Hypertensive Crisis Averted in Pheochromocytoma

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Background

Pheochromocytomas are rare endocrine neoplasms arising from the chromaffin cells of the adrenal medulla and sympathetic ganglia releasing norepinephrine, epinephrine, and dopamine.It is a potentially devastating tumor that is sporadic with an annual incidence of approximately 0.8 per 100,000 person-years most common in the fourth to the fifth decade with an equal male to female ratio. Location of tumor- Abdomen (85-90%), Extra-adrenal also called paragangliomas (10-15%); 10% of the tumors are metastatic.The classic triad of pheochromocytoma: Episodic headache, sweating, and tachycardia but most patients do not present with the classic triad. Less common symptoms include orthostatic hypotension, cardiomyopathy, paroxysmal hypertension.

Case Presentation

53-year-old gentleman presented with right-sided abdominal pain that was sharp and stabbing abdominal pain for one week. Associated symptoms included occasional palpitations and severe headaches for the last few weeks. He denied any fever, chills, cough or shortness of breath, vomiting, CT of the abdomen revealed a right adrenal 39 x 31 x 40 mm solid and cystic mixed enhancing lesion.

Labs showed Urine metanephrine 4015 mcg/ day (55-320), Urine normetanephrine 1961 mcg/day (114-865), Serum cortisol 6.6, ACTH 32, DHEA sulfate 159.4 (38-313), Serum aldosterone 3.4, plasma renin activity 0.1 and 24-hour urine volume was 1400 mL.

Asked to follow up for surgery planning, however he was lost for follow-up upon discharge. One year later, he returned for worse complaints and warranted repeat CT. Right heterogeneous multi-septated adrenal lesion measuring 39 x 43 x 41 mm proved an increase in size. He was started on phenoxybenzamine 10 mg 1 tablet twice a day and doxazosin 1 mg 2 tablets once a day with eventual pheochromocytoma surgery.

Image 1 Repeat CT Abdomen (Green Circle Around Right Pheochromocytoma measuring 39 x 43 x 41 mm)

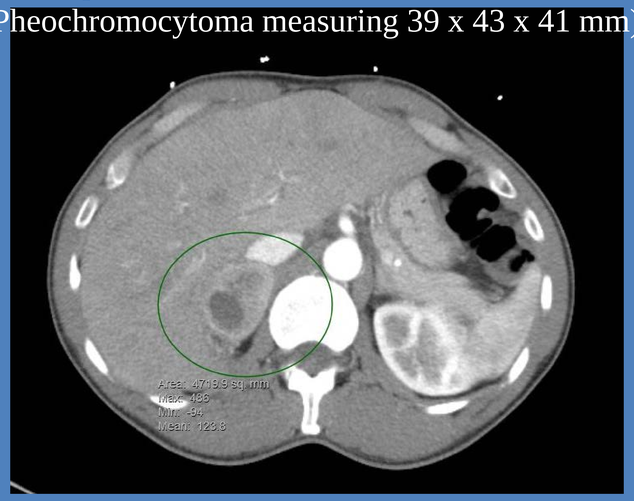
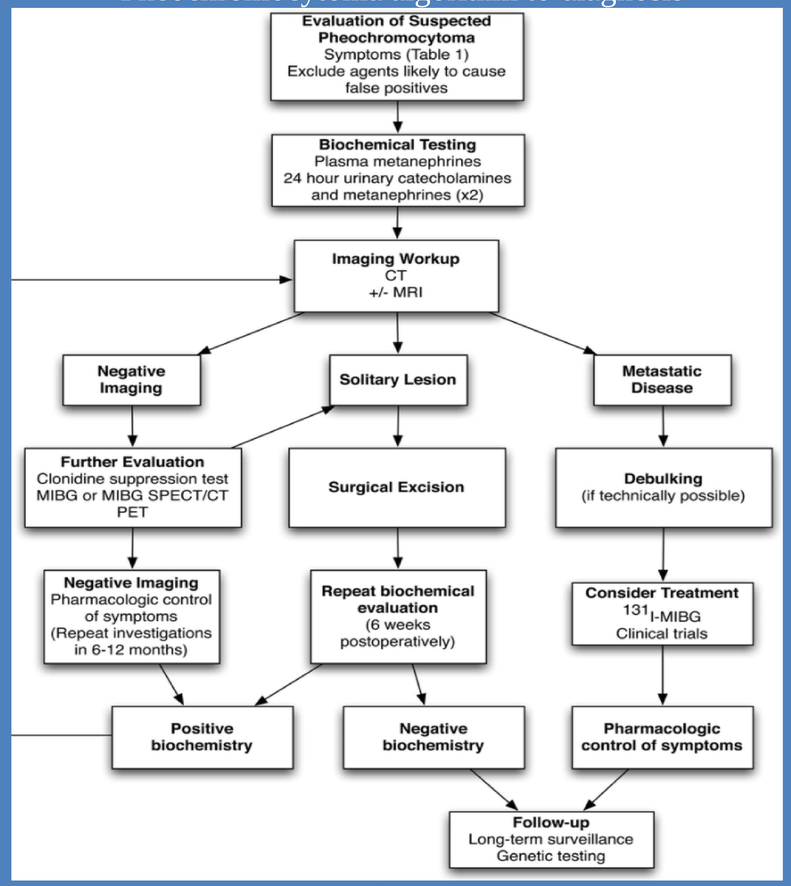


Table 1

Pheochromocytoma algorithm to diagnosis[1]



References

1. Adler, Joel & Meyer-Rochow, Goswin & Chen, Herbert & Benn, Diana & Robinson, Bruce & Sippel, Rebecca & Sidhu, Stan. (2008). Pheochromocytoma: Current Approaches and Future Directions. The oncologist. 13. 779-93. 10.1634/theoncologist.2008-0043.
2. Beard, C. M., Sheps, S. G., Kurland, L. T., Carney, J. A. & Lie, J. T. Occurrence of pheochromocytoma in Rochester, Minnesota, 1950
3. Guerrero, M. A. *et al.* Clinical Spectrum of Pheochromocytoma. *J. Am. Coll. Surg.* (2009) doi:10.1016/j.jamcollsurg.2009.09.022.
4. Stein, P. P. & Black, H. R. A simplified diagnostic approach to pheochromocytoma: A review of the literature and report of one institution's experience. *Med. (United States)* (1991) doi:10.1097/00005792-199101000-00004.
5. Bravo, E. L. Pheochromocytoma: new concepts and future trends. *Kidney Int* . **40**, 544–56 (1991).

Discussion

With a low index of suspicion, measure 24-hour urinary fractionated catecholamines and metanephrines.
With a High index of suspicion, only measure plasma fractionated metanephrines which is what was tested in our gentleman. For measurements of plasma metanephrines, blood should be drawn during a rested stated for at least 30 minutes and in a supine position. There is a strong influence of the sympathetic system in an upright or seated position from stimulation of norepinephrine. This increases risk of false positive results.

Once the biochemical confirmation is made, radiological imaging with CT or MRI of the abdomen and pelvis should be performed to locate the tumor. The definitive treatment is surgery which was unfortunately postponed by our gentleman. Medications that precipitate a hypertensive crisis are dopamine D2 receptor antagonists, opioid analgesics, serotonin reuptake inhibitors, norepinephrine reuptake inhibitors, monoamine oxidase inhibitors, corticosteroids, neuromuscular blocking agents. The risk for hypertensive crisis in our gentleman was high due to loss in follow up and was lucky to not suffer complications of hemorrhagic stroke nor permanent end organ damage.

Alpha-adrenergic blockers like phenoxybenzamine, prazosin, terazosin, or doxazosin are given for at least seven days preoperatively. Beta-adrenergic blockade with propranolol or metoprolol should be given two to three days preoperatively. Dual medical therapy prevents hypertensive crises.

Conclusion

Pheochromocytoma can be incidentally found on routine screening and should not be taken lightly. Hypertensive Emergency is a real risk that luckily our gentleman did not suffer even with loss of follow up for a year. It is important to create an organized pre and post procedure plan to limit the amount of catecholamine release into the bloodstream.