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Pheochromocytoma Revealed by Acute Coronary Syndrome: A Case Report.

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ABSTRACT

Pheochromocytomas are catecholamines producing tumors arising mostly from chromaffin cells of the adrenal medulla. The prevalence of the tumor is 0.1%-0.6% in the hypertensive population, It has been called 'the great mimic', capable of presenting itself as many other medical conditions. The most common clinical presentation is hypertension, mainly in the form of paroxysmal episodes. Cardiovascular manifestations include malignant arrhythmia and catecholamine cardiomyopathy, mimicking acute coronary syndromes and acute heart failure. We report a case of pheochromocytoma revealed by angina, and acute myocardial infarction without coronary artery disease (CAD).

Abbreviations

CAD, coronary artery disease.
CT, Computed tomography.
ACS, acute coronary syndrome. ECG, electrocardiogram.

Introduction

Pheochromocytomas are rare tumors arising from the adrenal gland with a highly variable clinical presentation but most commonly presenting with episodes of headaches, sweating, palpitations, and hypertension [1].

Typical presentations of pheochromocytoma include episodic headache, sweating, and tachycardia. Several patients with pheochromocytoma manifest paroxysmal hypertension and hypertensive crisis with sustained hypertension or normal blood pressure [2]. Other patients present cardiovascular events, including cardiac arrhythmias (sinus tachycardia or bradycardia and supraventricular arrhythmias), heart failure by toxic cardiomyopathy, angina, and acute coronary syndrome (ACS), in the absence of coronary artery diseases [3].

In patients with myocardial ischemia, catecholamine-induced increased myocardial oxygen demand and coronary vasospasm may play a role. Multiple case reports of pheochromocytoma presenting with acute myocardial infarction with ECG changes or non-ST elevation myocardial infarction (NSTEMI) [4, 5, 6, 7].

We present a 40 -year-old woman admitted with angina, headache and high blood pressure. Elevated cardiac biomarkers and non-ST elevation myocardial infarction on electrocardiography .Subsequent abdominal computed tomography revealed a right adrenal tumor of 12 × 15 cm in size. Four weeks after diagnosis was made the patient underwent right adrenalectomy.

Case report

A 40 -year-old woman, nonsmoker, with a history of hypertension treated with Amlodipine 5 mg, episodic palpitations, headache, and sweating over the previous 4 months. Her family history was negative for hereditary malignancies or syndromes.

The patient came to the Emergency Service complaining of chest pain, headache and palpitations; no dyspnea, nausea or vomiting.

Her physical examination revealed a blood pressure of 205 /115 mmHg, equal in both arms, with a heart rate of 100 bpm. She had no fever, had normal oxygen saturation, a respiratory rate of 20 breaths/min. and normal physical examination results. The electrocardiogram showed sinus tachycardia and non-specific ST-segment and T-wave changes. The patient's chest X-ray was clear with no pulmonary infiltrates or congestion and a normal aortic arc.

Laboratory examination results showed increased levels of troponine 133 ng/L, Creatinine was normal, serum electrolytes were also normal, the rest of results showed no significant abnormalities.

An emergency coronary angiography was performed with evidence of normal coronary arteries.

A pheochromocytoma crisis was suspected because of the previous history of hypertension in a young patient, an abdominal ultrasound showed a mass of 15 cm in her right adrenal area, completed by emergency abdominal CT scan, which revealed a right adrenal mass measuring 12,5 * 15 cm with a heterogeneous and solid structure (figure 1).

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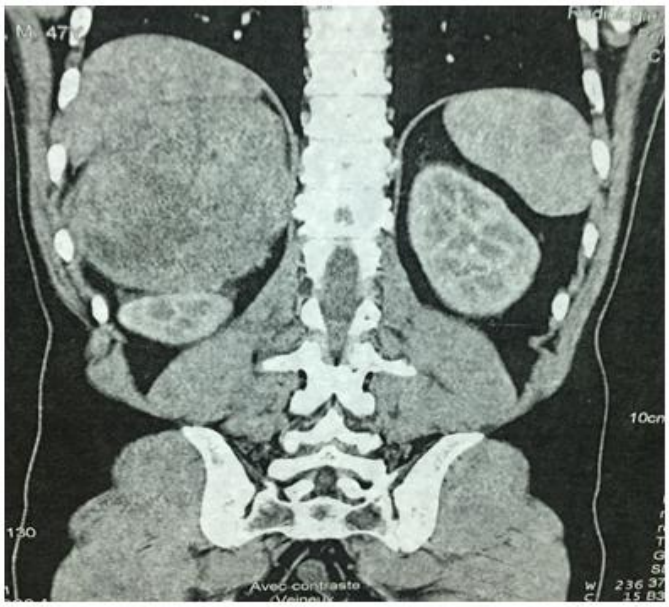

Figure 1.Coronal CT scan showing the right adrenal mass, with heterogeneous and solide structure.

The diagnosis of pheochromocytoma was supported by elevated 24-hour urinary catecholamines. Four weeks after diagnosis was made, a right sided open adrenalectomy was performed (Figure 2).

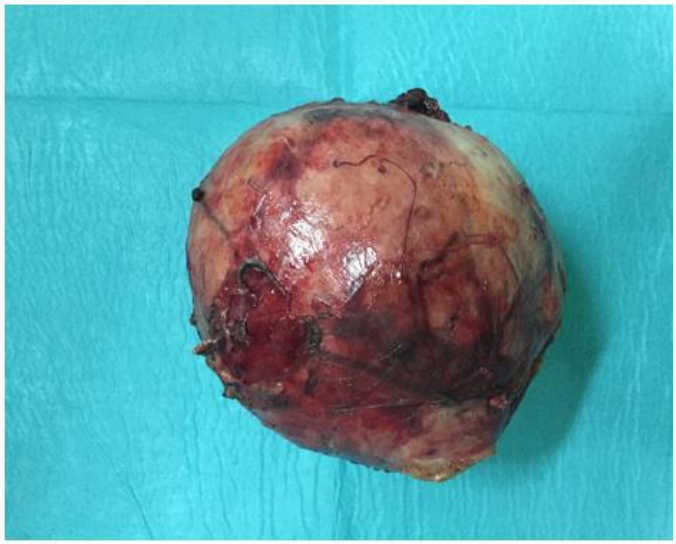

Figure 2. Tumor specimen measuring 12 x 15 cm.

There were no clinical problems during or after surgery. Histological examination of the tumor confirmed the diagnosis of pheochromocytoma. The patient was discharged 4 days after the surgery was performed.

Discussion

Although pheochromocytomas are rare tumors, a relatively high prevalence (up to 0.05%) has been reported in autopsy studies, suggesting that many tumors are missed, resulting in sudden death or premature mortality [8].

The tumors may display numerous clinical manifestations as a result from hemodynamic and metabolic actions of high in circulation catecholamine levels. This secretion can arise in a sudden burst leading to paroxysmal symptoms [9].

The clinical presentation is usually with paroxysms of headache, palpitations, and sweating associated with hypertension. When this triad is absent, pheochromocytoma can be excluded with 99.9% certainly [10].Hypertension can be episodic, but usually is constant and is also associated with orthostatic hypotension especially in the morning [11].

Whilst pheochromoytoma is found in less than 1% of patients with hypertension, between 77% and 98% of patients with pheochromocytoma are hypertensive [12]. In pregnancy pheochromocytoma can be misdiagnose from pre-eclampsia [13].

Different cardiovascular manifestations of pheochromocytoma including sinus tachycardia, sinus bradycardia, supraventricular arrhythmias, and ventricular premature contractions have all been reported. Angina and acute myocardial infarction can occur in the absence of CAD with catecholamine-induced increased myocardial oxygen demand and possible coronary vasospasm [14].

Pheochromocytoma may present as a real medical emergency, mainly where there are complications [15].

Multiple case reports of pheochromocytoma presenting with acute myocardial infarction with EKG changes or non-ST elevation myocardial infarction (NSTEMI) [4, 5, 6, 7], like our case, but pheochromocytoma presenting as acute STEMI in the absence of any obstructive CAD is very unusual [16, 17]. Also we should bear in mind that elevated levels of cardiac troponins are usually present in hypertensive crisis [18].

Other ardiovascular complications of pheochromocytoma include sudden death, heart failure due to toxic cardiomyopathy,hypertensive encephalopathy, acute cerebrovascular accident or neurogenic pulmonary edema [19, 20].

The most appropriate diagnostic tests for patients with suspected pheochromocytoma remain a matter of some debate. Biochemical presentation of excessive production of catecholamines is an essential step for the diagnosis of pheochromocytoma. Traditional biochemical tests include measurements of urinary and plasma catecholamines, urinary metanephrines (normetanephrin and metanephrine), and urinary vanillylmandelic acid (VMA); these tests have a sensitivity of over 76%. Measurements of plasma-free metanephrines (normetanephrine and metanephrine) represent a more recently available test.

However, since catecholamine release is often paroxysmal, a single measurement may give a false sense of security. Sensitivity may be improved by repeating tests two or more times, and especially following a paroxysmal episode [12].

Computed tomography (CT) is the first choice imaging modality in emergency situations; given a strong clinical suspicion, the presence of an adrenal mass is highly indicative of pheochromocytoma. Magnetic resonance imaging (MRI) is recommended in patients with metastatic pheochromocytomas, in patients with surgical clips that cause artifacts when using CT, in patients with allergy to CT contrast, and in patients in who radiation exposure should be avoided [21].

Other valuable diagnostic procedure include Radio-labeled metaiodobenzylguanidine (MIBG) scanning, is a functional imaging modality due to the particular affinity of this substance for chromaffin tissues.

All pheochromocytomas should be resected surgically, if technically feasible. Prior to surgery, patients should be medically prepared in order to avoid potentially lethal complications such as hypertensive crises, malignant arrhythmias and multiorgan failure. Usually, a period of 7-14 days is needed for an adequate preoperative preparation.

There is no available evidence from randomized controlled trials comparing different treatment regimens, but a number of retrospective studies suggest the use of alpha

adrenergic receptor blockers as first line medication that minimizes perioperative complications [22, 23].

Laparoscopic removal of adrenal and extra-adrenal pheochromocytomas is now the preferred surgical technique at experienced centers, since it reduces postoperative morbidity, hospital stay, and expense compared with laparotomy [24, 25], with a complication rate of <8% and a conversion rate of 5% [26].

However, open surgery may be necessary in extreme emergencies involving hemodynamic instability, where rapid action is crucial to patient survival.

After surgery, patients need to be under close surveillance for the first 24 hours, either in a recovery room or in the intensive care unit. The two major postoperative complications are hypotension and hypoglycemia.

If the pheochromocytoma is intra-adrenal, removal of the entire gland is usually performed. In some patients with hereditary pheochromocytomas and small tumors, partial adrenalectomy may be a feasible and safe option [27] and may prevent postoperative adrenal insufficiency and permanent hypocortisolism [28].

Regular follow up is recommended to diagnose recurrence.

Conclusion

Pheochromocytoma may have very variable clinical presentation. Heart failure is one of the most serious expressions, because it engages the vital prognosis. Intensive medical preparation with vasoactive drugs and surgical excision of the adrenal tumor represents the only modality of ultimate cure.

Conflict of Interests

The authors declare no conflict of interest.

Authors Contribution

All authors mentioned have contributed to the development of this manuscript. All authors also declare to have read and approved the final manuscript.

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