blood pressure. She was diagnosed with "hypertension" at a local hospital and prescribed antihypertensive medication, but blood pressure control was poor. A urinary CT scan conducted 15 days prior to hospital admission revealed a "left renal pelvis mass and right kidney atrophy." She reported no significant flank or abdominal pain, gross hematuria, chills, or fever. The patient sought further diagnostic evaluation and treatment at our hospital. Since the onset of her illness, the patient reported normal appetite and sleep, regular bowel and urinary habits, but occasional menstrual irregularities with prolonged periods. Her weight increased from 60 kg to 80 kg. Medical history: Two years ago, the patient underwent bilateral ureteroscopic laser lithotripsy at a local hospital. On admission on June 24, 2024, physical examination revealed: Temperature: 36.4°C, Pulse: 85 bpm, Respiration: 19 breaths/min, Blood Pressure: 135/110 mmHg, BMI: 31.25. The patient exhibited central obesity, buffalo hump, moon face, abdominal distension, soft abdomen, and no tenderness in the bilateral kidney regions.

3. DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS AND TREATMENT

Auxiliary examination: CT of the urinary system from an external hospital indicated right kidney atrophy and a possible left renal pelvis mass. Further CT plain and contrast-enhanced scans after admission revealed a slightly hypodense tumor at the left renal hilum, approximately 3.8×2.8 cm in size, with irregular shape and heterogeneous enhancement. The left kidney was not compressed. CT urography (CTU) showed normal imaging of the left renal pelvis and ureter with no filling defects (**Figure 1**).



Figure 1 CT enhanced imaging results. **A-B** : A mass at the left renal hilum with slightly lower density, irregular shape, approximately 3.8×2.8 cm in size (At the white arrow). The enhancement scan shows heterogeneous enhancement, with compression of the left renal pelvis. **C** . CTU showing normal visualization of the renal pelvis and ureter, with no filling defect observed.

MRI findings: A T2-weighted scan showed a 3.6×2.6 cm iso-signal tumor in the left renal sinus with visible fat signals. The tumor was traversed by the lower pole artery of the left kidney. Contrast-enhanced imaging revealed homogeneous enhancement of the tumor’s solid components. The tumor was closely associated with the renal vein. Differential diagnoses included ectopic adrenal gland and angiomyolipoma. Renogram: Severe impairment of perfusion, filtration, and excretion functions in the right kidney, with normal function in the left kidney. The total glomerular filtration rate (GFR) was 81.11 ml/min, with 67.96 ml/min from the left kidney and 13.14 ml/min from the right kidney(**Figure 2**).

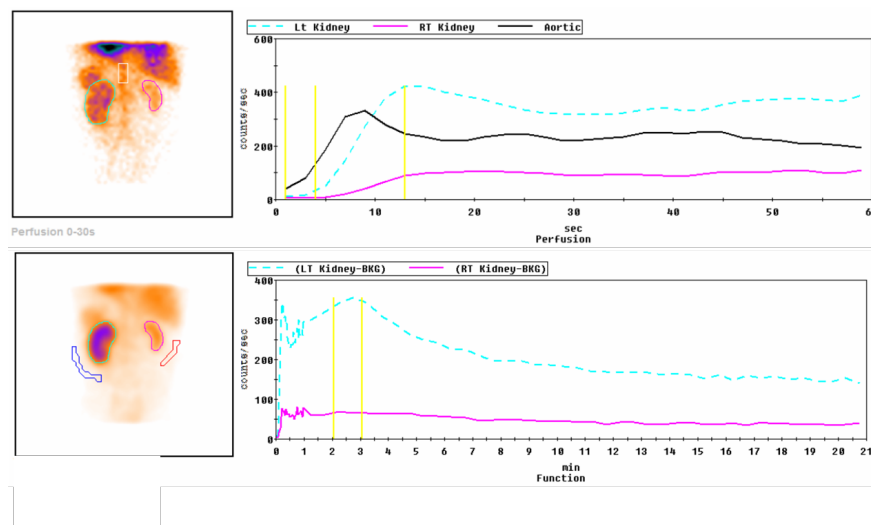


Figure 2 SPECT/CT renal dynamic imaging. **A** . Arterial perfusion imaging: Both kidneys showed early arterial filling 2 seconds after the abdominal aorta became visible. Perfusion of the left kidney was normal, while perfusion of the right kidney was reduced.**B**. Renal dynamic imaging: Both kidneys were visualized, but the right kidney was significantly smaller in size compared to the left kidney, with a normal anatomical position. The left renal cortex demonstrated normal uptake and clearance rates of the tracer, while the right kidney showed reduced uptake and delayed clearance.

PET/CT: A round, mixed-density tumor was observed in the left renal hilum. Both 18-FDG and 18F-PSMA PET/CT scans showed increased radiotracer uptake, with a maximum standardized uptake value (SUVmax) of 6.6. No significant abnormalities were noted in other regions (**Figure 3**). Based on the PET/CT findings, the tumor at the left renal hilum is considered likely to be an ectopic adrenal adenoma.

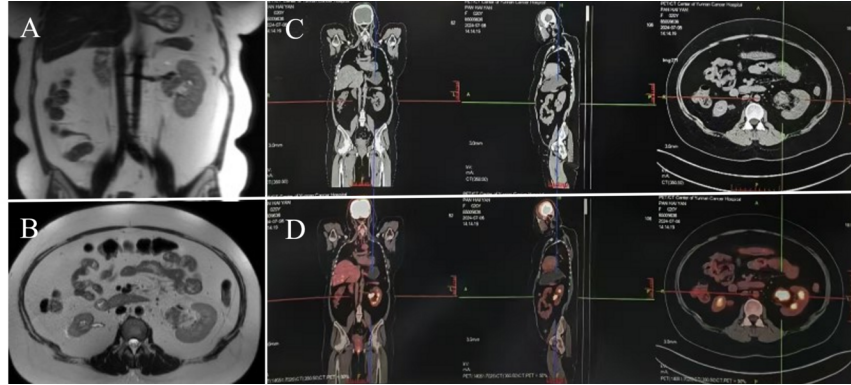


Figure 3 MRI and PET/CT findings suggestive of ectopic adrenal gland. **A**: MRI T2-weighted images show a 3.6×2.6 cm Tumor within the left renal sinus with fat signal. **B**: The enhancement scan reveals uniform enhancement in the parenchymal portion. The Tumour contains the left renal lower pole artery traversing it, and the Tumor is closely related to the renal vein. **C**: PET/CT: A round, isodense, mixed-density Tumor is observed in the left renal hilum. **D**: Both 18-FDG and 18F-PSMA PET/CT imaging show increased radioactive uptake, with a maximum standardized uptake value (SUVmax) of 6.6(SUVmax < 2.5: Typically indicative of benign lesions or normal tissue. SUVmax > 2.5: Suggests the possibility of malignancy, requiring further evaluation in conjunction with the clinical context. SUVmax > 10: Commonly observed in highly metabolically active malignant tumors, such as lymphoma or lung cancer.),. No significant abnormal findings were noted in other areas.

Laboratory results: Increased cortisol secretion and disrupted diurnal rhythm were observed, with elevated 24-hour urinary free cortisol(24h-UFC) (Table 1). ACTH level was 4.43 pg/ml, and levels of angiotensin, renin, aldosterone, and aldosterone-to-renin ratio (ARR) were within the normal range.. Overnight 1 mg low-dose dexamethasone suppression test and 5 mg high-dose dexamethasone suppression test showed no suppression of cortisol levels. Liver and renal function tests, electrolytes, and blood glucose levels were all normal.

Testing time	($\mu\text{g}/\text{dL}$)	($\mu\text{g}/\text{dL}$)	($\mu\text{g}/\text{dL}$)	($\mu\text{g}/24\text{h}$)
Preoperative	18.90	19.40	19.90	840.00
Postoperative	1.81	6.79	4.44	329.19

Table 1. Preoperative elevated cortisol levels with loss of diurnal rhythm, postoperative restoration of cortisol secretion rhythm.

Based on the patient’s clinical manifestations, hormonal test results, and imaging findings, the diagnosis included: ectopic adrenocortical adenoma, ACTH-independent Cushing’s syndrome, hypertension, and non-functioning right kidney. After confirming the diagnosis, the patient underwent robot-assisted laparoscopic resection of the ectopic adrenocortical adenoma at the left renal hilum under general anesthesia on July 11, 2024.

4. OUTCOME AND FOLLOW-UP

Intraoperatively, the tumor was located in the renal sinus, closely associated with the kidney, lacking an intact capsule, and adhered tightly to the renal hilar vessels. Some venous tributaries of the tumor drained into the left renal vein. The left renal artery was clamped intraoperatively to facilitate tumor resection. The tumor was completely removed, with a warm ischemia time of 20 minutes (Fig. 4 A, B). Postoperative pathology confirmed the diagnosis of an adrenocortical adenoma (Fig. 4 C). Immunohistochemistry results: CA9(-), CD10(focal+), CD117(-), CK(-), CK7(weak, scattered+), GATA3(-), Heppar-1(weak, scattered+),

HMB45(-), Ki-67(+5%), Melan A(+), Pax-8(-), S100(-), SDHB(+), TFE-3(partial+), Vimentin(partial+), a-inhibin(+), Calponin(-), CD34(vessels+), Desmin(-), MPO(partial+), SMA(-), EMA(-).

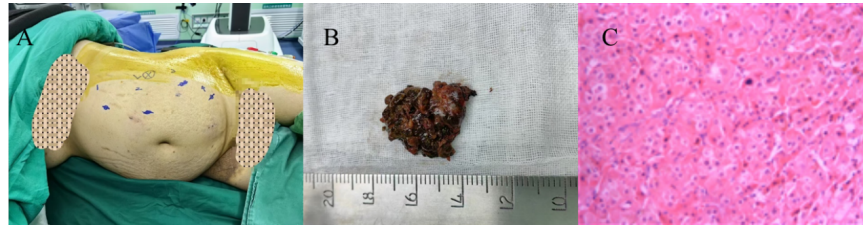


Figure 4 Intraoperative and postoperative findings. **A.** Robotic-assisted laparoscopic positioning and trocar placement via transabdominal approach. **B.** The grossly excised tissue post-surgery appears yellowish and solid, with nodular hyperplasia observed. **C.** Post-surgical hematoxylin and eosin (H&E) stained pathological section is consistent with adrenal adenoma findings ($\times 100$).

Postoperative hormone supplementation protocol: Hydrocortisone 100 mg was administered intravenously during the surgery. On the day after the surgery, hydrocortisone 200 mg was administered intravenously. On the first postoperative day, hydrocortisone 200 mg was given intravenously (125 mg at 8:00 AM and 75 mg at 4:00 PM), and the following day, the dose was reduced to 150 mg (100 mg at 8:00 AM and 50 mg at 4:00 PM). After resuming normal oral intake, prednisone 30 mg per day was prescribed orally, with a gradual reduction to 20 mg per day upon discharge, depending on the patient's condition. The dose was further reduced by 2.5 mg every 4 weeks.

Regular postoperative follow-up was conducted for 2 months, with no symptoms of adrenal insufficiency, such as anorexia, nausea, tachycardia, apathy, or fatigue and drowsiness. The patient's cortisol diurnal rhythm was restored (Table 1), menstruation was regular, and the patient's weight decreased by 5 kg. No significant symptoms of adrenal insufficiency or adrenal crisis were observed during the follow-up period. Plasma cortisol and ACTH levels will continue to be monitored, and medications may be gradually reduced or discontinued once adrenal cortical function is confirmed to have returned to normal.

5. DISCUSSION

Robotic-assisted laparoscopic technology offers significant advantages over traditional open surgery and conventional laparoscopic surgery in the resection of complex tumors[10], [11], such as those located at the renal hilum. The robotic system provides high-definition, 3D stereoscopic vision, which can magnify the surgical field more than 10 times, allowing for clearer visualization of critical anatomical structures, such as the complex vasculature and surrounding tissues of the renal hilum. Furthermore, robotic surgery reduces the physical strain on surgeons, enabling long, high-precision procedures and improving surgical efficiency[11]. Additionally, in partial nephrectomy for renal tumors, robotics can optimize the balance between tumor resection and renal function preservation, enhancing postoperative renal function retention[12]. In the case reported here, the patient had right renal atrophy and nonfunctional right kidney, necessitating maximal preservation of the left renal unit. Although the tumor's close adhesion to the renal hilum vessels increased the surgical difficulty, the superior ergonomics, 3D magnification of the surgical field, tremor filtration, and enhanced instrument dexterity of the robotic-assisted surgical system provided a significant advantage in handling complex tumors surrounded by large blood vessels and tightly adhered to the renal hilum. The tumor was successfully resected, with the kidney's warm ischemia time limited to only 20 minutes.

Adrenal function has long been suppressed by the high levels of cortisol secreted by ectopic tumors[13]. Although the source of high cortisol secretion is removed after surgery, adrenal function has not yet fully recovered, and the patient remains in a relative state of cortisol deficiency[13]. Therefore, glucocorticoid supplementation is necessary after the removal of any cortisol-secreting tumor to prevent symptoms of adrenal insufficiency or adrenal crisis[14]. Currently, there is no standardized replacement therapy, and there

are differences in medication practices and experiences among medical centers. Generally, hydrocortisone is administered intravenously during surgery and on the day after surgery. Once oral intake is tolerated, prednisone is switched to oral administration[14], [15]. Glucocorticoids need to be gradually tapered because cortisol secretion follows a diurnal and pulsatile rhythm in a physiological state. Thus, glucocorticoid supplementation is given in multiple doses, with higher doses in the morning and lower doses in the afternoon, and the dose may be increased during times of stress[14].

Ectopic adrenal glands are rare in adults and are commonly located in the retroperitoneal axis, broad ligament, or spermatic cord. Similar to normal adrenal glands, they can develop cortical hyperplasia, nodules, or adenomas. It is rare for ectopic adrenal adenomas to cause Cushing's syndrome. When the tumor is located near the renal hilum, the complex anatomy increases the difficulty of complete resection. The use of a robot-assisted surgical system allows for safer and more precise tumor removal.

PATIENT CONSENT STATEMENT

Written informed consent was obtained from the patient for the publication of this case report, including any accompanying clinical details and images. The original consent form is securely retained and can be provided to the Publisher upon request in accordance with journal policies.

AUTHOR CONTRIBUTIONS

Yunqiang Shi: Conceptualization, Data curation, Formal analysis, Project administration, Writing - original draft. **Kai Deng:** Conceptualization, Data curation, Formal analysis, Project administration, Writing - original draft. **Xiaofang Bi:** Data curation, Investigation, Writing - review & editing. **Jie Xiong:** Data curation, Investigation, Supervision. **Meng Yang:** Data curation, Investigation. **Lin Zhang:** Data curation, Investigation. **Chunhui Wang :** Conceptualization, Data curation, Funding acquisition, Project administration, Supervision, Writing - review & editing

CONFLICT OF INTEREST STATEMENTThe authors declare that they have no conflicts of interest regarding the publication of this article.

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