

Diagnostic and Perioperative Complexity when Retroperitoneal Hemangioma Mimics Sympathetic Paraganglioma: A Case Report

Ankith Laichetty

ankith.laichetty@wfusm.edu

Wake Forest University School of Medicine

Perry Shen

Wake Forest University School of Medicine

Jingmei Lin

Wake Forest University School of Medicine

Case Report

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Abstract

Background.

Primary retroperitoneal tumors, which comprise 0.1–0.2% of all malignancies, can encompass various etiologies, and possess nonspecific imaging features. These factors make them a diagnosis of exclusion, especially in adult patients. Additionally, fewer than 50 cases of adult primary cavernous hemangiomas in the retroperitoneal space have been reported. However, when a hypervascular retroperitoneal mass is accompanied by hypertension and ambivalent serum catecholamine levels, a functional sympathetic paraganglioma becomes the leading diagnosis as it requires a differential work up, driving a high-stakes perioperative pathway.

Case Presentation.

A 52-year-old man with insulin dependent type 2 diabetes mellitus, hypertension, hyperlipidemia and schizoaffective disorder had a well vascularized retroperitoneal mass incidentally identified on trauma imaging following a motor vehicular accident (MVA). Cross-sectional imaging and mildly-to-moderately elevated urinary and plasma catecholamines/Metanephrines raised concern for a functional paraganglioma. He underwent preoperative alpha then beta-adrenergic blockade and laparoscopic resection. Final Histopathology demonstrated a cavernous hemangioma with no neuroendocrine tissue.

Conclusions.

The combination of chronic hypertension, anxiety, and equivocal biochemistry produced a convincing phenotype for functional paraganglioma. Since unprepared resection of a functional paraganglioma carries substantial perioperative cardiovascular risk, alpha-blockade optimization was justified even though the lesion proved non-functional. Poorly controlled diabetes with marked perioperative glycemic variability added independent surgical risk. Since hypervascular retroperitoneal masses are rare in adults; if the diagnosis of a functional paraganglioma cannot be excluded, the safest course is to prepare as though the mass were one.

Background

Cavernous hemangiomas are benign vascular tumors typically found in the skin or liver; their presence in the retroperitoneum, especially in adults, is exceedingly rare, with so few cases that they represent a statistical anomaly.^{1–3} Inversely, paragangliomas are much more common, and are associated with many familial conditions and are frequently characterized by catecholamine hypersecretion.⁴ Intraoperative manipulation or ligation of hormonally active tumors can lead to profound hemodynamic shifts. Patients may exhibit chronic alpha receptor downregulation due to persistent catecholamine exposure, resulting in marked hypotension following resection. Preoperative preparation typically

includes alpha blockade and, if necessary, cautious beta blockade.^{5, 6} Anesthetic induction must also be approached carefully due to the risk of triggering catecholamine surges.⁶ The postoperative period is equally delicate. Blood pressures are labile in most patients following tumor excision, perhaps due to persistent alpha receptor desensitization, residual vasodilatory medications, and relative hypovolemia.^{4, 5, 7} Conversely, transient hypertension and hypoglycemia may also occur due to abrupt shifts in catecholamine and insulin dynamics. Glucose monitoring is crucial as perioperative hyperglycemia (especially > 200 mg/dL) is associated with increased mortality, postoperative myocardial infarction, and longer hospital stays.⁸

Case Presentation

A 52-year-old male with a medical history of insulin-dependent type 2 diabetes mellitus, hypertension, hyperlipidemia, and schizoaffective disorder and no personal medical history or family history of malignancy; presented for elective surgical excision of a retroperitoneal mass. The mass was initially incidentally identified on trauma imaging following pedestrian-on-vehicle MVA where the patient sustained multiple injuries. Cross-sectional imaging revealed a well vascularized retroperitoneal mass abutting the third portion of the duodenum and near the inferior vena cava, raising concern for a functional sympathetic paraganglioma.

Preoperative workup was initiated given the suspicion for catecholamine-secreting pathology. Endocrinology was consulted for optimization of glycemic control and perioperative risk stratification. The patient had persistently elevated random blood glucose levels (ranging from 130–230 mg/dL) and poor chronic glycemic control, with recent hemoglobin A1c values of 10.9% (2 months prior), 11.6% (3 months prior), and 7.5% (1 year prior), corresponding to an estimated average glucose of 266 mg/dL at the time of surgery. Biochemical evaluation demonstrated elevated 24-hour urinary norepinephrine (207 µg/24 hr) and normetanephrine (774 µg/24 hr), as well as mildly elevated plasma normetanephrine (172.3 pg/mL) and metanephrine (55.6 pg/mL) one month prior to surgery. These findings, in combination with the radiographic appearance of the mass, further supported the differential diagnosis of a functional paraganglioma.

Given the hemodynamic implications of tumor manipulation and catecholamine release, the patient was initiated on preoperative alpha-adrenergic blockade with doxazosin 4 mg daily and beta blockade with propranolol 20 mg three times daily. Non-selective beta-blockade was administered as the patient had previously been on metoprolol 100 mg daily for hypertension. Clonazepam 0.5 mg three times daily was continued to reduce preoperative anxiety and minimize sympathetic stimulation.

The patient underwent laparoscopic resection under general anesthesia without intraoperative complication. There was no gross invasion into adjacent structures. Complete resection was performed by a fellow-ship trained surgical oncologist and was achieved using a harmonic scalpel, and the specimen, measuring 92 g, 6.5 x 6.3 x 3.7 cm was retrieved via a hand port (Fig. 1). The operative field was hemostatic, and all incisions were closed in standard fashion. The patient was extubated and

transferred to the post-anesthesia care unit in stable condition. Postoperatively, the patient remained persistently hypertensive, with systolic pressures in the 140–150 mmHg range with continued glycemic variability despite insulin titration. His course was otherwise uneventful, and he was discharged to home on postoperative day 3. Final histopathologic analysis of the resected specimen demonstrated a well-circumscribed vascular lesion composed of variably sized, thin-walled, dilated vessels lined by a single layer of flattened endothelial cells; No evidence of neuroendocrine tissue or malignancy was identified (Fig. 2). This confirmed that the lesion was a benign vascular tumor, a cavernous hemangioma, rather than a sympathetic paraganglioma. Follow-up was conducted weekly and interval imaging did not show recurrence of mass at one month follow-up.

Gross pathology of the resected retroperitoneal mass. Serially sectioned surface of a well-circumscribed mass (92 g; 6.5 × 6.3 × 3.7 cm) resected laparoscopically. Cut surfaces are tan-brown with focal congested, hemorrhagic areas, consistent with a vascular lesion. Scale bar: 3 cm.

Histopathology of the resected mass. Hematoxylin and eosin–stained section (original magnification ×100) showing a well-circumscribed vascular proliferation composed of variably sized, thin-walled, dilated, blood-filled vascular channels lined by a single layer of flattened endothelial cells, without cytologic atypia. No neuroendocrine tissue, chief-cell nesting or malignancy is identified, findings diagnostic of a cavernous hemangioma.

Discussion and Conclusions

This case illustrates the diagnostic ambiguity and perioperative complexity that arise when a benign retroperitoneal cavernous hemangioma presents with the clinical signature of a functional paraganglioma. Three features converged to make paraganglioma the leading preoperative diagnosis: a hypervascular retroperitoneal mass on cross-sectional imaging, elevated urinary catecholamines with mildly elevated plasma metanephrines, and a background of hypertension and prominent anxiety. Because the imaging appearance of retroperitoneal hemangiomas is nonspecific and overlaps with hypervascular neoplasms including paraganglioma, preoperative discrimination on imaging is generally not possible; definitive diagnosis is histopathologic.⁹

The biochemical picture reinforced the suspicion in a physiologically coherent way. Because extra-adrenal sympathetic paragangliomas downregulate phenylethanolamine N-methyltransferase, they secrete predominantly norepinephrine and normetanephrine rather than epinephrine, and this patient's disproportionate urinary norepinephrine and normetanephrine elevations fit that noradrenergic pattern.¹⁰ The elevations were nonetheless only mild to moderate and were plausibly confounded by chronic hypertension, prominent anxiety, continued antihypertensive therapy, and the physiologic stress of recent trauma; a reminder that catecholamine and metanephrine assays are sensitive but imperfectly specific, and that borderline values accompanying a hypervascular mass rarely settle the question on their own.

The decision to proceed with alpha-blockade-first preparation was appropriate despite the ultimately benign pathology. Unprepared resection of a functional paraganglioma carries a materially higher risk of catecholamine-driven cardiovascular catastrophe than the hemorrhagic risk posed by a hemangioma, and adequate preoperative blockade is what reduces that mortality to the low single digits.^{4, 5} It is worth noting that the current PPGL literature increasingly debates whether routine blockade is necessary in patients with only marginal biochemical elevations, but consensus guidance continues to recommend it for suspected functional tumors, and this case fell squarely within that indication.^{4, 11}

The preparation a functional paraganglioma demands is itself worth spelling out, because it explains why a lesion that proved benign warranted so elaborate a pathway. Guideline-concordant preparation pairs 7–14 days of alpha-adrenergic blockade with deliberate volume repletion and a high-sodium diet, reversing the chronic catecholamine-induced volume contraction that otherwise precipitates severe hypotension once the tumor is devascularized.⁵ Beta-blockade is added only after alpha-blockade is established, because blocking β_2 -mediated vasodilation while α -receptors remain unopposed can drive a paradoxical hypertensive crisis – the same physiology that makes unopposed beta-blockade hazardous in other catecholamine-excess states. For that reason, cardioselective β_1 agents are preferred over nonselective propranolol, and combined α/β agents such as labetalol are avoided as primary therapy because their β -blocking activity greatly exceeds their α -blocking activity; intraoperatively the short-acting β_1 -selective agent esmolol is favored over labetalol for tachyarrhythmia.^{4, 11} However, in this case, propranolol was administered over the patient's home metoprolol as his systolic blood pressures were proving difficult to control.

Anesthetic planning also avoids drugs that provoke catecholamine release; notably metoclopramide and glucagon, and ketamine because of its sympathomimetic properties. After resection, roughly half of patients develop hypotension from the abrupt loss of circulating catecholamines against residual blockade and downregulated adrenergic receptors, and rebound hyperinsulinemia can produce clinically important hypoglycemia, so glucose is monitored postoperatively.⁶ This patient's course ran the opposite way, persistent hypertension and hyperglycemia; which in hindsight was concordant with a non-functional tumor superimposed on genuine essential hypertension and diabetes.

A second thread was the patient's poorly controlled diabetes and marked perioperative glycemic variability. Perioperative hyperglycemia is independently associated with increased surgical-site infection, reoperation, cardiovascular events, and mortality across general surgical populations, with a dose-response relationship above roughly 180–200 mg/dL; insulin-based control appears to attenuate that excess risk. Contemporary guidance targets perioperative glucose of 100–180 mg/dL and a preoperative A1c below 8%.^{6, 8} Despite active insulin titration, this patient remained hyperglycemic before and after surgery, underscoring how difficult this target can be in complex patients and how catecholamine-related concern for paraganglioma further complicated glycemic interpretation.

The teaching point is that comorbidities and equivocal biochemistry can convincingly imitate a high-stakes diagnosis. When a functional paraganglioma cannot be confidently excluded, preparing the

patient as though the tumor were functional is the prudent course, even when final pathology tells a different story.

Limitations. This is a single case, and biochemical elevations were mild and potentially confounded by chronic hypertension, anxiety, and continued antihypertensive therapy; functional imaging and germline testing were not performed which limited definitive exclusion of a mixed or non-secretory neuroendocrine process to clinical grounds alone.

Conclusion. This case underscores the reality that comorbidities and ambiguous pathology can frequently blur diagnostic clarity. Retroperitoneal hemangiomas are statistical anomalies, while sympathetic paragangliomas, though rare, represent a high-stakes diagnosis. Because its imaging, and occasionally, clinical picture can overlap with a functional sympathetic paraganglioma, and because the consequences of under preparing a true paraganglioma are severe, the safest strategy is to complete guideline-concordant alpha-blockade optimization whenever a functional tumor remains plausible. Definitive diagnosis rests on histopathology.

Abbreviations

MVA
Motor Vehicle Accident
OR
Operating Room

Declarations

Patient Perspective

None as patient could not be reached.

Ethics approval and consent to participate.

The patient was consented and use of the specimen was covered by the approved Wake Forest University School of Medicine tumor bank/biorepository protocol [IRB protocol number: IRB00000435] prior to his procedure, authorizing research use of the resected specimen. Written informed consent for publication of clinical details and accompanying images was obtained from the patient.

Consent for publication.

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Author's contributions. AL assisted PS with data collection, PS was responsible for data collection, analysis and management. AL was a major contributor in manuscript drafting. JL provided the histological examination and interpretation as well as manuscript review.

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Availability of data and materials.

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

Competing interests. The authors declare that they have no competing interests

References

1. Debaibi M, Sghair A, Sahnoun M, et al. Primary retroperitoneal cavernous hemangioma: An exceptional disease in adulthood. Clin Case Rep. 2022;10. 10.1002/ccr3.5850.
2. AlBishi N, Alwhabi M, Elhassan MAM. Retroperitoneal Tumor, a Primary Cavernous Hemangioma: A Case Report. Cureus. 2023;15. 10.7759/cureus.43442.
3. He H, Du Z, Hao S et al. Adult primary retroperitoneal cavernous hemangioma: a case report. *World Journal of Surgical Oncology* 2012 10:1. 2012-12-08;10. 10.1186/1477-7819-10-261
4. Neumann HP, Young WF Jr. Pheochromocytoma and Paraganglioma. N Engl J Med. 2019;381. 10.1056/NEJMr1806651.
5. Fagundes GFC, Almeida MQ. Perioperative Management of Pheochromocytomas and Sympathetic Paragangliomas. J Endocr Soc 2022/02/01;6. 10.1210/jendso/bvac004

6. Chen H, Sippel RS, Pacak K. The NANETS Consensus Guideline for the Diagnosis and Management of Neuroendocrine Tumors: Pheochromocytoma, Paraganglioma & Medullary Thyroid Cancer. *Pancreas*. 2010;39. 10.1097/MPA.0b013e3181ebb4f0.
7. José Viana Lima J, Kater CE. The Pheochromocytoma/Paraganglioma syndrome: an overview on mechanisms, diagnosis and management. *Int Brazilian J Urology: Official J Brazilian Soc Urol*. 2023;49. 10.1590/S1677-5538.IBJU.2023.0038.
8. RG MK, IB S. Perioperative hyperglycemia and risk of adverse events among patients with and without diabetes - PubMed. *Ann Surg*. 2015;261. 10.1097/SLA.0000000000000688.
9. Li A, Wolf EL, Fidvi S. Imaging approach to retroperitoneal vascular tumors: a narrative review. *Abdom Radiol (New York)*. 2025;51. 10.1007/s00261-025-05332-w.
10. Lee SE, Oh E, Lee B, et al. Phenylethanolamine N-methyltransferase downregulation is associated with malignant pheochromocytoma/paraganglioma. *Oncotarget*. 2016;7. 10.18632/oncotarget.8234.
11. Wang J, Liu Q, Jiang S, et al. Preoperative α -blockade versus no blockade for pheochromocytoma–paraganglioma patients undergoing surgery: a systematic review and updated meta-analysis. *Int J Surg (London England)*. 2023;109. 10.1097/JS9.0000000000000390.

Figures

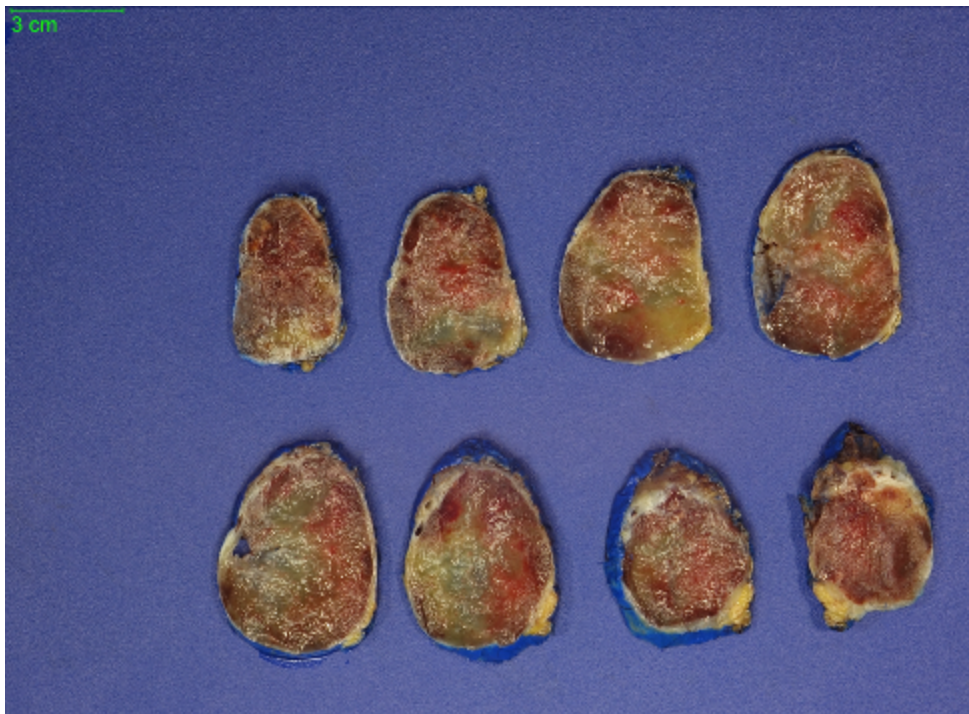


Figure 1

Gross pathology of the resected retroperitoneal mass. Serially sectioned surface of a well-circumscribed mass (92 g; 6.5 × 6.3 × 3.7 cm) resected laparoscopically. Cut surfaces are tan-brown with focal

congested, hemorrhagic areas, consistent with a vascular lesion. Scale bar: 3 cm.

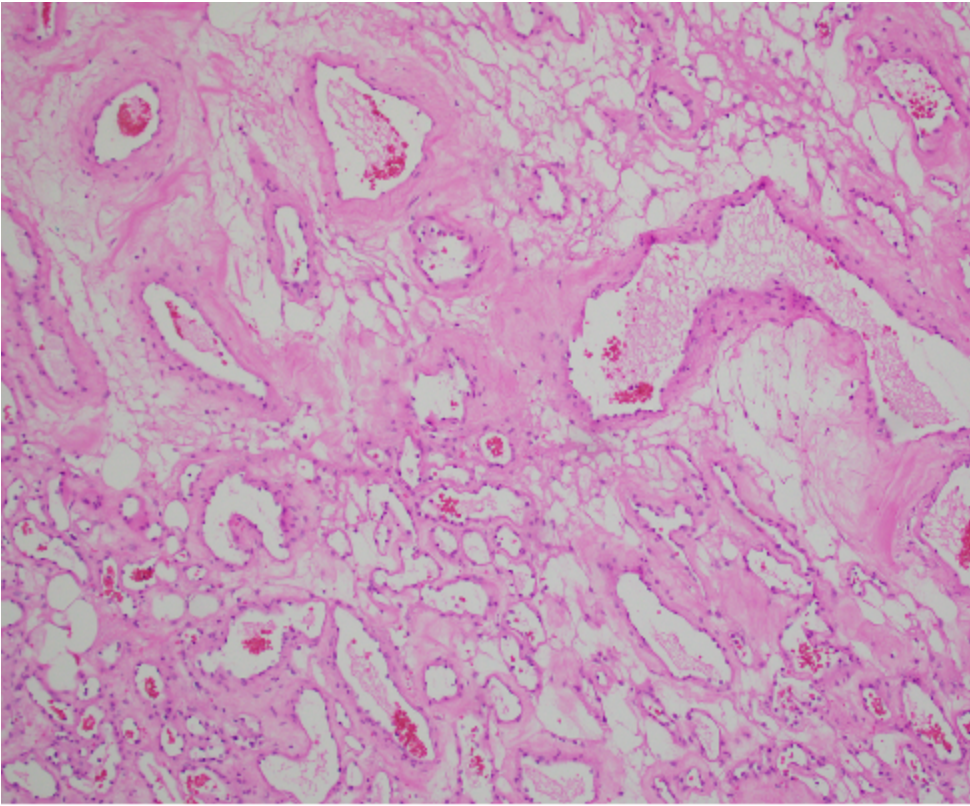


Figure 2

Histopathology of the resected mass. Hematoxylin and eosin–stained section (original magnification $\times 100$) showing a well-circumscribed vascular proliferation composed of variably sized, thin-walled, dilated, blood-filled vascular channels lined by a single layer of flattened endothelial cells, without cytologic atypia. No neuroendocrine tissue, chief-cell nesting or malignancy is identified, findings diagnostic of a cavernous hemangioma.