

Pheochromocytoma: A deadly tumour

Tirath Patel¹

¹Affiliation not available

March 7, 2023

Article type : Case Report

Correspondence : Tirath Patel

Institute: American University of Antigua

Department: Medicine

City: Antigua and Barbuda

Contact: +91-8128250661

Email: Tirathp611@gmail.com

Conflict of interest: None

Declaration: None

Funding: None

Abstract : This case report describes a 45-year-old man with a history of episodic headache, palpitations, and sweating who was diagnosed with pheochromocytoma. The diagnosis was confirmed by elevated levels of plasma catecholamines and metanephrines and imaging studies that revealed a 3 cm mass in the right adrenal gland with evidence of local invasion into surrounding tissues. The patient underwent a laparoscopic right adrenalectomy and was discharged on the third postoperative day with normal blood pressure. Histopathological examination confirmed the diagnosis of pheochromocytoma with evidence of capsular and vascular invasion, but no malignancy markers were identified. The patient was followed up for six months postoperatively with resolution of symptoms and no evidence of tumor recurrence on imaging. This case report highlights the importance of early diagnosis, appropriate management, and follow-up for pheochromocytoma.

Introduction : Pheochromocytoma is a rare tumor arising from the chromaffin cells in the adrenal medulla that produces catecholamines (epinephrine and norepinephrine) in an uncontrolled and excessive manner. It accounts for less than 0.2% of all diagnosed hypertension cases. This tumor can occur sporadically or as part of an inherited syndrome like Multiple Endocrine Neoplasia (MEN) or von Hippel-Lindau (VHL) disease. Pheochromocytoma can present with a wide range of symptoms including episodic headache, sweating, palpitations, anxiety, and hypertension. The diagnosis can be challenging due to the nonspecific nature of the symptoms, and hence, imaging and biochemical testing play a crucial role. Surgical resection is the mainstay of treatment.

Case Presentation : A 45-year-old man presented with a history of episodic headache, palpitations, and sweating for the past 6 months. He also reported feeling anxious and irritable during these episodes. He denied any history of hypertension or other chronic medical conditions. There was no family history of pheochromocytoma or other endocrine disorders. Physical examination was unremarkable except for an

elevated blood pressure of 170/100 mmHg. Laboratory investigations revealed elevated levels of plasma catecholamines (epinephrine: 550 pg/mL, norepinephrine: 700 pg/mL) and metanephrines (metanephrine: 1800 pg/mL, normetanephrine: 1200 pg/mL), confirming the diagnosis of pheochromocytoma. Further investigations were performed to determine the location of the tumor. Abdominal computed tomography (CT) scan showed a 3 cm mass in the right adrenal gland with evidence of local invasion into the surrounding tissues. Magnetic resonance imaging (MRI) of the brain was normal. Genetic testing for MEN and VHL was negative. The patient was started on alpha-blockers (phenoxybenzamine) to control his blood pressure and prevent intraoperative hypertensive crisis. He underwent a laparoscopic right adrenalectomy. Intraoperative findings confirmed the presence of a pheochromocytoma with local invasion into the adjacent tissues. The tumor was removed completely, and the postoperative period was uneventful. The patient was discharged on the third postoperative day with normal blood pressure. Histopathological examination confirmed the diagnosis of pheochromocytoma. The tumor measured 3.5 cm in diameter and showed evidence of capsular and vascular invasion. The mitotic rate was low, and Ki-67 index was 5%. The tumor was negative for malignancy markers. The patient was followed up in the clinic for six months postoperatively. He reported resolution of his symptoms, and his blood pressure was normal on follow-up visits. Repeat imaging did not show any evidence of tumor recurrence.

Discussion : Pheochromocytoma is a rare tumor that can present with a wide range of symptoms, making the diagnosis challenging. Biochemical testing for plasma catecholamines and metanephrines is the cornerstone of diagnosis. Imaging studies like CT and MRI can localize the tumor and determine its extent of invasion. Alpha-blockers are used to control hypertension and prevent intraoperative hypertensive crises during surgical resection. The prognosis of pheochromocytoma is generally good with surgical resection being the mainstay of treatment. However, the risk of recurrence and metastasis is higher in patients with malignant pheochromocytoma.

Conclusion : In conclusion, this case report highlights the diagnostic and therapeutic challenges associated with pheochromocytoma, a rare tumor arising from the chromaffin cells in the adrenal medulla. The diagnosis of pheochromocytoma requires a high index of suspicion, given its nonspecific symptoms. The biochemical testing for plasma catecholamines and metanephrines and imaging studies like CT and MRI play a crucial role in the diagnosis and management of this tumor. Surgical resection is the mainstay of treatment, with alpha-blockers being used to control hypertension and prevent intraoperative hypertensive crises. The prognosis of pheochromocytoma is generally good with surgical resection, although the risk of recurrence and metastasis is higher in patients with malignant pheochromocytoma. This case report underscores the importance of early diagnosis, appropriate management, and follow-up for pheochromocytoma to ensure the best possible outcomes for patients.

References :

1. Neumann, H. P., Young Jr, W. F., & Eng, C. (2019). Pheochromocytoma and paraganglioma. *New England journal of medicine*, 381(6), 552-565.
2. Reisch, N., Peczkowska, M., Januszewicz, A., & Neumann, H. P. (2006). Pheochromocytoma: presentation, diagnosis and treatment. *Journal of hypertension*, 24(12), 2331-2339.
3. Bravo, E. L., & Tagle, R. (2003). Pheochromocytoma: state-of-the-art and future prospects. *Endocrine reviews*, 24 (4), 539-553.
4. Tsirlin, A., Oo, Y., Sharma, R., Kansara, A., Gliwa, A., & Banerji, M. A. (2014). Pheochromocytoma: a review. *Maturitas*, 77 (3), 229-238.
5. Pacak, K., Eisenhofer, G., Ahlman, H., Bornstein, S. R., Gimenez-Roqueplo, A. P., Grossman, A. B., ... & Tischler, A. S. (2007). Pheochromocytoma: recommendations for clinical practice from the First International Symposium. *Nature clinical practice Endocrinology & metabolism*, 3 (2), 92-102.
6. Davutoglu, V., Soyuncu, S., Celkan, A., & Kucukdurmaz, Z. (2004). Left ventricular free-floating ball thrombus complicating aortic valve stenosis. *JOURNAL OF HEART VALVE DISEASE*, 13(2), 197-199.
6. Adler, J. T., Meyer-Rochow, G. Y., Chen, H., Benn, D. E., Robinson, B. G., Sippel, R. S., & Sidhu,

- S. B. (2008). Pheochromocytoma: current approaches and future directions. *The oncologist* , 13 (7), 779-793.
- Vinnakota, S., Jentzer, J. C., & Luis, S. A. (2021). Thrombolysis for COVID-19-associated bioprosthetic mitral valve thrombosis with shock. *European heart journal*, 42(39), 4093-4093.
7. Steinsapir, J., Carr, A. A., Prisant, L. M., & Bransome, E. D. (1997). Metyrosine and pheochromocytoma. *Archives of internal medicine* , 157 (8), 901-906.
8. Manger, W. M., & Gifford, R. J. (2012). *Pheochromocytoma* . Springer Science & Business Media.
9. Walther, M. M., Keiser, H. R., & Linehan, W. M. (1999). Pheochromocytoma: evaluation, diagnosis, and treatment. *World journal of urology*, 17, 35-39.
10. Bryant, J., Farmer, J., Kessler, L. J., Townsend, R. R., & Nathanson, K. L. (2003). Pheochromocytoma: the expanding genetic differential diagnosis. *Journal of the National Cancer Institute* , 95 (16), 1196-1204.

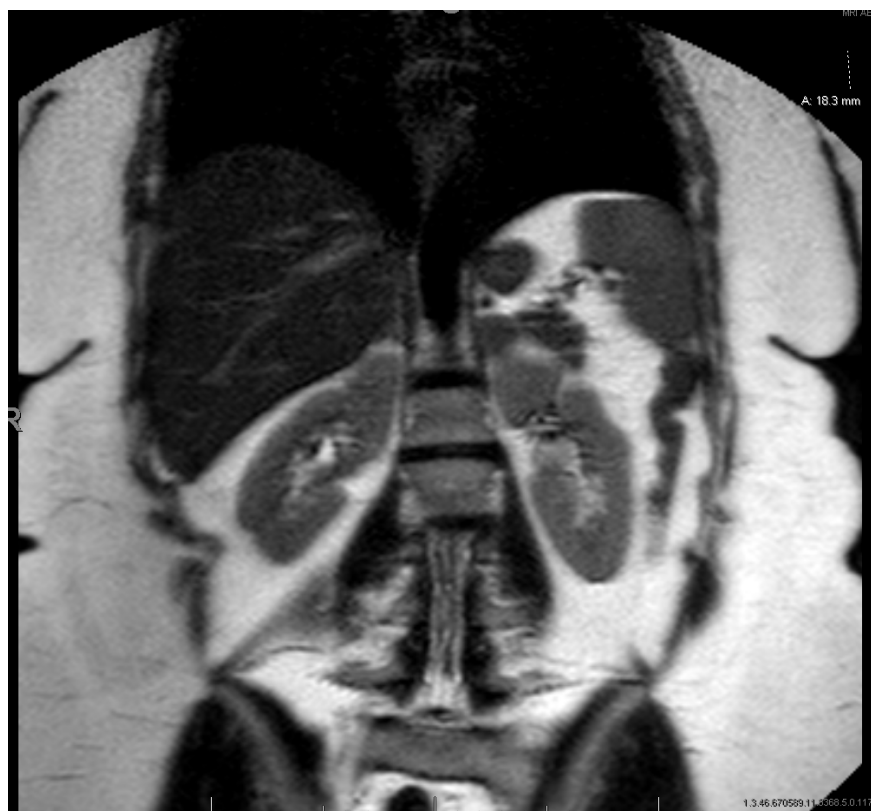


Figure 1: MRI scan representing the clearly viewed pheochromocytoma located in flank

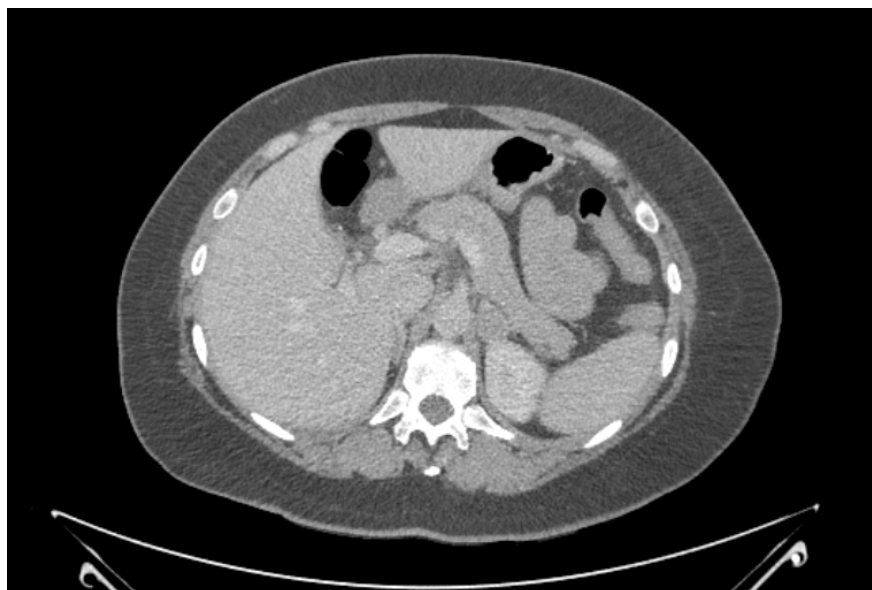


Figure 2: CT scan showing tumor deep down in flank region