

# Paraganglioma Displacing IVC: A Surgical Challenge

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## KEY WORDS

paraganglioma ► pheochromocytoma ► liver transplantation technique

## ABSTRACT

Paragangliomas are catecholamine-secreting tumors that originate from chromaffin cells in the sympathetic ganglia. Commonly presenting with excess catecholamine symptoms, these tumors are usually treated with surgical excision. We present a case of an abdominal paraganglioma located between the aorta and the inferior vena cava and discuss the challenges associated with the resection of this tumor in such a complex location.

## INTRODUCTION

Paragangliomas are neoplasms that arise from chromaffin cells in the sympathetic ganglia, which travel in the paravertebral region [1-3]. These rare tumors occur anywhere from the upper portion of the neck to the pelvis [2, 4]. The incidence of these tumors has been reported to be as low as 0.05% [5]. Within the abdomen, paragangliomas are commonly found in the superior para-aortic region featuring characteristic microscopic trabeculation and nesting patterns [6]. Patients who develop paragangliomas below the neck may have back pain, classic catecholamine excess symptoms, or may be completely asymptomatic [2, 4, 7]. We present a rare case of an intra-abdominal paraganglioma located between the aorta and the inferior vena cava and discuss the surgical challenges associated with resection of these types of tumors.

## CASE PRESENTATION AND MANAGEMENT

A 64-year-old non-hypertensive male presented with persistent back pain worsened by ambulation. Computed tomography (CT) of the lumbar spine detected an incidental right upper quadrant lesion. Dedicated abdominal CT with three dimensional reconstructions was obtained, demonstrating a highly vascularized 6.5 cm x 5.5 cm mass in the right upper quadrant. The tumor was located in the aorto-caval groove, displacing the inferior vena cava (IVC) to the right (Fig. 1). It was closely related to the right renal artery, the left renal vein, and the celiac and superior mesenteric arteries (Fig. 2). There was no evidence of invasion (Fig. 1) and the right adrenal gland was distinct from the mass.

Biochemical studies (serum epinephrine & norepinephrine and urinary metanephrines and normetanephrines) ruled out a functional lesion and the patient elected to undergo open surgery. Routine preoperative testing was performed to clear the patient for surgery. A bilateral subcostal incision was used. Estimated blood loss was 500 ml. The patient's postoperative recovery was uneventful. The specimen measured 9.0 cm x 6.0 cm x 4.0 cm. Histopathological examination revealed a pheochromocytoma with

positive immunohistochemical staining for synaptophysin and chromogranin antibodies. The patient is free of recurrence at six months follow-up.

## DISCUSSION

Extra-adrenal pheochromocytomas are rare cases of catecholamine-secreting tumors that originate from chromaffin cells in the sympathetic ganglia [1, 3]. These tumors comprise approximately 10% to 15% of all cases of pheochromocytoma [8]. A large series from the Mayo clinic reported the head and neck region to be the most common site for benign paragangliomas. Within the abdomen, the majority of these lesions are located in the superior periaortic and paracaval region [1, 2, 4].

Patients with paragangliomas may present with symptoms associated with catecholamine excess including palpitations, headache, perspiration, pallor, or orthostasis [4]. Sustained or episodic hypertension is observed in over 50% of patients [2, 7] and usually resolves following surgical resection [4]. Abdominal or flank pain may be the only presenting symptom secondary to mass effect in larger non-functional tumors [4].

An open approach utilizing a bilateral subcostal incision was selected owing to the size of the lesion and its proximity to multiple major vascular structures. A Rochard retractor was used to provide adequate exposure of the upper abdomen. At our institution we commonly undertake the resection of large renal masses with or without caval extension using liver transplantation techniques [9]. The right triangular and coronary ligaments are divided and the liver is mobilized to the left side of the abdomen to expose the bare area and the hepatic IVC. Short hepatic veins are ligated if necessary. The liver can then be rotated on the IVC and retracted into the left upper quadrant to expose the upper pole of the right kidney and the retroperitoneum. The IVC can also be mobilized paying particular attention to lumbar veins.

In this instance, mobilization provided exposure to the lateral border of the IVC and allowed further dissection to mobilize the



**Fig. 1.** Transverse CT section showing the inter-aorto-caval location of the tumor.

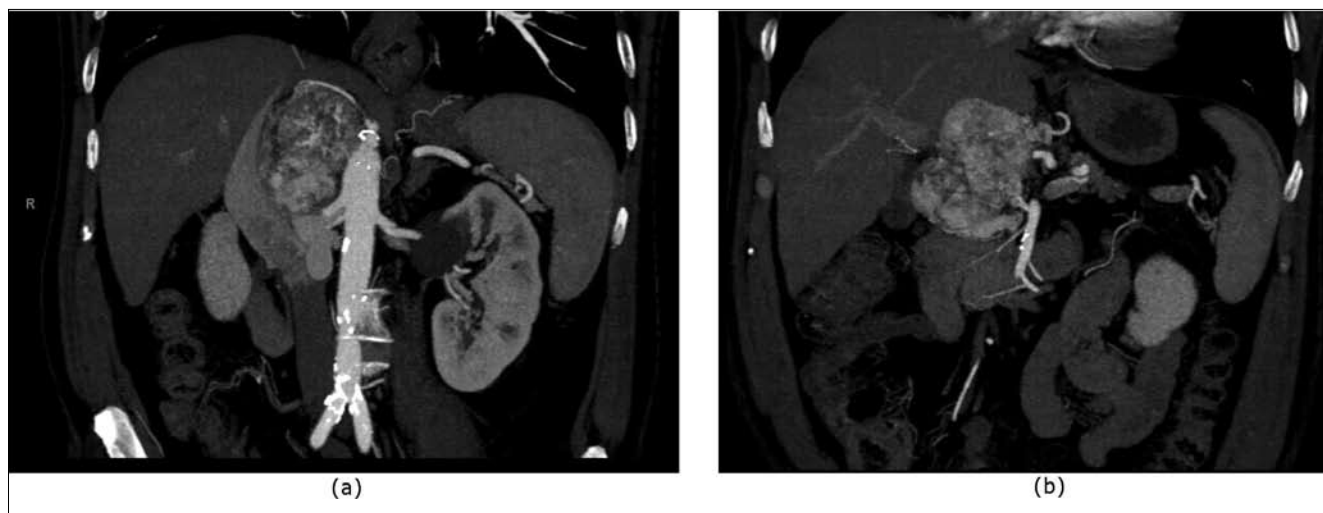


Fig. 2. Coronal CT sections showing (a) tumor locations between the IVC and aorta (b) and its relation to the superior mesenteric artery.

posterior and lateral aspects of the tumor. The right adrenal gland was identified and preserved when the tumor was separated from the upper pole of the right kidney. The duodenum was Kocherized and the lesser sac was also opened by incising the peritoneum forming the anterior wall of the lesser sac to allow access to the aorta and the upper and medial aspects of the tumor. Multiple arterial branches from the superior mesenteric and multiple venous tributaries draining into the left renal vein were ligated, thereby providing exposure to the inferior aspect of the tumor. The mass was finally released by careful dissection from the IVC, left renal vein, and aorta.

Compared to renal and adrenal lesions, paragangliomas require special attention owing to a fragile and ill-defined capsule, which can make dissection challenging. In addition, excessive manipulation should be avoided to prevent intraoperative hypertension, which may occur despite alpha and beta blockade.

A thorough understanding of the anatomy, careful dissection, and meticulous hemostasis are essential for a safe outcome. At our institution, these challenging cases are undertaken by an interdisciplinary team comprised of urologists and transplant surgeons. These techniques have proven useful for treating large renal and adrenal tumors as well as retroperitoneal masses [9]. Our findings add a new case to the literature on paragangliomas in the aorto-caval region.

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