

Vena cava and renal vein thrombosis with pheochromocytoma: A case report

 **Ramazan Gundogdu,¹**  **Kenan Caliskan,¹**  **Ufuk Uylas,²**  **Nazim Emrah Kocer³**

¹Department of General Surgery, Baskent University Dr. Turgut Noyan Training and Research Hospital, Adana, Turkiye

²Department of Gastroenterological Surgery, University of Health Sciences, Izmir Tepecik Training and Research Hospital, Izmir, Turkiye

³Department of Pathology, Baskent University Dr. Turgut Noyan Training and Research Hospital, Adana, Turkiye

ABSTRACT

Pheochromocytomas are potentially malignant and may manifest with vascular thrombi. We present the treatment of a patient with pheochromocytoma and tumor thrombosis of the renal vein and inferior vena cava. A thirty-eight-year-old male patient was admitted complaining of abdominal pain and headache. High levels of urinary catecholamine were detected. Magnetic resonance imaging revealed left pheochromocytoma with thrombosis of the inferior vena cava and the left renal vein. A left adrenalectomy and a thrombectomy with cavotomy were performed. The pathology results reported a 9.5 cm malignant pheochromocytoma. The patient's symptoms dissipated after the operation and there were no signs of recurrence at the two-month follow-up. Preoperative radiological diagnostic examinations are valuable in the planning of pheochromocytoma surgery. We believe that adrenalectomy and thrombectomy can be performed on pheochromocytoma patients with vascular thrombi with low morbidity.

Keywords: Adrenal; inferior vena cava; pheochromocytoma; renal vein; thrombus.

Cite this article as: Gundogdu R, Caliskan K, Uylas U, Kocer NE. Vena cava and renal vein thrombosis with pheochromocytoma: A case report. *North Clin Istanbul* 2024;11(6):583–585.

Pheochromocytomas are tumors derived from chromaffin cells that release catecholamines and appear most commonly in the adrenal medulla [1]. Pheochromocytoma with vascular thrombi coexist is rarely present and is only reported in case reports in the literature [2, 3]. IVC thrombosis accompanying pheochromocytoma can be treated with adrenalectomy and thrombectomy. The five-year surveillance of life average is 36% in malignant pheochromocytoma [3]. Here, we present a case of pheochromocytoma of the left adrenal gland, in which a thrombus of the renal vein and IVC was diagnosed. The patient was treated by adrenalectomy and total thrombectomy with cavotomy.

CASE REPORT

A thirty-eight-year-old male patient with abdominal pain was admitted to our clinic. Physical examination and blood pressure were normal. His laboratory test results showed dehydroepiandrosterone sulfate of 119.6 µg/dL, total testosterone of 5.56 ng/mL, and 17-hydroxyprogesterone of 1.34 µg/dL. The patient's metanephrine was 134 µg/day, and normetanephrine was 13162 µg/day in 24-hour urine. Abdominal magnetic resonance imaging (MRI) and 18F-fluorodeoxyglucose positron emission tomography (PET) revealed a mass in the left adrenal gland. Thrombus which extended to the IVC through

Received: May 07, 2022

Revised: February 03, 2023

Accepted: December 12, 2023

Online: November 22, 2024

Correspondence: Ufuk UYLAS, MD. Saglik Bilimleri Universitesi, Izmir Tepecik Egitim ve Arastirma Hastanesi, Gastroenterolojik Cerrahi Klinigi, Izmir, Turkiye.

Tel: +90 544 723 36 85 e-mail: ufukuylas@hotmail.com

Istanbul Provincial Directorate of Health - Available online at www.northclinist.com



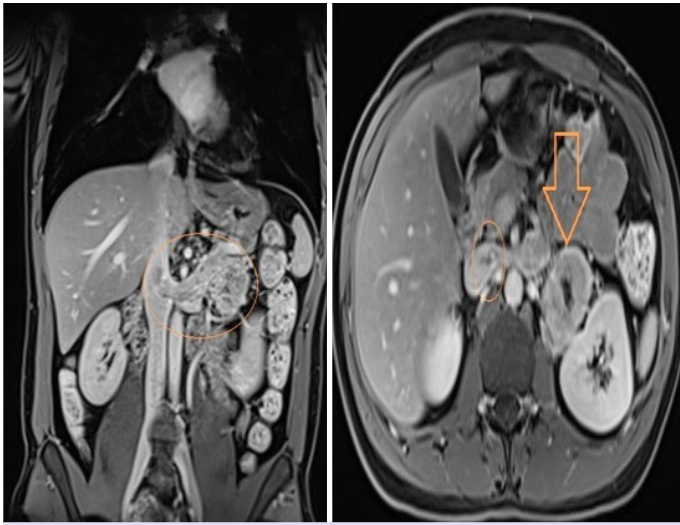


FIGURE 1. Magnetic resonance image of the abdomen.



FIGURE 2. Positron emission tomography images.

the adrenal and renal vein was detected (Fig. 1, 2). The patient was diagnosed with malignant pheochromocytoma and surgery was scheduled. The patient signed the informed consent form prior to the surgery.

Laparotomy was performed by a professor of a general surgeon. Exploration was shown a tumoral lesion of approximately 9 cm in the left adrenal gland and a thrombus extending from the left adrenal vein to the vena cava was detected. The left renal vein was clamped distal to the adrenal vein and the right renal vein and vena cava were clamped closed. A cavotomy was performed. The thrombus in the vena cava was mobile and completely excised. The openings in the renal vein and IVC were closed by 6/0 polypropylene.

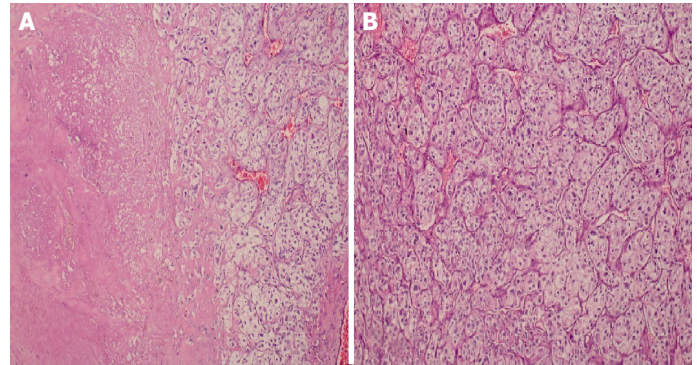


FIGURE 3. (A) Photomicrograph revealing central tumor necrosis (HE x200). (B) Cell nests composed of cells with hyperchromatic nuclei (HE X100).

As there were no further symptoms and no postoperative complications, he was discharged the fifth day. The pathology results reported a 9.5x6x5 cm malignant pheochromocytoma. Synaptophysin and chromogranin were found to be A+. There was staining on the S-100. Ki-67 was <1%. There was no tumor capsule or vascular invasion (Fig. 3). Pheochromocytoma of the adrenal gland scaled score (PASS) was 6. There was no recurrence at the second postoperative month.

DISCUSSION

Pheochromocytomas are associated with varying symptoms depending on the levels of catecholamine secreted. The most common of these are hypertension, headaches, sweating, and palpitations [1]. In some patients, a pheochromocytoma crisis can occur. This involves multiple systems, including the cardiopulmonary, neurological, gastrointestinal, renal, hepatic, and metabolic systems [4]. There were no additional medical findings for our patient and his preoperative symptoms were limited to left flank pain.

Some clinicians argue that preoperative PET is more effective in the detection of additional pheochromocytoma lesions than other types of imaging [5]. MRI scans are also valuable, particularly for the detection of IVC thrombi [3]. In the preoperative period, an MRI on our patient detected both the thrombus extending to the renal vein and IVC and the left adrenal mass. The MRI and PET scan were performed and no additional lesions or metastases were detected.

The preoperative blockade is recommended with functional pheochromocytomas to prevent perioperative cardiovascular complications [6]. Our patient was given daily doses of doxazosin mesylate (4 mg/day) in the 14 days prior to surgery.

The coexistence of pheochromocytoma and vena cava thrombosis is quite rare. In a study of 206 patients who underwent adrenal surgery for malignant tumors, two renal vein thrombi and four adrenal vein thrombi were observed and pathologically confirmed. The average size of the masses was 11.5 cm [7]. In such cases, IVC thromboses can be treated surgically with cavotomy and thrombectomy or conservatively with heparin and warfarin. After the left adrenal mass of our patient was excised, the thrombus extending from the left renal vein to the IVC was completely excised by cavotomy.

Approximately 10% of pheochromocytomas are malignant but differential diagnosis is very difficult. Generally, the detection of invasion and metastasis in nonchromatin tissues is indicative of malignancy [3]. Pheochromocytomas with a PASS score of four or above have the potential for biologically aggressive behavior [8]. The pathology results for our patient reported a 9.5x6 cm capsule and pheochromocytoma without vascular invasion. It was evaluated as a malignant pheochromocytoma. This was due to the PASS score of 6 and because the PET scan indicated the involvement of the thrombus in the IVC.

Systemic chemotherapy can be used to control rapid growth and disease progression. Cyclophosphamide, vincristine, and dacarbazine are standard treatment regimens for these patients [4]. Since our patient's tumor and tumor thrombi were completely excised and there was no residual focus, no adjuvant treatment was planned. However, meticulous follow-up was ensured.

Conclusion

Radiological imaging on surgical planning of pheochromocytoma presenting with vascular thrombi is very valuable. We believe that adrenalectomy and thrombectomy with low morbidity can be applied to these patients.

Authorship Contributions: Concept – UU, RG; Design – KC, RG; Supervision – UU, RG; Materials – RG, KC, NEK; Data collection and/or processing – RG, KC, NEK; Analysis and/or interpretation – KC, RG; Literature review – UU, RG; Writing – UU, RG; Critical review – UU, RG.

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

Conflict of Interest: No conflict of interest was declared by the authors.

Use of AI for Writing Assistance: Not declared.

Financial Disclosure: The authors declared that this study has received no financial support.

Peer-review: Externally peer-reviewed.

REFERENCES

1. Pacak K. Preoperative management of the pheochromocytoma patient. *J Clin Endocrinol Metab* 2007;92:4069–79. [\[CrossRef\]](#)
2. Poudyal S, Pradhan M, Chapagain S, Luitel BR, Chalise PR, Sharma UK, et al. Marker-negative pheochromocytoma associated with inferior vena cava thrombosis. *Case Rep Urol* 2017;2017:6270436. [\[CrossRef\]](#)
3. Shigemura K, Tanaka K, Arakawa S, Hara I, Kawabata G, Fujisawa M. Malignant pheochromocytoma with IVC thrombus. *Int Urol Nephrol* 2007;39:103–6. [\[CrossRef\]](#)
4. Aygun N, Uludag M. Pheochromocytoma and paraganglioma: from treatment to follow-up. *Sisli Etfal Hastan Tip Bul* 2020;54:391–8. [\[CrossRef\]](#)
5. Nockel P, El Lakis M, Gaitanidis A, Merkel R, Patel D, Nilubol N, et al. Preoperative 18F-FDG PET/CT in pheochromocytomas and paragangliomas allows for precision surgery. *Ann Surg* 2019;269:741–7. [\[CrossRef\]](#)
6. Lenders JW, Duh QY, Eisenhofer G, Gimenez-Roqueplo AP, Grebe SK, Murad MH, et al; Endocrine Society. Pheochromocytoma and paraganglioma: an endocrine society clinical practice guideline. *J Clin Endocrinol Metab* 2014;99:1915–42. [\[CrossRef\]](#)
7. Osman Y, Haraz A, El-Mekresh M, Gomha AM, El-Ghar MA, Eraky I. Adrenal tumors with venous thrombosis: a single-institution experience. *Urol Int* 2011;87:182–5. [\[CrossRef\]](#)
8. Thompson LD. Pheochromocytoma of the Adrenal gland Scaled Score (PASS) to separate benign from malignant neoplasms: a clinicopathologic and immunophenotypic study of 100 cases. *Am J Surg Pathol* 2002;26:551–66. [\[CrossRef\]](#)