Pheochromocytoma Presenting with Acute Abdomen

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Abstract

Pheochromocytoma, a neuroendocrine tumor of the adrenal medulla, arising from chromaffin cells, usually presents with the clinical triad of paroxysmal headache, profuse sweating and palpitations, associated with labile hypertension. Here, we present the case of an adult male with an unusual manifestation of pheochromocytoma in the form of acute pain abdomen with nausea and abdominal guarding, mimicking acute peritonitis. He had fluctuating blood pressure recordings. On subsequent investigation, a mass lesion in the left suprarenal area on an abdominal CT scan and a 24-hour urinary metanephrine assay confirmed the diagnosis of pheochromocytoma.

Introduction

Pheochromocytomas are tumors of the adrenal medulla arising from the chromaffin cells. Less commonly, chromaffin cell tumors also arise in extra-adrenal sites (paragangliomas). They are neuroendocrine tumors which lead to a variety of systemic manifestations owing to their ability to secrete and release catecholamines. The classical presentation is constituted by the triad of episodic headache, palpitations and diaphoresis associated with labile hypertension.

Other manifestations include, postural hypotension, visual disturbances, fever, tremors, nausea, weakness, pallor, anxiety, a sense of impending doom, chest pain, and very rarely, pain abdomen.

They may occur either sporadically or as a component of heritable syndromes (in about 25-33% of the cases) like MEN (Multiple Endocrine Neoplasia) 2A and 2B, Neurofibromatosis 1 and VHL (von Hippel Lindau) syndrome.

The tumors are popularly said to follow a ‘rule of tens’: ~10% are extra-adrenal, ~10% are bilateral, ~10% are malignant, ~10% can recur after surgical resection, ~10% are found in children, ~10% are familial, ~10% are not associated with hypertension, ~10% contain calcification.

Case Report

A 40 year-old male patient, farmer by occupation, visited the ER with complaints of sudden onset of pain in abdomen associated with profuse sweating and nausea. On examination, abdominal guarding was found. The case was initially seen by a surgeon. Acute peritonitis was suspected and investigations were duly ordered. No abnormalities were detected on plain erect abdominal radiography and abdomino-pelvic ultrasound. The case was subsequently referred to the physician.

On examination, the patient was febrile and looked anxious. His pulse was bounding with a rate of about 130 beats per minute. His blood pressure recordings were fluctuating. The initial recording yielded a reading of 90/70 mm Hg. A subsequent reading after 15 minutes was 210/120 mm Hg. The patient was duly shifted to the ICU.

Precordial examination revealed a heaving apical impulse with a loud second heart sound. ECG showed features of severe LVH with no signs suggestive of ischemic heart disease. The Creatine Kinase MB value was within normal limits. A subsequent 2D echocardiogram showed severe concentric left ventricular hypertrophy with mild mitral regurgitation and aortic regurgitation.

An fundoscopy was performed and features suggestive of Grade-3 Hypertensive Retinopathy were found.

The investigations obtained are shown in Table 1.

In view of the swinging blood pressures, florid hypertensive features and other constitutional signs, pheochromocytoma was suspected.

An abdominal computerized tomograph (CT) with contrast study revealed a solid well-circumscribed heterogeneous mass lesion measuring 4.7cm x 3.5cm x 4.5cm with areas of necrosis, with no evidence of calcification or hemorrhage in the left suprarenal area (Figure 1).

A subsequent 24 hour urine metanephrines assay yielded a high value of 650µg / 24hours (normal value: <350µg/ 24 hours), confirming the suspicion of pheochromocytoma.

A diagnosis of ‘Left Adrenal Pheochromocytoma’ was made. The
patient was started on oral Prazosin and subsequently on Propranolol. Further surgical intervention was planned.

The patient was further evaluated for syndromic forms of pheochromocytoma. No clinical features or family history suggestive of features of Multiple Endocrine Neoplasia 2A and 2B, Neurofibromatosis-1 or von Hippel Lindau disease were found. The thyroid function tests of the patient were within normal limits (T<sub>3</sub>, 61.09 mg/dl, T<sub>4</sub>, 7.12 µg/dL, TSH - 1.53 µIU/ml). No skin or ocular features of the aforementioned syndromes were encountered. Endolymphatic sac tumor, a feature of von Hippel Lindau disease, was not found on otological examination.

**Discussion**

The manifestations of pheochromocytoma may vary from an asymptomatic state to a life-threatening emergency. The condition can mimic primary cardiovascular, gastrointestinal and neurological disorders owing to the effect of elevated levels of catecholamines on the various systems. It has been rightly called ‘the great masquerader’.

In this patient, who mainly presented with acute severe abdominal pain, nausea, and local guarding, there were very few clinical pointers suggestive of the underlying condition. A high index of suspicion is therefore necessary in such situations, where labile hypertension, fever, tachycardia and diaphoresis are encountered.

Pheochromocytoma presenting de novo as abdominal pain is a relatively rare occurrence, reported in only a handful of studies.2,4

Pheochromocytoma may present as acute abdomen by various mechanisms which may be a direct result of changes in the tumor mass or a consequence of increased circulatory catecholamines.

Hemorrhage and necrosis in the tumor mass may be responsible for acute abdominal pain. This is the most likely mechanism in the case discussed above. Hemorrhagic necrosis may result in rupture and subsequent hemorrhagic shock. Also, it may further worsen the hemodynamic instability due to the sudden drop of circulating catecholamines which often follows tumor necrosis. The most catastrophic of changes in the tumor is the rupture of the pheochromocytoma. It is usually precipitated by increased intratumoral intravascular pressure resulting from paroxysms of hypertension. This may cause intraperitoneal or retroperitoneal bleeding, causing marked hemodynamic instability. It is often lethal, owing to a low index of suspicion and dramatic clinical course. The presentation often mimics that of a ruptured aortic aneurysm.

Pheochromocytomas may also cause acute abdominal pain as a result of the effects of catecholamines on the gastrointestinal system. Catecholamines cause relaxation of the intestinal smooth muscle (with a decrease in the frequency and intensity of peristalsis), constriction of the pyloric sphincter and ileocecal valve, and vasoconstriction in the splanchnic circulation. This underlies the intestinal pseudo-obstruction, bowel ischemia and rarely, gastrointestinal perforation in pheochromocytoma, which may present as an acute abdominal emergency. The hemodynamic instability that accompanies these presentations often leads the surgeon to perform an emergency surgery with insufficient investigations and an uncertain preoperative diagnosis. Such operative interventions are associated with a high mortality rate in the setting of an unrecognized functional pheochromocytoma.

Handling the tumor inadvertently without the knowledge of its functional status can cause massive release of catecholamines into the circulation, precipitating cardiac arrhythmias and malignant hypertension. Peri-operative mortality often results from bleeding, severe post-operative hypertension, shock, acute pulmonary edema and multi-organ failure.5

Hence, it would be prudent to include pheochromocytoma in the differential diagnosis of an abdominal emergency accompanied by hemodynamic instability.

Emergency surgical intervention must be restricted to patients who are unresponsive to maximal medical resuscitation.

The imaging appearances of pheochromocytomas are varied with all imaging modalities, primarily due to the multiple pathologic processes that they may undergo, such as hemorrhage, necrosis, calcification, fibrosis, lipid degeneration and cystic changes. Pheochromocytoma currently remains a true imaging chameleon, and its diagnosis should always be considered in the imaging evaluation of a patient with an adrenal mass.

Radionuclide studies such as MIBG (meta-iodo-benzyl-guanidine) scintigraphy have great specificity in localisation, characterization and staging of pheochromocytomas. But their use is limited as they are expensive and time consuming.6

Demonstration of elevated plasma and urine levels of catecholamines or their metabolites, metanephrines, is the cornerstone for the diagnosis. Urinary 24-hour total metanephrine excretion level is the best single biochemical test for screening of pheochromocytoma.

**Conclusion**

Complete surgical resection of pheochromocytoma prevents lethal hypertensive crises. It has to be borne in mind that this rare neuroendocrine tumor possesses the potential to mimic several conditions and its possibility should be entertained whenever labile hypertension with associated constitutional features is encountered.

Judicious use of radio-imaging studies and demonstration of an elevated 24-hour total urinary metanephrine level are useful in diagnosing this condition.

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**References**


