

Pre-operative arterial embolization of a large functional abdominal paraganglioma

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Introduction

Paragangliomas are rare, highly vascular tumors that arise from autonomic nerve ganglia. Paragangliomas that originate from the parasympathetic ganglia in the neck are also known as "carotid body tumors," "chemodectomas," "glomus jugulare tumors," and "non-chromaffin paragangliomas." These carotid body tumors are usually non-functional; however, their highly vascular nature poses a significant risk of severe hemorrhage during resection. Several techniques, such as intra-operative artery ligation, selective embolization, and balloon occlusion have been used with variable success to control operative field bleeding of carotid body tumors [1, 2].

Paragangliomas derived from sympathetic ganglia arise in the mediastinum, heart, retro-peritoneum, and pelvis. These sympathetic paragangliomas are often described as "extra-adrenal pheochromocytomas." Like the carotid body tumors, sympathetic paragangliomas are highly vascular; however, these tumors may also produce catecholamines that lead to significant hypertension both before and during surgical treatment.

Sympathetic paragangliomas are rarely embolized because of the technical challenges and fear of precipitating hypertensive crisis. A small number of cases using embolization of adrenal pheochromocytomas have been reported [3-5]. These procedures are typically done several days before surgical resection to allow medical stabilization of the patient's catecholamine-induced hypertension.

Case Presentation

The patient is a 44-year-old man with a long-standing history of hypertension. Physical exam at the time of diagnosis was notable for a supine blood pressure of 172/112 with a pulse of 60, and standing blood pressure of 140/106 with a pulse of 60. The patient was taking several medications for his blood pressure: labetalol, prazosin, benazepril, and hydrochlorothiazide.

After multiple episodes of recurrent epigastric pain, the patient had a thin-section abdominal CT scan showing a 7.1 x 6.0 x 7.2 cm mass inferior and posterior to the head of the pancreas, likely retroperitoneal, with possible invasion of the adjacent vena cava and duodenum (Figure 1A).

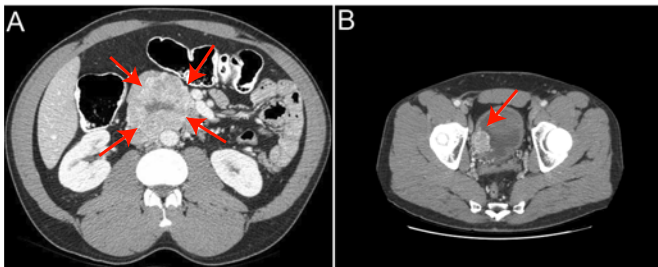


Figure 1: Contrast-enhanced axial computed tomographic (CT) images. (A) a 7-cm retroperitoneal mass (arrows) ventral to the aorta, and (B) a 2-cm mass (arrow) in the right bladder wall.

A second mass measuring 3.1 x 2.9 cm was present in the right bladder wall (Figure 1B), consistent with a second paraganglioma. A 1-cm left adrenal adenoma was also identified. Whole-body 123I-metaiodobenzyl-guanidine (¹²³I-MIBG) scanning showed avid uptake in the retroperitoneal mass, mild uptake in the bladder wall mass, and no unusual uptake in the adrenal glands.

Diagnosis

A 24-hour urine collection for fractionated metanephrines and catecholamines confirmed the diagnosis of secretory paragangliomas. The fractionated values were as follows (with normal values):

Metanephrines	694 nmol/d (NI <1136 nmol/d)
Normetanephrines	>24,478 nmol/d (Normal <2,738 nmol/d)
Total metanephrines	>49,209 nmol/d (NI <3,498 nmol/d)
Epinephrine	38.22 nmol/d (NI <591 nmol/d)
Norepinephrine	9113 nmol/d (NI <131 nmol/d)
Dopamine	4087 nmol/d (NI <3139 nmol/d)

The patient's plasma fractionated metanephrine, catecholamine, and serum chromogranin A levels are presented in Table 1.

	Meta nephrines	Normeta nephrines	Total Metanephrines	Epinephrine	Norepi nephrine	Dopamine	Total Catecholamine	Chromogranin A (CgA)
Normal pper limit (SI Units)	0.42 nmol/L	0.98 nmol/L	1.34 nmol/L*	0.27 nmol/L	3.88 nmol/L	0.065 nmol/L	3.95 nmol/L*	39 ug/ml
At diagnosis (day+40)	0.84	23.94	24.26	0.44	---	<0.13	---	310.00
Admission (day-1)	0.61	19.25	19.45	0.27	>47.28	<0.13	>103.66	330.00
After embolization (day 0, 11:40 am)	0.69	16.12	16.47	0.30	>47.28	<0.13	>47.44	---
After tumor resection (day 0, 1:20 pm)	0.26	10.36	10.39	0.37	>47.28	<0.13	85.71	---
Postop. Day 1	---	---	---	0.61	20.50	<0.13	21.08	---
Postop. Day 9	0.23	1.09	1.31	0.62	9.88	<0.13	10.52	16.50
1 year followup	0.24	0.64	0.89	0.28	4.02	<0.13	4.31	---

Embolization

The patient was scheduled for sequential embolization and surgical resection of the abdominal and bladder masses on the same day. The patient's blood pressure was monitored with an arterial transducer. Using iohexol non-ionic contrast (iohexol, Amersham Health) to reduce the risk of hypertensive crisis, trans-femoral aortography revealed that the abdominal paraganglioma was vascularized via the right lumbar L2 artery (Figure 2A). A 5-French selective visceral C-2 (Cook) catheter was used to select the right L2 lumbar vessel and a 3-French micro-catheter (Boston Scientific) was used to super-selectively catheterize branches of the lumbar artery (Figure 2B). The tumor was embolized with 3x2 mm platinum tornado micro-coils (Cook) to prevent inadvertent embolization of arteries distal to those supplying the tumor. Gelfoam slurry was then injected. After the embolization, repeat abdominal aortography (Figure 2C) confirmed a reduction of the visualized tumor vasculature. There was no rise in blood pressure during or after the embolization procedure, and plasma normetanephrines did not rise.

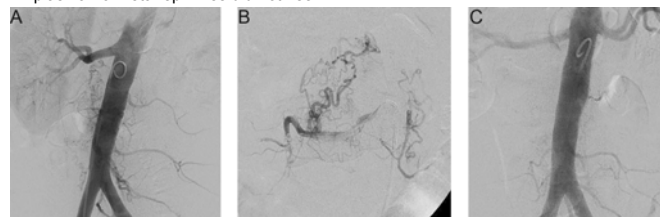


Figure 2: (A) Abdominal aortogram confirmed the presence of a hyper-vascular mass supplied predominantly by the lumbar arteries. (B) Selective contrast injection into the right L-2 lumbar artery showed neovascularity within the mass. (C) Repeat aortogram confirmed a successful reduction of visualized tumor vessels after embolization of the tumor with Gelfoam slurry.

Surgery

After the abdominal paraganglioma embolization, the patient was transferred directly to the operating room where he underwent a laparotomy to resect the retroperitoneal paraganglioma. A Kocher maneuver revealed a large mass posterior to the second and third portions of the duodenum. Manipulation of the retroperitoneal mass was associated with elevated systolic blood pressures and bronchospasm that resolved when the mass was left undisturbed for several minutes. Multiple vascular branches from the duodenum and aorta were identified and ligated. No invasion into the aorta or the inferior vena cava was noted; however, there were multiple short, delicate vessels bridging the neoplasm to the aorta. A Satinsky clamp was placed on the aorta before the mass was separated from the aorta, and the clamp was removed after multiple friable tumor vessels arising from the aorta were controlled. Estimated blood loss for this portion of the surgery was 350 cc. The left adrenal adenoma was not removed because of the benign appearance on abdominal imaging.

A partial cystectomy with en-bloc resection of the bladder tumor was then performed through the same incision. The bladder was closed in layers and an indwelling Foley catheter was left in place. The catheter was removed 2 weeks later after a cystogram showed no leakage of urine.

After his surgical procedures, the patient remained intubated for 2 days in the intensive care unit. His blood pressure was easily managed with moderate doses of carvedilol and lisinopril. He was subsequently discharged on post-operative day 5. Pathologic analysis of the tumor masses showed multiple nests of peripherally dilated and delicate vasculature in the Zellballen pattern, with polygonal cells having amphophilic cytoplasm. Immunohistochemical staining of both tumors was positive for chromogranin A and synaptophysin, confirming the diagnosis of multiple paragangliomas.

Discussion

Although the advent of α -receptor and catecholamine release blockers has improved blood pressure management, surgical resection of a large functional paraganglioma remains a complicated yet necessary component of treatment. Paragangliomas express high levels of vascular endothelial growth factor (VEGF) [6] making them extremely vascular and increasing the risk of severe hemorrhage during resection [1]. Paragangliomas also exhibit more variable vascular anatomy than pheochromocytomas. These features make pre-operative embolization an attractive adjunct before surgical resection.

Given the large size of this patient's retroperitoneal paraganglioma, we believe that the moderate surgical blood loss and relatively stable hemodynamics during surgery were significant benefits of the pre-operative tumor embolization. Removing the retroperitoneal tumor in our patient provided hemodynamic stabilization before the bladder wall paraganglioma was removed in the second stage of the surgery.

Summary

The patient described here exemplifies the feasibility and potential benefits of rapid, sequential pre-operative embolization and surgical resection of a large, functional abdominal paraganglioma. This combination of immediate pre-operative embolization and surgical resection may help decrease operative complications, including hypertensive crisis, improve the surgical outcome in patients with large secretory vascular tumors, may provide palliation for patients with unresectable tumors.

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