CASE REPORT

Spontaneous rupture of adrenal pheochromocytoma

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Summary Spontaneous rupture of pheochromocytoma is an extremely rare, but deadly condition with a mortality rate of approximately 32%. We report a case of spontaneous rupture of adrenal pheochromocytoma, which resulted in a retroperitoneal hematoma and respiratory distress. The 69-year-old female visited our emergency department because of progressive abdominal pain for 3 days and fever and shortness of breath for 1 day. Abdominal computerized tomography was performed and an 8 cm mass, with a hematoma in the retroperitoneal space, was revealed. Retroperitoneal exploration was performed on account of progressive respiratory distress and suspected superimposed infection. Elevated blood pressure was noted during anesthesia induction. During the operation, the tumor was found to have originated from the left adrenal gland. A drop in blood pressure was noted after removal of the tumor. Normal blood pressure was restored after fluid challenge and administration of vasopressive agents. The histopathological study of the tumor demonstrated adrenal pheochromocytoma. The patient was in good health, without any sequel at a 6-month postoperative follow up, and eventually fully recovered.

1. Introduction

Pheochromocytoma is an uncommon, catecholamine-secreting tumor derived from chromaffin cells of the adrenal gland, or extra-adrenal chromaffin tissue that failed to involute after birth. The prevalence of pheochromocytoma in patients with hypertension is about 0.1–0.6% and the pheochromocytoma accounts for 4% of
Incidentally found adrenal tumors.\textsuperscript{1,2} The clinical manifestations, resulting from the secretion of catecholamine, include hypertension, tachycardia, headache, pallor, tremor, weight loss, and a feeling of panic or anxiety. Surgical resection of the tumor is the treatment of choice for patients with pheochromocytoma.\textsuperscript{3} Spontaneous rupture of pheochromocytoma is an even rarer event, with only about 50 cases reported in the literature; it is however potentially fatal,\textsuperscript{4} and therefore important. We report a case of spontaneous rupture of a left adrenal pheochromocytoma, presenting with abdominal pain, in a 69-year-old woman with no past history of hypertension, paroxysmal headache or palpitation. The patient recovered very well after surgical excision.

2. Case report

A 69-year-old female visited our emergency department because of progressive abdominal pain for 3 days and fever for 1 day. She was malnourished, with acute malaise, and her body weight was 37 kg. She denied having a past history of hypertension or experience of paroxysmal headache or palpitation. Her blood pressure was 95/57 mmHg, and her body temperature was 38.7°C on arrival. The laboratory analysis revealed an elevated white blood cell count (18.170 x 10\(^9\)/L), C-reactive protein (12.89 mg/L) and blood sugar (13.88 mmol/L), with a decrease in serum albumin (20 g/L). Abdominal computerized tomography (CT) was performed at the emergency department and revealed an 8 x 5 x 4.5 cm\(^3\) retroperitoneal mass with a hematoma anterior to the left kidney (Fig. 1). The abdominal CT failed to identify a direct linkage of the tumor to the adrenal gland (Fig. 2). The patient was admitted to the intensive care unit for further evaluation and treatment. Unfortunately, progressive respiratory distress, high fever, and metabolic acidosis were noted after admission. Retroperitoneal exploration was performed on the next day of admission, on account of respiratory distress and suspected superimposed infection. Elevated blood pressure up to 200/150 mmHg was noted during anesthesia induction. Upon retroperitoneal exploration, a ruptured retroperitoneal tumor, apparently derived from the left adrenal gland, with hemorrhage causing a hematoma was noted. There was no significant bleeding during the operation; however, a sudden drop in blood pressure (60/40 mmHg) was noted after removal of the tumor. Normal blood pressure was restored after fluid challenge, with intravenous administration of dopamine and norepinephrine. The dosages of vasopressor agents were tapered and stopped 4 days after the operation and since then, her blood pressure remained in the normal range. The patient was kept intubated after the operation, due to inadequate lung expansion. A chest X-ray was taken, and chest CT was performed; these showed bilateral lower lobe pneumonia with pleural effusion. The pulmonary condition improved after proper chest care, antibiotic treatment, nutritional support, and bilateral pigtail catheter pleural drainage. She was extubated 8 days after the operation, and discharged from our hospital on the 13th postoperative day. Her blood pressure and blood sugar levels became normal after tumor excision. The histopathological study of the tumor revealed adrenal pheochromocytoma with hemorrhage (Figs. 3 and 4). The patient remained in good health, without any sequel during a 6-month postoperative follow up. The sonography performed 4 months later, revealed no tumor recurrence and the patient refused postoperative pheochromocytoma-related endocrinological studies.

3. Discussion

Various neoplasms can arise from the retroperitoneum with manifold histological diversity, including those of mesodermal, neurologic, and embryonic origin, and the diagnosis of these tumors is often challenging. Sarcomas are most common (43%) among them, followed by lymphomas (23%), benign tumors (11%), undifferentiated malignant tumors (11%), carcinomas (8%), and germ cell tumors (4%).\textsuperscript{5} Spontaneous retroperitoneal hemorrhage is an uncommon
entity and in most cases the underlying cause is from the kidney. Renal angiomyolipoma is the major source of spontaneous kidney hemorrhage.6

Rupture of an adrenal pheochromocytoma is extremely rare and can be lethal, with a mortality rate of approximately 32%. Most of the patients present abdominal pain of acute onset, while some patients complain of lumbar or chest pain.7 The exact mechanism of pheochromocytoma rupture remains unknown. A massive release of catecholamines is probably associated with vasoconstriction in the tumor and subsequent necrosis and hemorrhage. Consequently, elevated intracapsular pressure may result in a tear in the capsule, causing further hemorrhage into the retroperitoneum.8

The clinical diagnosis of ruptured pheochromocytoma may be difficult. Only 30% of patients have been diagnosed preoperatively.9 Several biochemical tests have been used for the diagnosis of pheochromocytoma. The choice of diagnostic test should be based on the clinical suspicion of a pheochromocytoma. Plasma-free metanephrine testing has the highest sensitivity (99%) for detecting a pheochromocytoma, but it has a lower specificity (89%). In comparison, a 24-hour urinary collection for measuring catecholamines and vanillylmandelic acid has, respectively, a sensitivity of 86% and 64% and a specificity of 88% and 95%.1 Ultrasound, CT scanning and magnetic resonance imaging have been suggested for the diagnosis of ruptured adrenal pheochromocytoma. CT scanning is the diagnostic tool of choice, because it is more feasible in the emergency condition as compared with magnetic resonance imaging.10

Surgical exploration has been suggested for the treatment of ruptured pheochromocytoma. In 1976, Van Way reviewed 12 reported patients with pheochromocytoma with hemorrhagic necrosis, with or without rupture. Six patients had no surgery and only one survived, while the other six had immediate surgery with four surviving.11 Surgical approaches for ruptured pheochromocytoma include either emergency or elective surgery. Proper preoperative management of patients with pheochromocytoma is essential to prevent intraoperative complications. Traditional regimens include the blockade of adrenoceptors, and phenoxybenzamine (7–10 days) is often preferred because it blocks adrenoceptors non-competitively. Propranolol can be used when cardiac arrhythmias occur. If blood pressure control is still not adequate, metyrosine, which can decrease catecholamine synthesis, may be used.12 The criteria to ensure adequate preoperative preparation include: (1) keep the blood pressure below 160/90 mmHg for at least 24 hours; (2) blood pressure in the upright position of not below 80/45 mm Hg; (3) no more than one ventricular premature contraction in 5 minutes; and (4) no S-T changes or T-wave inversions for 1 week.13 In 2005, Kobayashi et al reviewed 50 reported cases of adrenal pheochromocytoma rupture (38 cases) or hemorrhage (12 cases).9 They concluded that emergency surgery is associated with a high mortality rate, whereas no mortality has been reported in patients undergoing elective surgery with good control of blood pressure, using α-adrenergic blockers and proper fluid replacement therapy.1 The shortcoming of this study, is that the severity of disease was not accounted for. Patients with life-threatening conditions usually need emergency surgery and these patients would necessarily bear a higher mortality rate. Early control of the adrenal vein is essential while resecting a pheochromocytoma, because this procedure minimizes catecholamine surge during surgical manipulation. Informing the anesthesiologist before the ligation of the adrenal vein is important, because a significant fall in blood pressure may occur at this point.14

With the advent of advanced angiographic equipments and techniques, transcatheter arterial embolization by selective adrenal artery angiography, using a microcatheter, has been successfully applied to hemorrhage control in patients with adrenal tumor bleeding.15,16 Selective tumor excision has been arranged after the stabilization of vital signs and well-conducted preoperative preparation.7

Acute pheochromocytoma rupture is extremely rare and it bears diagnostic, medical and surgical challenges because of the massive release of catecholamines and
hemodynamic instability caused by the hemorrhage. This case report reminds us that pheochromocytoma rupture should be put into consideration in the differential diagnosis of spontaneous retroperitoneal or intraperitoneal hemorrhage. If the vital signs of the patient are stable, proper preoperative preparation with elective surgery may become possible, and yield results for patients.

References