An unusual presentation of pheochromocytoma

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Abstract

Pheochromocytoma is a rare catecholamine-secreting tumor that may arise from chromaffin cells of the adrenal medulla, or extra-adrenal paraganglion tissue. Their prevalence in the general population is estimated at about 1 per 100,000 adults. Characteristically, patient presents with sustained or paroxysmal hypertension and the classic triad of symptoms which include episodic headache, sweating and tachycardia. There are numerous reports in the literature of unusual presentations of benign or metastatic pheochromocytomas including but not limited to multisystemic failure, hypertensive crisis, arrhythmias, catecholamine-induced myocarditis and cardiomyopathy, myocardial ischemia and infarction, pulmonary, cardiovascular, gastrointestinal, nephrologic and neurologic emergencies. This is the case of a 43-year-old male patient with previous medical history of hypertension. He went to a nearby clinic for evaluation of 6 day abdominal distention associated with nausea and vomits and pressure sensation on the epigastric area. Patient was admitted to an intensive care unit (ICU) with diagnosis of hypertensive crisis and upon arrival to the ICU an abdominal computed tomography (CT) scan was done where a 7.7 cm heterogeneous mass at the right adrenal gland was described, renal cell carcinoma was suspected. Patient was then transferred to our center. Upon arrival anti-hypertensive medication, nicardipine, was started. A new abdominal CT revealed a large right adrenal gland hemorrhage. Pheochromocytoma was considered among the working diagnosis. He was started on phenoxybenzamine and beta-blocker therapy. Urine/plasma metanephrines, catecholamines, creatinine and vanillylmandelic acid (VMA), were done. A magnetic resonance imaging (MRI) confirmed that the mass was of suprarenal origin. The diagnosis of pheochromocytoma was finally done. His hospital course was also remarkable for acute renal failure (ARF), supraventricular tachycardia, elevated liver transaminases and pancreatic enzymes, and mechanical intestinal obstruction. After acute clinical conditions resolved, patient tolerated diet and was discharged to continue outpatient therapy with phenoxybenzamine and metoprolol, to undergo surgery in the mainland United State of America (USA).

Key words: Pheochromocytoma, multiorgan failure, hypertension, catecholamines, adrenal gland mass, methanephrines.

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Case Report

A 43-year-old male patient with previous medical history of hypertension went to a nearby clinic for evaluation of 6 day abdominal distention associated with nausea and vomits and a pressure sensation on the epigastric area. He was admitted to ICU with diagnosis of hypertensive crisis and upon arrival to the ICU an abdominal CT scan was done where a 7.7cm heterogeneous mass at the right adrenal gland was described, renal cell carcinoma was suspected. Patient was then transferred to our medical ICU where anti-hypertensive medication, nicardipine, was started. At initial evaluation in our ICU his physical exam was remarkable for an acutely ill appearance patient with dry oral mucosa, diminished bowel sounds and associated tenderness to deep palpation at epigastric and left upper quadrant abdominal area. A new abdominal CT scan revealed a large right adrenal gland hemorrhage (**Figure 1**), in addition to previous findings, pheochromocytoma was considered the working diagnosis. He was started on phenoxybenzamine and beta-blocker therapy. Urine and plasma metanephrines, catecholamines, creatinine and VMA were obtained, and found subsequently elevated (**Tables 1** and **2**). An abdominal MRI (**Figures 2** and **3**) confirmed that the mass was of suprarenal origin. Hence the diagnosis of pheochromocytoma was established.

His hospital course was also remarkable for ARF of pre-renal origin, which resolved with isotonic fluids. In addition, he had episodes of supraventricular tachycardia, elevated liver transaminases and pancreatic enzymes, and mechanical intestinal obstruction. After acute clinical conditions resolved, patient tolerated diet and was discharged with ambulatory therapy with phenoxybenzamine and metoprolol, to undergo surgery in the mainland USA. Approximately one month after been discharged from our institution the patient underwent surgery where a pheochromocytoma with internal bleeding was removed from the right adrenal gland. Histopathology was reported as benign with no local invasion. After discharge home patient has remained free of all hypertensive medications.

Discussion

Pheochromocytoma is a rare catecholamine-secreting tumor that may arise from chromaffin cells of the adrenal medulla, (1) or extra adrenal paraganglionic tissue. It presents as paroxysmal or sustained hypertension episodes in young to middle-age patients, probably occurring in less than 0.2% (2-3) of patients with hypertension. Its prevalence in the general population is estimated at about 1 per 100,000 adults. Pheochromocytomas produce, store, and secrete catecholamines. The clinical features are due predominantly to the release of catecholamines. (4-6) Pheochromocytomas are highly vascular. Characteristically, patient presents with sustained or paroxysmal hypertension and the classic triad of symptoms which include episodic headache, sweating and tachycardia. (6-7) Most patients come to medical attention as a result of a hypertensive crisis, (6) paroxysmal symptoms suggestive of seizure disorder or anxiety attacks, or hypertension that responds poorly to conventional treatment. Less commonly, unexplained hypotension or shock in association with surgery or trauma will suggest the diagnosis. (4) There are numerous reports in the literature of unusual presentations of benign or metastatic pheochromocytomas including but not limited to multisystem failure, hypertensive crisis, arrhythmias, catecholamine-induced myocarditis and cardiomyopathy, myocardial ischemia and infarction, pulmonary, cardiovascular, gastrointestinal, nephrologic and neurologic emergencies. (4-6,8) In rare cases, as in our patient, multiorgan system failure can be the main manifestation of a pheochromocytoma. (6) Patients may exhibit complications such as: cardiovascular (myocardial infarction, arrhythmias, hypertensive crisis, congestive heart failure, etc), pulmonary (pulmonary edema, acute respiratory distress syndrome), gastrointestinal (pancreatitis, cholecystitis, megacolon, ileus, peritonitis, perforation, bleeding, etc), renal (acute renal failure, hematuria, renal artery stenosis, etc), neurologic (seizures, hemiplegia, muscle weakness), metabolic (diabetes ketoacidosis, lactic acidosis). Gastrointestinal complaints as initial manifestation may be considered related to high levels of circulating catecholamines resulting in decrease intestinal peristalsis, motility, and tone, intermittent constipation ileus or a megacolon. (4,6) Hemorrhage of the tumor can lead to excretion of vast amounts of catecholamines and worsened clinical picture; in which emergency surgery would be required and delay in diagnosis can be fatal. (8) In non emergent cases, surgery for these subjects should be planned with experienced surgeons and patient should be clinically stable, as a catecholamine storm could be developed at the time of the surgery.

This case represents an unusual presentation of a pheochromocytoma, one of multiorgan involvement with acute abdominal non-specific complains, hypertensive crisis, ARF, arrhythmias, and an initial working diagnosis of renal cell carcinoma in a 43-year-old male patient with hypertension as previous medical history whom conventional therapy failed to improve clinical presentations and in which symptoms were considered to be unspecific for the clinician that has not seen so frequently these tumors.

Table 1. Urine catecholamines and metanephrines

	Value	Units	Ranges
Catecholamines			
Norepinephrine	>1,446	μg/24 hr	15-100
Epinephrine	>1,033	μg/24 hr	2-24
Dopamine	385	μg/24 hr	52-480
Norepinephrine+epinephrine	>2,479	μg/24 hr	26-121
Metanephrines			
Metanephrines	>9,692	μg/d	58-203
Normetanephrines	>9,578	μg/d	88-649
Total metanephrines	>19,270	μg/d	182-739
Vanillylmandelic acid	64.4	mg/24 hr	<6

Table 2. Plasma catecholamines and metanephrines

	Value	Units	Ranges
Catecholamines			
Norepinephrine	>8,000	pg/ml	112-658
Epinephrine	>4,697	pg/ml	<50
Dopamine	<40	pg/ml	<10
Metanephrines		pg/ml	
Metanephrines	>937	pg/ml	≤57
Normetanephrines	>2,046	pg/ml	≤148
Total metanephrines	>2,983	pg/ml	≤205

Figure 1. Abdominal CT scan with large right adrenal gland hemorrhage, coronal view

Figure 2. Abdominal MRI showing a 7.1 cm x 7 cm x 7.2 cm right adrenal mass lesion

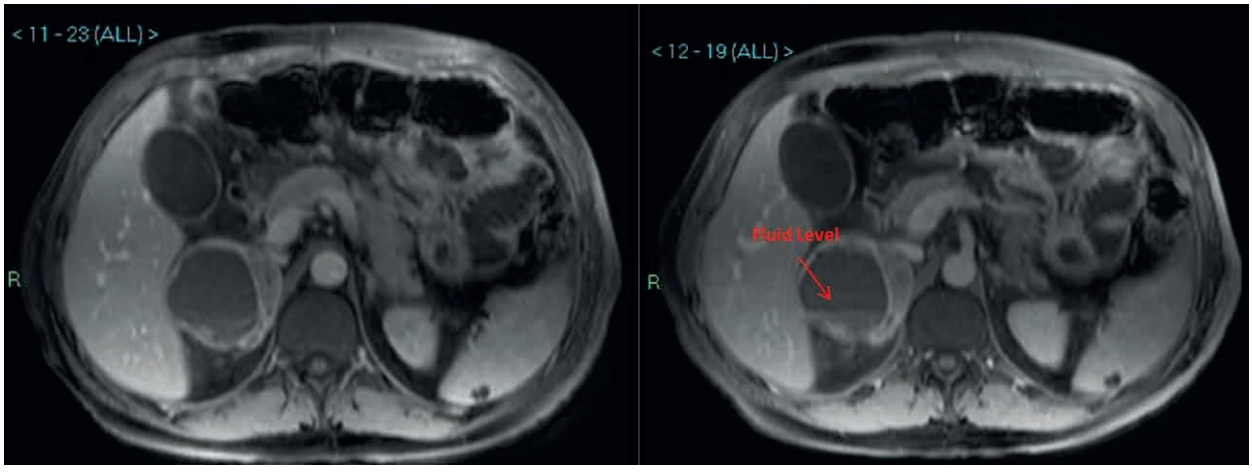
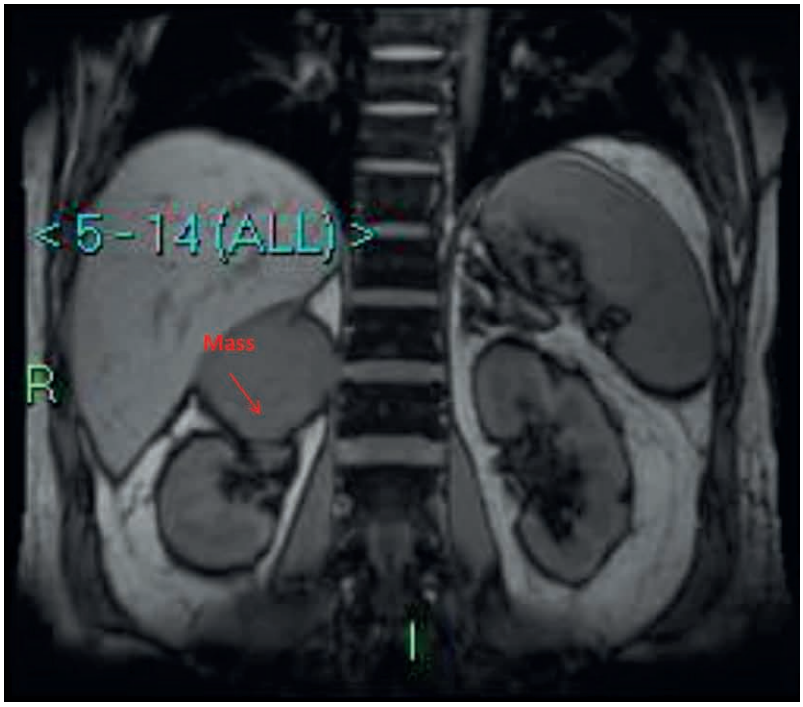


Figure 3. Abdominal MRI, coronal view, showing the right adrenal mass lesion



References

1. Li C, Xu YM. Spontaneous intraperitoneal bleeding caused by adrenal pheochromocytoma. Chin Med J (Engl) 2009;122:2193-5.
2. Stein PP, Black HR. A simplified diagnostic approach to pheochromocytoma. A review of the literature and report of one institution's experience. Medicine (Baltimore) 1991;70:46-66.
3. Pacak K, Linehan WM, Eisenhofer G, Walther MM, Goldstein DS. Recent advances in genetics, diagnosis, localization, and treatment of pheochromocytoma. Ann Intern Med 2001;134:315-29.
4. Sweeney AT, Malabanan AO, Blake MA, de las Morenas A, Cachecho R, Melby JC. Megacolon as the presenting feature in pheochromocytoma. J Clin Endocrinol Metab 2000;85:3968-72.
5. Kolhe N, Stoves J, Richardson D, Davison AM, Gilbey S. Hypertension due to phaeochromocytoma--an ununsual cause of multiorgan failure. Nephrol Dial Transplant 2001;16:2100-4.
6. Brouwers FM, Eisenhofer G, Lenders JW, Pacak K. Emergencies caused by pheochromocytoma, neuroblastoma, or ganglioneuroma. Endocrinol Metab Clin North Am 2006;35:699-724.
7. Young WF Jr. Clinical practice. The incidentally discovered adrenal mass. N Engl J Med 2007;356:601-10.
8. Chan MK, Tse HW, Mok FP. Ruptured phaeochromocytoma--a lesson in acute abdomen. Honk Kong Med J 2003;9:221-3