



## Case Report

# Left Adrenal Venous Drainage into the Inferior Vena Cava in a Pheochromocytoma Patient with Ectopic Pelvic Kidney

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### Abstract

In this report, we describe a very rare variant adrenal venous anatomy in a left-sided pheochromocytoma case with left adrenal vein draining into the inferior vena cava (IVC). A 66-year-old female with an incidentally discovered left adrenal mass was referred to our clinic for further diagnostic work up. She had hypertension for the past three years. Abdominal magnetic resonance imaging which has been performed for essential thrombocytopenia revealed a left adrenal mass. She was diagnosed as pheochromocytoma by further laboratory workup. Pre-operative imaging with computed tomography (CT) angiography showed that left kidney was ectopically localized in pelvis, and left adrenal vein was draining directly into the IVC. A laparoscopic transabdominal left adrenalectomy was performed. Final pathology result was consistent for pheochromocytoma. Variant adrenal venous anatomy is rare. There are only few case reports on patients with left adrenal vein draining into the IVC. Although there are handful of autopsy studies, these studies were performed on normal adrenal glands. The number of clinical reports on variant adrenal venous anatomy is limited. Clinical studies revealed that most commonly seen variant adrenal venous anatomy was number based and were associated with larger tumor size and pheochromocytoma. Pre-operative CT imaging could be used to improve the identification of venous anatomy. In this report, we present a very rare case of an adult patient with left ectopic kidney who had undergone laparoscopic adrenalectomy for left adrenal pheochromocytoma and had left adrenal vein draining into the IVC. To the best of our knowledge, it is a first in the literature. Pre-operative CT imaging identified the variant adrenal venous anatomy in this patient.

**Keywords:** Adrenal tumors, Adrenal vein anomalies, Left adrenal vein, Pheochromocytoma

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Minimally invasive adrenalectomy has become the mainstay treatment for adrenal diseases.<sup>[1]</sup> Apprehensive knowledge of the anatomy prior the surgery should be obtained on case-by-case basis to construct a proper dissection plan. The textbook venous anatomy for each adrenal gland is described as a single vein draining directly into vena cava on the right side, and a left adrenal vein joining with inferior phrenic vein and then draining into

left renal vein on the left side. However, variations of these patterns have been reported in case reports, clinical, and cadaveric studies.<sup>[2]</sup> In majority of patients, kidneys and adrenal glands are anatomical neighbors. In the case of renal agenesis or ectopy, most patients' adrenal glands lie in its original anatomical location.<sup>[3]</sup> In this case report, we present an anomalous left adrenal venous drainage into the inferior vena cava (IVC) in an adult patient with a left ectopic

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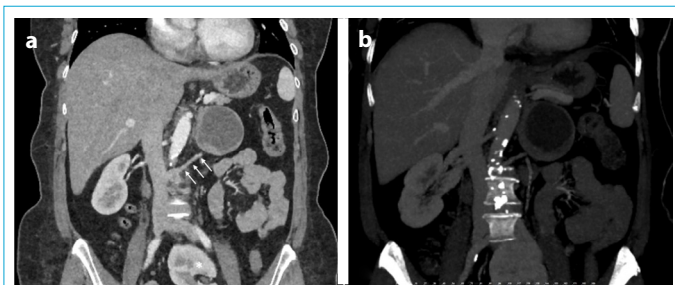


pelvic kidney and a left adrenal tumor which previously has been described once in the literature but, to the best of our knowledge, this is the first case describing a patient with pheochromocytoma.<sup>[4]</sup>

## Case Report

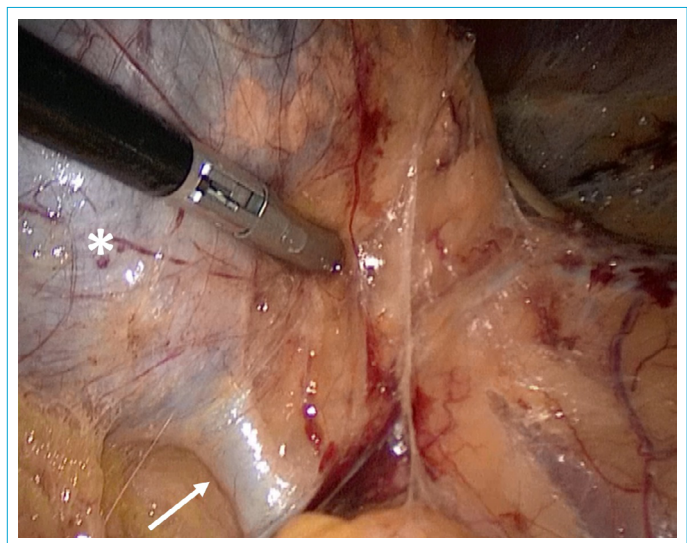
A 66-year-old woman with a 3-year history of essential thrombocytosis (ET) and hypertension has been referred to our clinic after the incidental detection of a 5 cm left adrenal mass on abdominal magnetic resonance imaging (MRI) performed for ET surveillance. Laboratory results revealed that serum sodium and potassium levels were within normal range. The results of further laboratory assays were as the following; serum aldosterone level: 134.6 ng/dL (Range: 70–300), plasma renin activity: 1.33 ng/mL/h (range: 0.1–6.56), and spot aldosterone-to-renin ratio: 10.1. Suppression test with 1 mg dexamethasone resulted in 08.00 and 08.30 AM cortisol levels of 1.05 and 1.11  $\mu\text{g}$ , respectively (range: 4.8–19.5). A 24 h urinary analysis revealed metanephrine level of 4859  $\mu\text{g}/24\text{ h}$  (range: 30–350), ~ 14 times the upper limit of normal (ULN), normetanephrine level of 1622  $\mu\text{g}/24\text{ h}$  (range: 50–650), ~ 3 times the ULN, and vanillylmandelic acid level of 13.7 mg/24 h (range: 1.6–7.3), ~ 2 times the ULN. The basal creatinine level was 0.84 mg/dL with estimated glomerular filtration rate of 73 mL/min/1.73m<sup>2</sup>.

T2-weighted MRI images on the left adrenal gland revealed a 51 × 52 mm hyperintense nodule with rim contrast enhancement in its original anatomic position. The patient was medically treated with phenoxybenzamine, diltiazem, doxazosin preoperatively for 3 weeks to ensure the adequate alpha and beta blockage has been sustained. Abdominal triphasic computed tomography angiography (CTA) showed that left kidney was ectopically localized in pelvis, and left adrenal vein was draining directly into the IVC posterior to the aorta (Fig. 1).



**Figure 1.** (a) On computed tomography angiography (CTA), arrows show left adrenal vein that stems from a left adrenal tumor, draining into the inferior vena cava (IVC). Ectopic pelvic kidney is marked with an asterisk. (b) Reconstructed CTA image of left adrenal venous drainage into the IVC.

She was taken into the operating room for a laparoscopic transabdominal left adrenalectomy. The patient was placed in right-lateral decubitus position. Abdominal incision was made in left anterior axillary line left of umbilicus and 10 mm port was placed with open (Hasson) approach. Subsequently, two more 10 mm ports were placed in subcostal left anterior and mid axillary lines, respectively. Left paracolic gutter dissection was performed with monopolar hook energy device and a vessel sealer. Splenic flexure was taken down and left kidney was not visualized. A 5 cm left adrenal was seen and surrounding fatty tissue was dissected off the mass without interrupting its capsule. The left adrenal vein draining into the IVC was visualized and clamped with hemostatic clips proximally and distally (Fig. 2). The vein was then cut with scissors, the dissection was further continued, and the specimen was extracted using endoscopic retrieval bag. Following hemostasis, a drain was left in place and operation was finalized without any intraoperative complications. The patient was normotensive during the procedure and followed up in intensive care unit without any postoperative complications. Patient was started on regular diet 6 h after the operation. The drain was removed on postoperative day 1. The patient was discharged on postoperative day 3 and remained normotensive during her postoperative course. Final histopathology of the adrenal specimen was consistent with pheochromocytoma. Patient has signed informed consent regarding treatment and agreed to participate in the study.



**Figure 2.** In this laparoscopic image, left adrenal tumor (marked with an asterisk and left adrenal vein is marked with an arrow) is seen in its original location but left kidney is ectopic and quadratus lumborum can be visualized instead.

## Discussion

The identification of adrenal vein is very important for diagnosis and treatment of adrenal disease. Bilateral venous sampling can be used to differentiate a unilateral adenoma from bilateral hyperplasia. Minimally invasive adrenalectomy requires identification and ligation of adrenal veins.<sup>[1]</sup> Therefore, variations of adrenal venous anomaly can present a challenge during diagnosis and surgery. Right adrenal vein most commonly drains into posterolateral the IVC while left adrenal vein drains into left renal vein.

The data on anatomical variations of adrenal veins are scarce. Historically, studies performed on cadavers reported higher rate of variations on the right side: Right adrenal vein draining into inferior hepatic vein, right renal vein, or combination of all three.<sup>[5]</sup> On the left side, there are a few variants and are usually accompanied by anatomical anomalies of kidneys and renal veins.<sup>[6]</sup> If there is a duplicate of left renal veins, left adrenal vein drains into anterior renal vein as described by Field and Saxton.<sup>[7]</sup> If the renal vein is retro-aortic, left adrenal vein can be seen draining directly into the IVC as described by Stack et al.,<sup>[8]</sup> It can also drain into left lumbar veins or azygos-hemiazygos system.<sup>[7]</sup> Majority of patients with renal agenesis and ectopia, will have adrenal glands in its original anatomical position.<sup>[3]</sup> Therefore, the left adrenal vein would retain its connection to the IVC and drain directly into it. Kenney et al.<sup>[4]</sup>, reports an anomalous left adrenal vein draining directly into the IVC in a 47-year-old male with bilateral renal ectopia and left adrenal adenoma. Similarly, El-Sherief. describes a left adrenal vein draining directly into the IVC in a 1-year-old child with multiple congenital malformations and aplastic left kidney.<sup>[9]</sup> In our case, the patient had a congenital pelvic ectopia of the left kidney with left adrenal gland in its original anatomic location. The abdominal CTA revealed that the patient's adrenal tumor was draining directly to the IVC behind the aorta.

In a study conducted by Scholten et al.<sup>[2]</sup>, 546 consecutive laparoscopic adrenalectomies for anatomical variants of adrenal veins were analyzed. They reported 28 anomalies on the left side (9%) of which only 4 are related to location-based variations. Patients with variant anatomy had more pheochromocytomas. Mean operative time and estimated blood loss was higher for the variant anatomy group.

In a more recent study by Sun et al.<sup>[10]</sup>, 303 consecutive minimally invasive adrenalectomies were studied. Variant anatomy was detected in 20.5% and they were associated with increased tumor size, larger adrenal veins and pheochromocytomas. Similar to the previous study number base variations were more common than location based ones. Most common location based left adre-

nal anomaly was accessory adrenal vein draining into left inferior phrenic vein. No patients with left ectopic kidneys or drainage of the left adrenal vein into the IVC were seen. Variant anatomy was also associated with increased blood loss, more need for transfusion, longer operative time and post-operative stay, overall associated with worse surgical outcomes.

The minimally invasive surgery for adrenal tumors provides better visualization of relevant anatomy although controlling bleeding may provide difficult with massive intraoperative hemorrhage.<sup>[1,11]</sup> Traction injury of adrenal veins can occur during dissection therefore preoperative assessment of vascular anatomy with imaging studies is crucial to construct a proper dissection plan which can predict and prevent major complications.<sup>[1]</sup>

## Conclusion

We presented a case with an anomalous left adrenal vein drainage directly into the IVC. Although left adrenal venous anomalies are rare, they can be seen in patients with renal anomalies. Pre-operative assessment of vascular anatomy should be considered in all patients with adrenal tumors.

## Disclosures

**Informed consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

**Peer-review:** Externally peer-reviewed.

**Conflict of Interest:** None declared.

**Authorship Contributions:** Concept – Y.I., Y.G.S., F.T.; Design – B.S.; Supervision – Y.I., I.C.S.; Materials – F.T., Y.G.S., I.C.S.; Data collection &/or processing – B.S., N.A., A.P.; Analysis and/or interpretation – B.S., Y.I., A.P.; Literature search – B.S., Y.I., N.A., Y.G.S.; Writing – B.S., Y.I., Y.G.S.; Critical review – Y.G.S., F.T., N.A., I.C.S.

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