International Medicine 2019; 1(4): 237-240

International Medicine

https://www.theinternationalmedicine.com/

Case Report

A case of ruptured pheochromocytoma causing loss of life of a patient due to delay in diagnosis

Osman Toktas[1], Abdussamet Batur[2], Recep Eryılmaz[3], Tolga Kalaycı[1], Abdulselam Özdemir[1]

[1] *Yüzüncü Yıl University, Medical Faculty, Department of General Surgery, Van, Turkey*

[2] *Yüzüncü Yıl University, Medical Faculty, Department of Radiology, Van, Turkey*

[3] *Yüzüncü Yıl University, Medical Faculty, Department of Urology, Van, Turkey*

Received: 09 May 2019 / Accepted: 04 June 2019

Abstract

Ruptured pheochromocytoma is an unusual condition, with a mortality rate reaching 31–50%. We describe a case of ruptured pheochromocytoma that was initially mistaken for acute inferior myocardial infarction. On computed tomography, a hemorrhagic mass surrounding the kidney in the region of the right adrenal was observed, but its origin could not clearly be distinguished. On laparotomy, a ruptured and hemorrhagic mass with common necrotic areas was seen, and this was excised successfully. After surgery, the patient experienced a sudden drop in blood pressure. In the postoperative period in the intensive care unit, cardiac arrest occurred. The patient could not be saved. The histological evaluation of the mass showed that it was pheochromocytoma. A surgery combined with early diagnosis and appropriate preoperative treatment may be life-saving. We think that this phenomenon is reminiscent of the adage that an *"acute abdomen is filled with surprises".*

Keywords: acute abdomen, hemorrhage, pheochromocytoma, rupture

Introduction

Pheochromocytoma is an uncommon tumor, it generally originates from chromaffin cells of the adrenal medulla and causes catecholamine secretion. The primary symptoms are persistent or paroxysmal hypertension, tachycardia, diaphoresis, and headache. Rarely, the rupture of the tumoral mass or bleeding inside the mass can lead to the acute abdomen [1]. Because it is a vascular tumor, small hemorrhages and hemorrhagic necrosis are usually found in resected tumors; severe hemorrhage and necrosis, which can occur due to the acute abdominal syndrome are uncommon [2]. A total of 50 cases have been reported worldwide. The death rate from ruptured pheochromocytoma is approximately 31% [3]. Our aim is to present a case who had ruptured pheochromocytoma and was diagnosed as inferior myocardial infarction (MI) in an external emergency room.

Address for Correspondence: Osman Toktas, Yüzüncü Yıl University, Medical Faculty, Department of General Surgery, Van, Turkey.
E-mail: osmantoktas@windowslive.com

DOI: 10.5455/im.46978

Case

A 43-year-old man presented with upper abdominal and anterior chest pain and nausea/vomiting. Systemic examination did not show any abnormalities. His electrocardiogram (ECG) indicated an ST-segment depression in the D2 and D3 derivations. Besides this, elevated CK-MB levels were detected. The patient admitted to the intensive care unit under the suspicion of acute inferior MI and treatment with anticoagulants (300 mg of acetylsalicylic acid and 300 mg clopidogrel) was initiated. He later underwent coronary angiography (CA). The CA did not show any abnormalities. After several hours, due to the worsening of the patient's clinical condition, abdominal ultrasonography (USG) was performed to assess the extra pathology. A hematoma in the right retroperitoneal area, measuring 6 cm in the thickest part, was observed in USG. Computed tomography angiography (CTA) was performed to assess a possible vascular complication after CA. CTA showed no significant vascular pathology, however, the right pararenal area showed a 12 cm hematoma and active bleeding (contrast enhancement), with no clear origin (Figure 1 and 2).

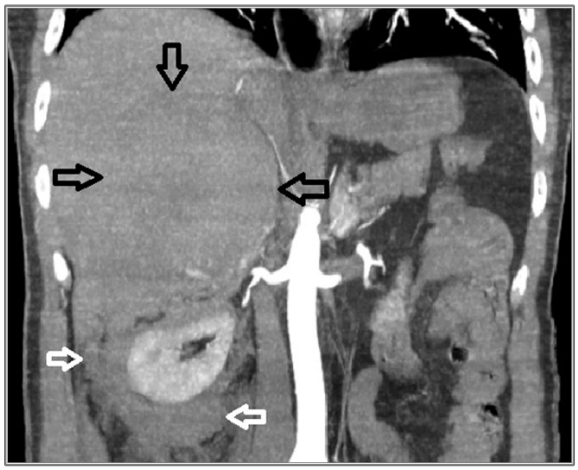

Figure 1. Coronal contrast-enhanced computed tomography (CT) image demonstrating the right adrenal mass and pararenal hematoma.

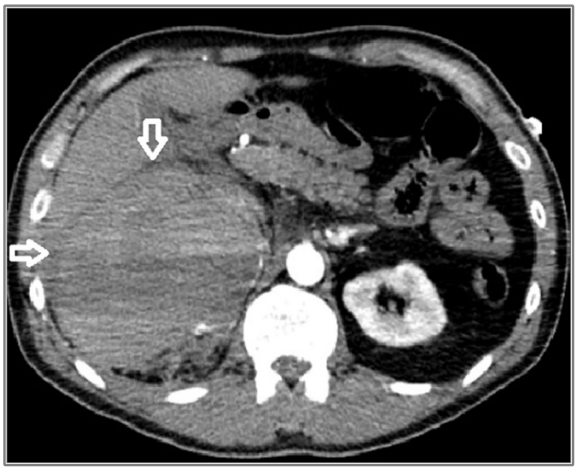

Figure 2. Axial contrast-enhanced computed tomography (CT) image demonstrating the large hematoma localized in the right surrenal gland.

Due to the lack of improvement in the patient's condition, he was referred to our center. When the patient came to the emergency department, his conscious was open, oriented, and cooperated. He had sensitivity on the right side of the abdomen, but there was no tenderness or rebound on physical examination. The patient's blood pressure (BP) was 100/70 mmHg, while his initial pulse rate (PR) was 130 beats/min. The initial biochemical analysis results are shown in Table1. After three hours, the patient's consciousness became hazy and he exhibited restlessness and paleness in the skin. The biochemical analysis showed Hgb: 8.8 g/dL, HCT: 33.8%, BP: 65/40 mmHg, PR: 140 beat/min.

Table 1. Initial biochemical analysis

	Value	Reference range (RR)
Glucose	467 mg/dL	70-105
Creatine	2.6 mg/dL	0.9-1.5
Alanine aminotransferase (ALT)	64 U/L	0-41
Aspartate aminotransferase (AST)	94 U/L	0-37
Amylase	719 U/L	25-125
Creatine kinase (CK)	403 U/L	38-174
CK-myocardial isoenzyme (CK-MB)	143 U/L	0-25
Hemoglobin (Hgb)	10.3 g/dL	11.5-16.5
Hematocrit (HCT)	31.4%	35-55
Platelets (PLT)	221 mm^3	100-400

Emergency surgery was performed to the patient, accompanied by urology department. Intraoperatively, a hematoma was observed; this caused overexpression of the right retroperitoneal region and extended to the right mesocolon. On continued exploration, it was found that the renal artery and renal parenchyma were normal, but a mass lesion was observed in the right adrenal gland that was ruptured, middle necrotic and actively hemorrhagic, with several sites of intact capsules. Because most of the mass was necrotic, only the intact capsule was removed.

Compression of the gauzes in the region and bleeding control were carried out. Intraoperatively, after removal of the mass, the patient underwent cardiac arrest twice. Cardiopulmonary resuscitation with medical support was performed for four minutes the first time and six minutes the second time before the patient's heartbeats returned to normal sinus rhythm. During the operation, three units of fresh frozen plasma and three units of erythrocyte suspension were administered. Bleeding control was performed by removing the gauzes from the region, and the operation was terminated.

When receiving postoperative intensive care, the patient went into cardiac arrest again after 30 minutes. Although all cardiopulmonary resuscitation and medical treatments were performed, the patient could not be rescued. Histopathological examination of the mass revealed that it was pheochromocytoma.

Discussion

Ruptured adrenal pheochromocytoma is exceptionally uncommon and may be fatal [4]. The reasons for and mechanisms of the rupture are unknown. However, a rapid intratumoral hemorrhage can increase the pressure to the capsule and bring about the rupture. After the rupture, a lot of catecholamines are released into circulation and causes a strong headache, abdominal pain, nausea/vomiting, pale skin, and sweating [2]. Changes in the ECG may be due to an increase in circulating catecholamine levels and the possible mechanism for this may be an incompatibility between the needed and available oxygen in the system or another cause of coronary spasm. CA is normal in most of these cases [5,6].

When the patient was admitted to another hospital with upper abdominal pain, nausea, and vomiting complaints, he probably had a hemorrhage in the tumor, but there was no rupture. The intratumoral hemorrhage caused catecholamines to be released in large quantities into the systemic circulation, and this caused changes in the ECG. Because there was ST depression in the ECG, the patient underwent CA, but a pathology was not observed. We estimate that the retroperitoneal necrotic mass ruptured due to the antithrombolytic treatment given to the patient and secondarily the systemic stress that arose from CA.

It is difficult to accurately identify a ruptured pheochromocytoma before surgery in cases like the one described here. Kobayashi et al [3] reported that the ratio of correct preoperative diagnosis is just 30.2%. This difficult diagnosis is due to the low incidence of ruptured pheochromocytoma and catecholamine tests are not performed in both serum and urine under emergency conditions [AQ1]. It is difficult to consider this diagnosis in a hemorrhagic patient with hypotension and tachycardia [2]. We diagnosed retroperitoneal hemorrhage using USG and CT after the worsening of the patient’s clinical condition. After histopathological examination of the removed specimen, the ruptured pheochromocytoma was diagnosed.

The appropriate treatment for ruptured pheochromocytoma is surgical resection via emergency or elective surgery. The most important prognostic factor is careful BP management using α-adrenergic blockade and fluid substitution preoperatively. In the literature, it has been found that emergency surgery without suitable BP checks and an incorrect preoperative diagnosis are strongly associated with a worse prognosis [3]. The death rate for the 54 cases in the literature was 34% [1,7]. Because the diagnosis was not made preoperatively, we could not give the patient α-adrenergic blockage therapy, instead, the patient was only given crystalloid fluid and erythrocyte suspension. This is another reason why our patient died.

In conclusion, a correct preoperative diagnosis is extremely important when it comes to saving the lives of patients with ruptured pheochromocytoma. We have to keep abdominal pathologies in our mind, even if we are suspected an inferior MI. The present case, in which the patient could not be saved, reminds us of the adage that the *“acute abdomen is filled with surprises”.*

Conflict of interest

All authors declare that they have no conflict of interest.

Funding

There was no funding received for this paper.

References

1. Okutur K, Küçükler K, Öztekın E, Borlu F, Erdem L, Demir G. A rare cause of acute abdomen: ruptured adrenal pheochromocytoma. Turk J Gastroenterol 2010;21:467-9.

2. Jee YS. Spontaneous ruptured pheochromocytoma: an unusual case report and literature review. Ann Surg Treat Res 2017;93:170-2.
3. Kobayashi T, Iwai A, Takahashi R, Ide Y, Nishizawa K, Mitsumori K. Spontaneous rupture of adrenal pheochromocytoma: review and analysis of prognostic factors. J Surg Oncol 2005;90:31-5.
4. Oh SH, Oh KJ, Kim KH, Kwon DD, Oh BR, Park YI. Spontaneous rupture of adrenal pheochromocytoma. Chonnam Med J 2002;38:285-7.
5. Chen HY. Ruptured pheochromocytoma presents as acute myocardial infarction. Am J Emerg Med 2009 27;1018.5-6.
6. Menke-van der Houven van Oordt CW, Twickler TB, van Asperdt FG, Ackermans P, Timmers HJ, Hermus AR. Pheochromocytoma mimicking an acute myocardial infarction. Neth Heart J 2007;15:248-51.
7. Takeshita T, Takeshita K, Tagawa Y, Horie S, Furui S. Ruptured pheochromocytoma presenting with acute abdomen and pulmonary edema. Intern Med 2006;45:933-4.