



Arterial embolization for ruptured adrenal pheochromocytoma

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ABSTRACT

Background

Spontaneous rupture of adrenal pheochromocytoma is an extremely rare condition that can be lethal. Optimal treatment in these cases is still unclear.

Methods

We conducted a comprehensive review of MEDLINE articles on ruptured pheochromocytomas to locate all research done on this topic. Based on the literature review and one additional case at European Gaza Hospital, we analyzed clinical presentations, hemorrhage site, tumour side, mortality rate, and treatment options.

Results

In addition to our case, we identified 53 cases of ruptured pheochromocytoma. A review of all 53 cases revealed that 17 (32%) of the patients died, and that no mortality occurred among the 12 patients who received an alpha-blocker (to control high blood pressure) and fluid infusion therapy before surgery. Only 3 patients, including our case, underwent elective surgery after transcatheter arterial embolization (TAE).

Conclusions

Surgical treatment should be considered for ruptured pheochromocytoma. Surgical approaches involve either emergency or elective surgery. It has been reported that emergency surgery is commonly associated with a high mortality rate; no deaths were reported in patients who underwent elective surgery. We therefore consider that, if a patient has hemodynamic instability, TAE can be an effective and a safe procedure for achieving hemostasis and maintaining the patient in good condition until surgery can be performed.

KEY WORDS

Adrenal pheochromocytoma, rupture, arterial embolization, treatment

1. INTRODUCTION

Adrenal pheochromocytomas are uncommon catecholamine-secreting tumours. In general outpatient clinics, the prevalence of pheochromocytoma in patients with hypertension is 0.1%–0.6%^{1–3}. The classic triad of pheochromocytoma presentation is episodic headache, sweating, and palpitations. Pheochromocytoma is typically found with a diverse set of symptoms, which may include anxiety, chest and abdominal pain, visual blurring, papilloedema, nausea and vomiting, orthostatic hypotension, and transient electrocardiographic changes⁴. Spontaneous rupture of adrenal pheochromocytoma is extremely rare, with only about 50 cases reported in the literature to the date of writing⁵. The mechanism of pheochromocytoma rupture is unknown, but high intracapsular pressure may tear the capsule and cause necrosis of the tumour⁶.

Here, we describe a patient with ruptured right adrenal pheochromocytoma presenting with hypertension and abdominal pain, and we review the literature for similar cases. To best of our knowledge, this case is the first to be reported showing ruptured adrenal tumour with large retroperitoneal and subcapsular hematoma of the right kidney and with safe coil embolization for the adrenal and renal arteries.

2. CASE DESCRIPTION

A 42-year-old Palestinian man referred to the European Gaza Hospital emergency room presented with sudden onset of right-side upper abdominal pain radiating to the right flank and accompanied by sweating and palpitations for the preceding 24 hours.

In the 2 weeks before this presentation, the patient had recurrently experienced similar symptoms. He had been admitted to another hospital, where he

underwent an urgent abdominal computed tomography (CT) scan with contrast, which demonstrated a large tissue mass in the right adrenal gland region with intracapsular bleeding and a large hematoma in the right retroperitoneal space (Figure 1) and subcapsular hematoma in the right kidney (Figure 2). At that time, he was urgently referred to Egypt, but unfortunately, he was denied entry.

The patient was then referred to our hospital. On admission, his blood pressure was 170/120 mmHg, and his pulse, 105 bpm. A complete blood count revealed elevated white cells (10,200/ μ L), hemoglobin (10.2 g/dL), hematocrit (38.3%), and platelets (415,000/mL). Blood chemistry showed some abnormal values: total protein, 8.4 g/dL; serum albumin, 5.1 g/dL; aspartate aminotransferase, 32 IU/L; alanine aminotransferase, 41 IU/L; serum creatinine, 1.7 mg/dL; and fasting plasma glucose, 6.3 mmol/L. Urinalysis showed urinary protein 2+ and urinary occult blood 3+. Electrocardiography showed sinus tachycardia. Findings on chest radiography were within normal limits, with a cardiothoracic ratio of 50% and no lung abnormalities. The patient had no medical history of hypertension. Laboratory tests of 24-hour urine demonstrated high vanillylmandelic acid, at 31 mg in 24 hours (reference range: 1–10 mg).

We performed abdominal aortography using a right femoral approach. Selective angiograms of the celiac, superior mesenteric, and right renal artery were obtained, with subsequent selective cannulation of the right renal and adrenal arteries using a Judkins right 6 French catheter (Figure 3).

Because of the subcapsular hematoma in the right kidney, urinary protein 2+, urinary occult blood 3+, an arterial supply of the right suprarenal gland was performed from the right renal and medial and lateral suprarenal arteries. With back and abdominal pain and hypertension continuing despite medical treatment, the patient was at high risk for surgery. We therefore suggested transcatheter arterial embolization (TAE) for the renal and suprarenal arteries.

A TAE using coils was then performed on the medial and lateral branches of the right adrenal and renal artery using 3 French microcatheters. Subsequent dye injection demonstrated complete obliteration of the tumour's vascular supply. Immediately after embolization, the patient's vital signs stabilized, and his blood pressure normalized to 130/90 mmHg. After 48 hours, his abdominal and flank pain had completely resolved.

A follow-up abdominal CT examination 2 weeks post TAE showed significant resolution of the retroperitoneal hematoma. In addition, the tumour was also better delineated than it had been in the imaging done before the TAE procedure (Figure 4).

One month after the coil TAE, the patient underwent elective right adrenalectomy and nephrectomy. Pathology findings confirmed the diagnosis of pheochromocytoma. The adrenal gland tumour was

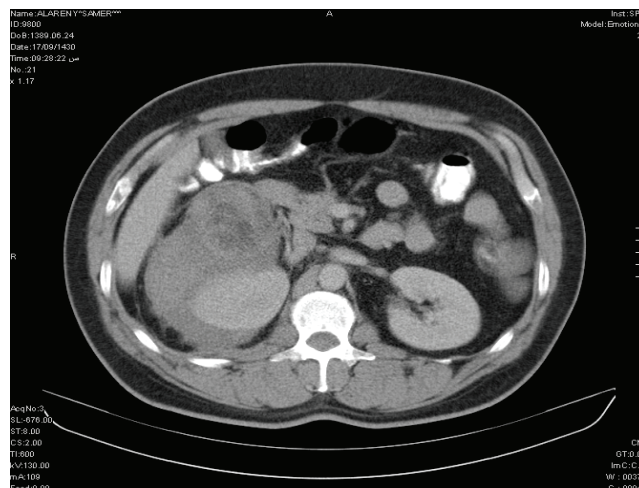


FIGURE 1 Right adrenal mass with retroperitoneal hematoma and intratumoral bleeding.



FIGURE 2 Subcapsular hematoma in the right kidney.

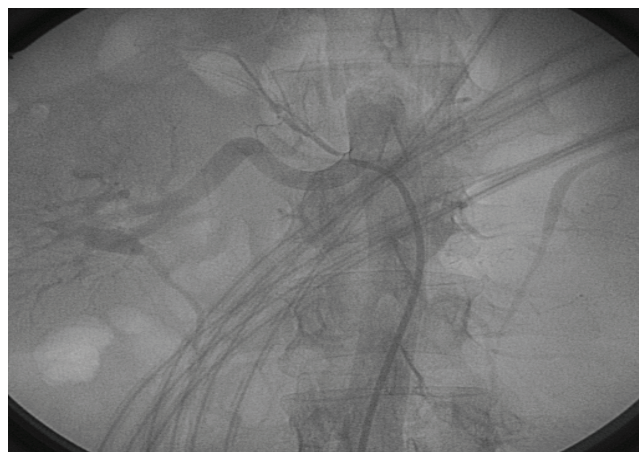


FIGURE 3 Adrenal gland supply by medial and lateral branches of the right adrenal and renal artery.

composed of cells possessing round nuclei and abundant eosinophilic cytoplasm, arranged in a Zellballen

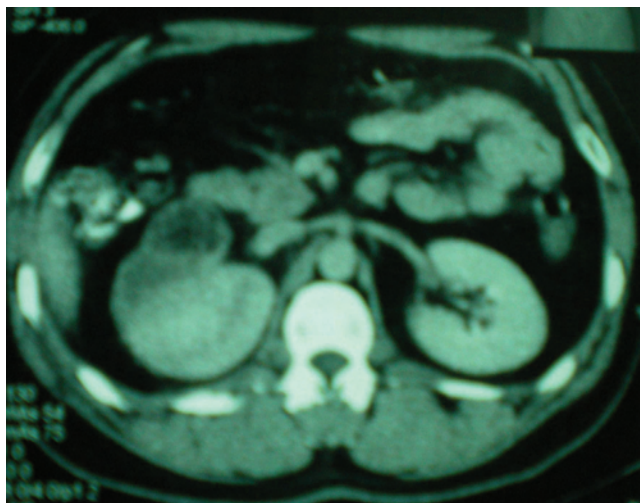


FIGURE 4 Significant resolution of the retroperitoneal hematoma.

pattern. Hemosiderin pigmentation and coagulation necrosis were frequently seen.

3. LITERATURE REVIEW

Since the first description of ruptured pheochromocytoma by Cahill in 1944⁷, a total of 53 cases, including the present one, have been reported in the literature. No difference in frequency between men and women has been reported, and median age of the patients was 50.1 years. Table 1 summarizes the 53 cases^{8–56}.

3.1 Presentation

Analysis of the literature shows that 79% of the patients presented with acute abdominal pain, 19% with chest pain, 24.5% with lumbar pain, and 57% with shock. One case presented with acute myocardial infarction⁵².

3.2 Treatment

Surgical excision was performed in 44 cases. In 12 patients, the surgery was elective after the patient had been stabilized with an alpha-blocker (to control high blood pressure) and fluid infusion therapy. In 3 patients, the surgery was elective after TAE. In 29 cases, the surgery was emergent, and in 9 patients, surgery was not performed.

3.3 Outcome

The mortality rate was high: one third of the patients [17 (32%)] died. No mortality occurred among the 15 patients who received alpha-blocker and fluid infusion therapy, or who were treated by TAE before elective surgery. All 17 who died were among the 38 patients (44.7%) who did not undergo elective surgery after appropriate medical preparation.

TABLE 1 Clinical characteristics of 53 patients with spontaneous ruptured pheochromocytoma

Characteristic	Value
Age (years)	
Median	50.1
Range)	15–80
Sex (n)	
Male	27
Female	26
Tumour location [n (%)]	
Right	30 (56.6)
Left	22 (41.5)
Bilateral	1 (1.9)
Symptoms [n (%)] ^a	
Acute abdomen	42 (79)
Shock	30 (57)
Lumbar pain	13 (24.5)
Chest pain	10 (19)
Surgical setting [n (%)]	
Emergency	29 (55)
Elective	12 (23)
Elective surgery after TAE	3 (5)
Not performed	9 (17)
Hemorrhage site [n (%)]	
Inside tumour	13 (25)
Rupture	
Retroperitoneal	27 (51)
Intraperitoneal	13 (24)
Outcome [n (%)]	
Survived	36 (68)
Died	17 (32)

^a Multiple symptoms reported when applicable.

TAE = transcatheter arterial embolization.

3.4 Transcatheter Arterial Embolization

The literature shows that only 3 patients (including our case) underwent elective surgery after TAE (Table II):

Case 1

A 67-year-old man was admitted to the emergency department with a 1-day history of severe right abdominal and back pain associated with diaphoresis, nausea, and vomiting. Imaging by CT demonstrated a right adrenal mass with retroperitoneal hemorrhage. A TAE using polyvinyl alcohol particles was done to the right adrenal artery, and 2 months later, the patient underwent elective right adrenalectomy⁵³.

Case 2

A 32-year-old man presented with sudden right abdominal pain, and urgent CT imaging demonstrated

TABLE II Summary of the 3 cases of ruptured pheochromocytoma treated with transcatheter arterial embolization (TAE)

	Case		
	1	2	3
Age (years)	67	32	42
Sex	Male	Male	Male
Tumour location	Right	Right	Right
Embolization material	Polyvinyl alcohol particles	Coils	Coils
Time of surgery after TAE	2 Months	21 Days	1 Month
Hemorrhage site	Retroperitoneal	Intratumoral	Retroperitoneal; subcapsular in the right kidney and intratumoral

a bilateral adrenal mass with hemorrhage in the right adrenal mass. Coil embolization was done to the right adrenal artery, and 21 days later, resection of the right adrenal gland was performed⁵⁴.

Case 3 (present case)

A 42-year-old man with a large retroperitoneal hematoma, hemorrhage in the right adrenal mass, and hypertension underwent TAE by coils to the right renal and the adrenal arteries to achieve hemostasis. Surgery was performed 1 month later.

4. DISCUSSION

Pheochromocytomas are uncommon catecholamine-secreting tumours. Rupture of an adrenal pheochromocytoma is extremely rare and can be lethal because of dramatic changes in the circulation.

The exact mechanism of pheochromocytoma rupture is unknown, but high blood pressure attributable to a massive release of catecholamine into the circulation is probably associated with vasoconstriction in the tumour and subsequent necrosis, hemorrhage, and elevated intracapsular pressure. Additionally, rapid tumour growth may contribute to high intracapsular pressure. High pressure in the intracapsular region may also result in a tear in the capsule, causing retroperitoneal hemorrhage.

Abdominal pain may be related to stimulation of alpha-adrenergic receptors, constriction of intestinal vascular smooth muscle, and contraction of the ileocolic sphincter. High levels of circulating catecholamines will therefore result in a decrease of intestinal motility and tone⁵⁵.

The application of TAE allows for hemostasis, with consolidation of the hematoma; surgery can subsequently be performed easily and without complications⁵⁶.

5. CONCLUSIONS

When adrenal pheochromocytoma has been diagnosed, surgical treatment should be considered. Surgical approaches for ruptured pheochromocytoma involve either emergent or elective surgery. Immediate surgery is associated with a high mortality rate (44.7%); no mortality has been reported in patients who undergo elective surgery after good blood pressure and total body water control has been achieved using alpha-adrenergic blocker and fluid infusion therapy, or after stabilization by TAE.

Phentolamine mesylate is a useful alpha-adrenergic blocker to control blood pressure in patients with pheochromocytoma. If initial conservative therapy can control retroperitoneal hemorrhage and blood pressure, a watch-and-wait approach is safe and less invasive for the patient, avoiding the risk of re-bleeding. If, despite medical treatment, the patient is unstable because of uncontrolled blood pressure or recurrent abdominal pain or persistent bleeding, we therefore conclude that TAE should be the first treatment option in the attempt to achieve hemostasis before surgery—an approach that should reduce mortality and maintain the patient in good condition.

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