

Pheochromocytoma: A Case Report and Literature Review

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Abstract

Objective: We present a case of pheochromocytoma with fatal paroxysmal dyspnea hemoptysis and shock. A systematic review of literature on pheochromocytoma was performed to improve the understanding of the clinical manifestation, diagnosis and treatment of a typical pheochromocytoma.

Methods: The clinical manifestation, diagnostic examination, operation mode and pathological characteristics of the typical pheochromocytoma, were analyzed and summarized. Simultaneously relevant literature was reviewed.

Results: A 29-year-old male was diagnosed with pheochromocytoma with hemorrhagic cysts though without any typical clinical manifestation such as paroxysmal hypertension headache diaphoresis and palpitation. We treated this case as functional pheochromocytoma during the perioperative preparation although there were not enough evidence supporting that the tumor was pheochromocytoma. After sufficient preoperative preparation one of the adrenal grands was surgical resected and the diagnosis was confirmed on histopathology. Postoperation follow-up showed that the patient had a good prognosis after six months.

Conclusion: The manifestations as we reported in this article fatal paroxysmal dyspnea hemoptysis and shock are quite rare as initial manifestations in pheochromocytoma. It showed that the initial manifestations of pheochromocytoma are complicated and deserve an appropriate examination and careful handling, which might reduce the misdiagnosis and surgical risk.

Keywords: Pheochromocytoma; Atypical; Diagnosis

Introduction

The typical manifestations of pheochromocytoma, are the paroxysmal hypertension, headache, palpitation and diaphoresis. This paper is reporting a case of pheochromocytoma with paroxysmal dyspnea hemoptysis and shock as the initial symptoms.

Case Report

A 29-year-old male was admitted to the emergency department with sudden difficulty in breathing coughing up pink frothy sputum and vomiting stomach contents for One day and in a mental confusion for half an hour. He was previously healthy with normal blood pressure.

On physical examination, his blood pressure was 70/50 mmHg with obvious hypoxemia. The X-ray showed a butterfly shaped effusion with shadows in the bilateral pulmonary hilum. Hence, he was diagnosed as having acute pulmonary edema and shock. He was in remission, & after 4 days of assisting ventilation by endotracheal intubation and a breathing machine & anti-shock treatment with supporting treatment. The dynamic monitoring of his blood pressure, had shown the systolic blood pressure was ranging from 98 to 125 mmHg and the diastolic blood pressure ranging from 54 to 78 mmHg; and the heart rate fluctuated between 68 and 105 bpm. The cardiac ultrasound showed his left ventricular ejection fraction was lower than normal and then, it went back to normal one week later. The results of the magnetic resonance imaging (MRI) revealed that the left ventricular free wall the interventricular septum and the papillary muscle were thickened and the end-diastolic volume and end-systolic volume were both reduced. Coronary angiography indicated that the left main coronary artery, and the left circumflex and the right coronary artery had not been seen in significant stenosis. The enhanced CT scan of the adrenal had revealed a lesion occupying a space in the left adrenal (5.4 cm \times 5.3 cm \times 6.5 cm) and a hemorrhagic cyst cannot be exclude. The right adrenal gland showed no abnormal changes. 24-hour urinary catecholamines and their metabolites were at normal levels in repeated tests. The cortisol rhythm aldosterone activity, renin activity and the ratio were within normal limits. Diagnosis as" hemorrhagic cysts of left adrenal gland, pheochromocytoma had not been ruled out", the selective a1 and β 1 receptor blockers were given to him as combination therapy before operation. One month later, the patient was admitted again to hospital for surgery. After expert consultations of urinary surgery and anaesthesia the operation called "the left adrenal cyst resection by laparoscopic" was performed. The blood pressure remained stable during the operation and only when contacting with the tumor the blood pressure, rose to 200/105 mmHg with heart rate 130 bpm for a few seconds. On the right post operation day, the blood pressure fluctuated between 105/65 and 130/75 mmHg, while the preoperative blood pressure fluctuated between 87/47 and 130/76 mmHg. The heart rate changed from 63-116 bpm before the operation to 77-90 bpm after the operation. The α 1 receptor blocker was stopped on the right post operation day and the β 1 receptor blocker was stopped on the 3rd day after operation. And his blood pressure and heart rate were stable. The final diagnosis was confirmed by pathology as adrenal pheochromocytoma with bleeding and cystic change. The macroscopic findings showed: that the tumor measured 6 cm × 5 cm × 2 cm, and the thickness of cystic wall was about 0.2-0.4 cm, and there were some taupe substances in the cyst. Immunohistochemical results showed: CgA/Syn (+), s100 (+), Ki67 (< 5%), P53 (partly +), CK/EMA (-). During the regular follow-up period for half a year, the blood pressure fluctuated between 110/70 and 115/75 mmHg and the heart rate fluctuated between 70 and 75 bpm. The patient had no difficulty in breathing and hemoptysis.

Discussion

This paper reported a case of pheochromocytoma which developed in a young age with uncommon clinical manifestations. Without any symptoms before the onset however the patient had a sudden acute pulmonary edema haemoptysis and shock. Pheochromocytoma arises from the chromaffin cell which is the location where the generation storage metabolism and secretion of catecholamine occur [1-3]. It's also called a neuroendocrine tumor. Large amounts of catecholamines including norepinephrine epinephrine and dopamine are released by pheochromocytoma continuously or discontinuously which cause persistent or paroxysmal hypertension and the functional and metabolic disorders of multiple organs. The clinical manifestations depend on the quantity scale and release of epinephrine and norepinephrine which are secreted by the tumor. Pheochromocytoma is typically characterized by high blood catecholamine, which causes persistent or paroxysmal hypertension with throbbing headache, palpitation and sweating as the main clinical symptoms. However about 15 percent of patients never suffer from the clinical manifestation of high blood pressure. They may suffer from uncommon clinical manifestations such as acute coronary syndrome [4,5] abnormal ventricular wall activity and ventricular dilatation [6] acute abdominal pain [7,8] limb weakness [8] lumbago [9] orthostatic hypotension upper gastrointestinal hemorrhage [10] and diabetes [5,10] or even have no symptoms. There are some reports about Heterologous Cushing's syndrome [11,12] (centripetal obesity) and ectopic adrenocorticotropic hormone (ACTH) syndrome [13,14] but rarely about non cardiogenic pulmonary edema [15]. Typical pheochromocytoma can usually find adrenal occupying lesions by ultrasound and CT, MRI and other imaging examination. Higher levels of catecholamines and their metabolites can be detected in blood or urine. However the value of the examinations above, in the diagnosis of atypical pheochromocytoma is limited.

These less common clinical presentations might be related to the necrosis hemorrhage cystic change of pheochromocytoma. There were case reports about the hematoma and hemorrhagic shock caused by spontaneous rupture and traumatic rupture of the tumor [16,17]. A study found that there was some degree of hemorrhage with 48 in 70 cases of pheochromocytoma patients detected by using spiral computed tomography scan [18]. The case that we reported started with a sudden fatal acute pulmonary edema and hypovolemic shock without typical manifestations, such as high blood pressure palpitations headache and hyperhidrosis previously. The bigger size of the tumor, might lead to spontaneous rupture and hemorrhoea which

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might be related to the imageological change of the adrenal cyst and hemorrhage. Reportedly at least one-third of the life-threatening bleeding tumors have atypical symptoms [19]. The most common symptoms are acute abdominal pain tachycardia, and peripheral vascular contraction and other signs include unstable blood pressure hyperhidrosis, vomiting leukocytosis and fever etc. Clinicians should be alert to pheochromocytoma with internal bleeding, when they encounter severe vasoconstriction, such as chills, sweating and acra pale associated with hypotension. These might be the body's early responses to a large number of secretions of catecholamines.

In the preoperative preparation for suspected pheochromocytoma it has been found that even in the clearly diagnosed patient the death rate of the emergency adrenal resection is far higher than the one of the scheduled surgery after conservative treatment [20]. We treated this case, as functional pheochromocytoma during the perioperative preparation, although there were not enough evidence supporting that the tumor was pheochromocytoma, before the operation. We used selective alpha 1 and beta 1 blockers as the patient's blood pressure was more than one month before the left adrenalectomy and the blood capacity was supplemented actively one week before the surgery. The surgery was performed using a retroperitoneal approach with the patient in the lateral position which could avoid the abdominal organs without affecting the full exposure of the left adrenal. Simultaneously we were prepared, for the possibility that the blood pressure would be sharply fluctuated during the contact and removal of the tumor in the operation. The blood pressure had risen not too badly for once during the surgery and it restored to a steady state soon after giving phentolamine. The patient was well followed-up for 10 months and his blood pressure and heart rate were relatively stable in the normal range and the related symptoms had disappeared (Figures 1-4).



Figure 1: The scanning of transected T1 weighted images showed a $5.4 \times 5.3 \times 6.5$ cm heterogeneous mass in the left adrenal with high signal on the edge and low in the center (arrow) (2014.5.26).

Clinicians should improve their understanding of atypical pheochromocytoma in order to reduce the misdiagnosis and missed diagnosis. At the same time, every patient with suspected adrenal pheochromocytoma should be prepared as a functional tumor whatever the result of the preoperative examination is functional or not.

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Figure 2: The scanning of transected T2 weighted images showed there was a boundary clearly round abnormal signal in the left adrenal, with high signal intensity in the center and low on the edge (arrow) (2014.5.26).



Figure 3: The macroscopic findings: the left adrenal tumor measured $6\text{cm} \times 5\text{cm} \times 2\text{cm}$. The cutting surface was soft and dark tan with focal gray-yellow, and the thickness of cystic wall was about 0.2-0.4 cm. In the cystic there were some taupe substances. 2014.7.1.



Figure 4: Tumor cells formed like a nest and were made up of large cells which were polygonal (arrow) and a small number of small cells, which were cylindrical and oval. The cells contain abundant cytoplasm with basophilic or eosinophilic granules. The boundary of cells is not clear (H&E: \times 400) (2014.7.3).

The blood capacity should be supplemented actively before operation, and invasive arterial blood pressure monitoring is very important during surgery. Try not to squeeze the tumor. Norepinephrine or dopamine should be transfused appropriately so that we can cope with the situation that the intraoperative blood pressure might fluctuate sharply. Finally, the postoperative patients should be regularly followed-up in order to timely avoid relapse which may be due to the incomplete resection of the tumor.

Compliance with ethics guidelines

Authors declare that they have no conflict of interest. This manuscript is a review article and does not involve a research protocol requiring approval by the relevant institutional review board or ethics committee. The authors have no multiplicity of interest to disclose.

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