Functional Hilar Paraganglioma Invading the Renal Veins: A Case Report Mohammed Alfozan

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KEYWORDS

ABSTRACT

A 39-year-old man with poorly managed hypertension was diagnosed with a large left retroperitoneal tumor compressing the renal vein. A CT scan of the abdomen and pelvis with contrast revealed a large, well-defined, capsulated, highly vascular, heterogeneous retroperitoneal para-aortic mass that was compressing the renal vein. Tests for catecholamines in the blood and urine revealed a functioning tumor. A multidisciplinary team, including anesthesiologists, endocrinologists, and urologists, prepared the patient for surgery. Due to the CT findings of the tumor being inseparable from the pancreas, a decision to utilize a midline trans-peritoneal approach. The tumor was compressing the vein but no effect on the blood flow can be noted in that imaging. The tumor noted to be draining into the renal vein during the surgery. The renal vein's width was reduced by less than 20% after the tumor was removed and the vein was reconstructed. The patient was weaned off medications and maintained blood pressure within acceptable ranges.

Introduction

The term "paraganglioma" refers to any tumor of the paraganglia, regardless of the location. The only exception is adrenal medulla paraganglioma, sometimes referred to as pheochromocytoma (1). Although extra-adrenal paragangliomas can develop anywhere in the paraganglia system, they most frequently occur in the retroperitoneum (2). Extra adrenal paragangliomas can be mistaken for a number of other disorders depending on their location and size. A neuroendocrine tumor called a phenochromocytoma often grows on the adrenal medulla's chromaffin cells (3). Paraganglia, which are groups of specialized neural crest cells symmetrically dispersed throughout the aorta in close connection with the sympathetic chain, are the source of extraadrenal retroperitoneal paragangliomas (4). Due to their rarity, these tumors can readily mislead the pre-operative diagnostic, which might have negative consequences, particularly if they develop in unusual places. Paraganglioma lacks a well-defined imaging characteristic; the tumor's vascularity should show a hyper-enhancing mass, but this isn't always the case (5). Given the unusual tumor site and vague symptoms, it is crucial to have a high index of suspicion for diagnosing paragangliomas because a significant percentage of patients encounter significant intraoperative hemodynamic difficulties even after taking premedication (6).Typical symptoms include lumbar discomfort, hypertension, and nonspecific symptoms including headache, impaired vision, heart palpitations, and flushing brought on by elevated catecholamines. There is no hormonal activity in 40% of retroperitoneal pheochromocytomas (7). This study aims to report a rare case of functional paraganglioma invading the renal veins and required reconstruction of the renal vein.

Case presentation

A 39-year-old man who had a large left retroperitoneal tumor compressing the renal vein was referred to our clinic. He has experienced poorly managed hypertension for the past three years. A CT scan of the abdomen and pelvis with contrast revealed a large, well-defined, capsulated, highly vascular, heterogeneous retroperitoneal para-aortic mass that was compressing the renal vein and

could not be ruled out as a result of renal vein invasion (Figure 1). The mass was not distinguishable from the inferior aspect of the pancreatic body or the lateral limb of the adrenal gland. Tests for catecholamines in the blood and urine revealed a functioning tumor.

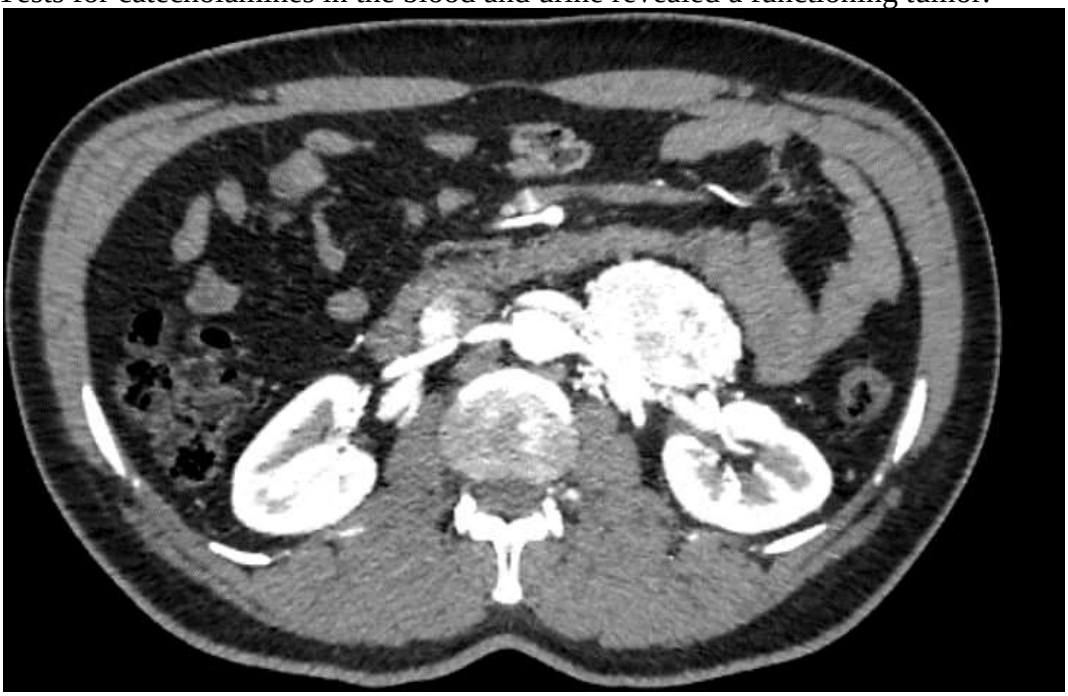


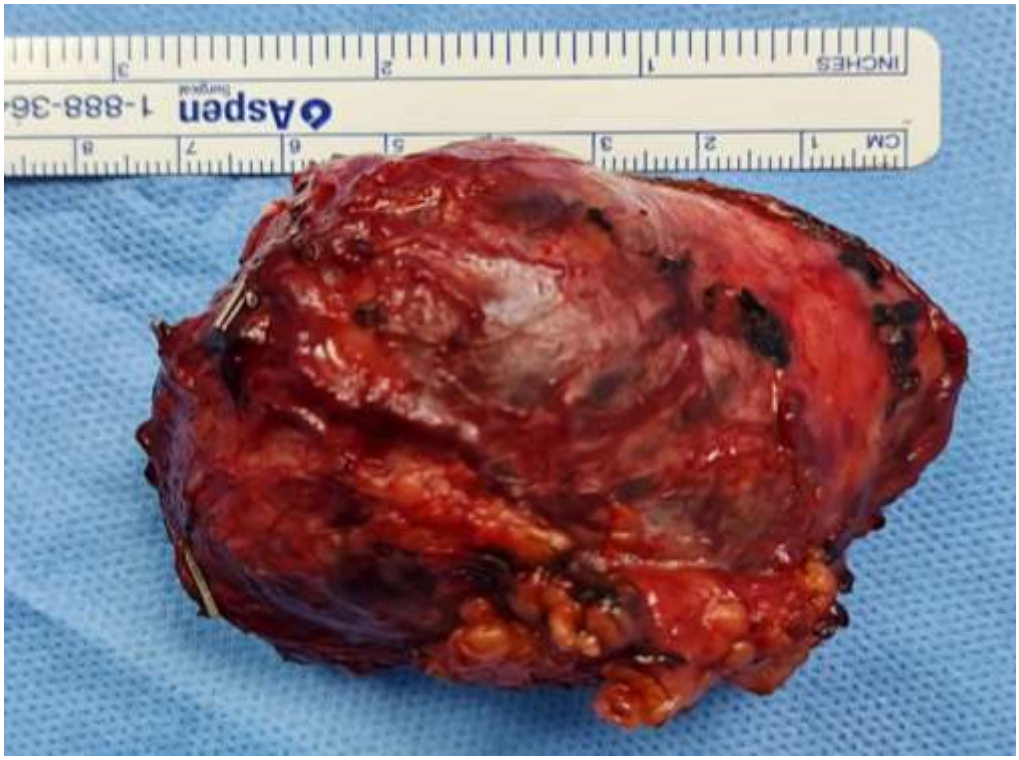
Figure 1: CT scan of the abdomen and pelvis with contrast revealed a large, well-defined, capsulated, highly vascular, heterogeneous retroperitoneal para-aortic mass.

The endocrinologist received the patient's referral. Long-acting beta-blockers and alpha-blockers were initiated as a part of the routine preoperative treatment protocol. The blood pressure was under control after two weeks, and he was prepared for surgery. Anesthesiologists, endocrinologists, and urologists collaborated as part of a multidisciplinary team to prepare the patient for surgery and post-operative care.

We selected a trans-peritoneal technique with a midline incision since the CT scan indicated that the tumor was inseparable from the pancreas. Our team succeeded in separating the pancreatic body from the mass and controlling the collateral vessels (Figure 2). The renal vein wall was within the tumor and inseparable. After a statinsky clamp was used to partially control the vein, the anesthesiologist observed a decrease in blood pressure, indicating that the renal vein was the main route by which the tumor was draining. The renal vein's width was reduced by less than 20% after the tumor was removed and the vein was reconstructed. Inotrops first regulate blood pressure throughout the first 48 hours. Prior to being discharged, the patient was weaned off of all medications and maintained his blood pressure within acceptable ranges.

Figure 2: The mass after separation

The diagnosis of functional paraganglioma was validated by histopathology. Post-operative



catecholamine tests were negative.

Discussion

Pheochromocytoma frequently represents an adrenal tumor that arises from neural crest germinal cells. Retroperitoneal paragangliomas develop from specialized neural crest cells that are dispersed throughout the aorta and connect to the sympathetic chain (8). The majority of patients are in the 30–40 age range, and males are more likely than women to be impacted (9). In the general population, their estimated incidence is 1 per 100,000 person-years, but in hypertension patients, it is 0.1% (10). The organ of Zuckerkandl, bladder wall, retroperitoneum, and heart are the most often occurring extra-adrenal sites for paragangliomas, which can develop anywhere from the base of the brain to the bladder (11).

If there are clinical signs of excessive catecholamine secretion, paragangliomas can be identified early (12). According to pathologic analysis of specimens using the (grading system for adrenal pheochromocytoma and paraganglioma) GAPP, which takes into account histologic pattern, cellularity, comedo-type necrosis, capsular/vascular invasion, Ki-67 labeling index, and catecholamine type, 10 to 30% of paragangliomas may be malignant (13, 14). The existence of distant metastases is still the sole reliable indicator of malignancy (15).

It is challenging to differentiate primary renal tumors from kidney and hilum paragangliomas. Determining the origin of these masses may be difficult using traditional imaging methods like CT, MRI, MIBG scans, or even angiography. Radioisotope imaging may be helpful when there is biochemical and clinical evidence of a tumor that secretes catecholamines but cross-sectional imaging is negative (16).

Variable anatomical presentation and the possibility of a catecholamine spike during operational manipulation have always raised concerns about laparoscopic surgery (17). For paragangliomas in general, open resection with the possibility of a laparoscopic procedure with minor lesions is recommended in surgically advantageous sites (4). Laparoscopic treatment may be linked to safe

intraoperative care, speedy recovery following surgery, and no influence on local recurrence (18). Ten to thirty percent of paragangliomas and pheochromocytomas are malignant and have the potential to spread, however surgical excision will cure the majority of these individuals (19).

The current complicated case comprised a highly vascular retroperitoneal tumor compressing the renal vein, necessitating a multidisciplinary approach to effective management. The patient's tumor was indistinguishable from the pancreas, necessitating a trans-peritoneal approach with a midline incision. Similar to retroperitoneal leiomyosarcoma and liposarcoma (20, 21), the present case included successfully separating the pancreatic body from the mass and controlling collateral vessels. Similar cases of renal hilar paraganglioma were previously reported, nephrectomy was performed in four cases (10, 22-24) and in two cases (25, 26) excision of the tumor only as in our case. The tumor's effect on venous drainage was found following partial closure of the renal vein. Inotropes are also used to maintain blood pressure after surgery. This case study demonstrates the value of a multidisciplinary approach and the efficacy of preoperative care in managing blood pressure and preparing patients for surgery.

Conclusion

The case emphasizes the importance of preoperative pharmacological management in controlling hypertension in patients with paragangliomas, the role of a multidisciplinary team in planning and carrying out complex surgical interventions, and the importance of meticulous intraoperative technique and postoperative care in treating complex retroperitoneal tumors.

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