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A rare cause of acute abdomen: Ruptured adrenal pheochromocytoma

Akut batının nadir bir nedeni: Rüptüre adrenal feokromositoma

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Pheochromocytoma is a tumor of the chromaffin cells which secretes catecholamines and 90% of it originates from adrenal medulla. The main symptoms and signs are hypertension, tachycardia, sweating and headache. Rarely, acute abdomen may occur as a result of the rupture of tumoral mass or bleeding inside the mass. Here we present a 43 year old male patient who applied with acute abdominal syndrome and severe hypertension. Abdominal MRI showed a large mass hemorrhagic in nature above the right kidney. At laparotomy, ruptured adrenal mass was seen and excised successfully. The histological evaluation confirmed the diagnosis as pheochromocytoma. Ruptured adrenal pheochromocytoma is a mortal situation potentially and it should be considered in patients who present with an acute obdominal syndrome and hypertension or shock. It should be known that early diagnosis and surgey with proper preoperative treatment is a life saver.

Key words: Pheochromocytoma, rupture, acute abdomen, treatment

Feokromositoma katekolamin sentezleyen kromaffin hücrelerden orjin alan, %90'ı adrenal medulla kaynaklı bir tümördür. Başlıca belirti ve bulguları hipertansiyon, taşikardi, terleme ve başağrısıdır. Nadiren tümör içine kanama veya kitlenin rüptürü sonucu akut batın gelişebilmektedir. 43 yaşında erkek hasta akut batın sendromu ve ciddi hipertansiyon ile başvurdu. Abdomen magnetik rezonans görüntülemesinde sağ böbreğin prosimalinde hemorajik kitle saptandı. Laparotomide sağ adrenal kitle görüldü ve başarıyla eksize edildi. Histolojik incelemede feokromositoma tanısı doğrulandı. Rüptüre adrenal feokromositoma potansiyel olarak mortal bir durumdur ve akut batın sendromuna eşlik eden hipertansiyon veya şok kliniğiyle başvuran hastalarda düşünülmelidir. Erken tanı ve uygun preoperatif tedaviyle beraber uygulanan cerrahinin hayat kurtarıcı olduğu bilinmelidir.

Anahtar kelimeler: Feokromositoma, rüptür, akut batın, tedavi

INTRODUCTION

Pheochromocytoma is a tumor that secretes catecholamines and originates from chromaffin cells of the sympathoadrenal system. In 90% of cases, it is located in the adrenal medulla, but it may also be localized in any region harboring chromaffin cells. Secreted catecholamines are responsible for the clinical signs and symptoms. Hypertension is the initial symptom in more than 50% of cases and it is persistent in 60% of cases; for the remaining, paroxysmal attacks of hypertension are encountered. Orthostatic hypotension may be seen because of the extent of volume depletion.

Pheochromocytoma presented as acute abdominal syndrome is a rare event. Since it is a vascular tumor, small hemorrhages and hemorrhagic necrosis in resected tumors are detected commonly, but severe hemorrhage and necrosis, which can lead to acute abdominal syndrome, are rare (1). Acute abdominal syndrome generally results from rupture of the tumoral mass, or retroperitoneal or intraperitoneal bleeding associated with hemorrhagic necrosis in the tumor without occurrence of rupture.

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Here we present a case of ruptured pheochromocytoma who presented with severe hypertension and acute abdomen.

CASE REPORT

A 43-year-old man admitted to the hospital with sudden abdominal pain, vomiting, sweating, and palpitation. On physical examination, he was observed to be anxious and also had flushing. His body temperature was 38°C, heart rate 130/minute and blood pressure (BP) 230/120 mmHg. In the abdominal examination, a paraumbilical mass was palpated on the right side. Defense and rebound were present on the same side. Total blood count parameters were as follows: white blood cells (WBC): $12.5x10^3/mm^3$, hemoglobin (Hb): 13 g/dl and platelets: $330x10^3/mm^3$. All biochemical parameters were in normal ranges except glucose (270 mg/dl). ECG demonstrated sinus tachycardia. Abdominal ultrasonography revealed a large mass located in the right retroperitoneal region. Abdominal magnetic resonance imaging (MRI) detected a mass in the right suprarenal gland, with a capsule formation with necrotic and hemorrhagic areas and calcification in the peripheral regions, with contrast enhancement and measuring 11x10x6 cm (Figure 1).

Na-nitroprusside and nitroglycerine infusion were initiated and surgical operation was planned. Because no effective control of high BP was maintained with these two agents, phentolamine mesylate, an alpha-blocker agent, was applied, and BP decreased rapidly to within normal range. During the laparotomy, 350 ml hemorrhagic fluid was aspirated from the peritoneal space. Right suprarenal gland was excised without further complications. Immediately after the complete removal of the tumor, the patient's BP rapidly descended to 80/50 mmHg. All hypotensive infusions were stopped abruptly and colloid infusions and ephedrine infusion were given. After BP was stabilized, the operation was completed.

The excised encapsulated hemorrhagic specimen measured 12.5x10x6.5 cm, and contained necrotic and hemorrhagic cystic fields divided by septae (Figure 2). Histopathological investigation confirmed the diagnosis as pheochromocytoma (Figure 3). No metastasis was detected on iodine-^{131}metaiodobenzylguanidine scintigraphy. During the 18-month follow-up period, no signs of recurrence or metastasis were detected.

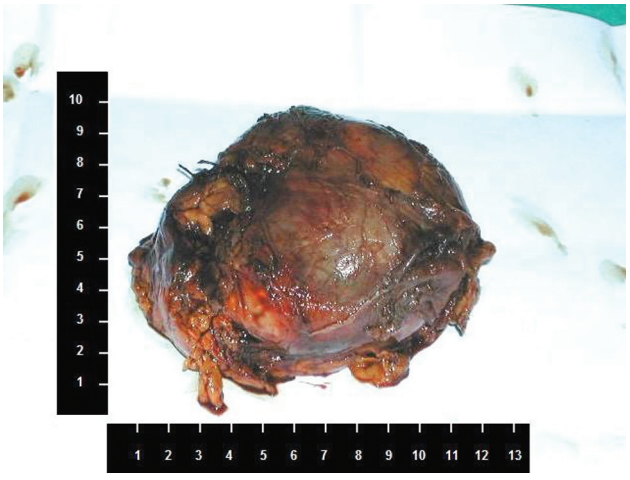

Figure 2. Macroscopic inspection of the resected adrenal pheochromocytoma.

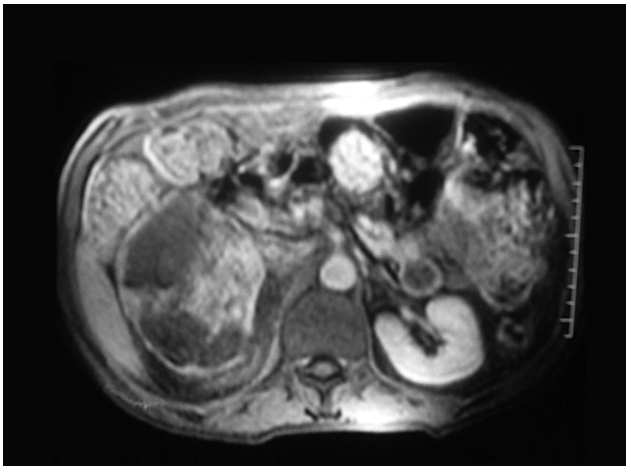

Figure 1. Abdominal MRI (T1-weighted image) reveals a right adrenal mass, hemorrhagic in character, having necrotic areas and septations. The right kidney was pushed inferiorly by the mass.

DISCUSSION

Ruptured pheochromocytoma may progress silently (1), but in some patients shock may also develop associated with massive retroperitoneal bleeding (2). Hypertension, sweating and tachycardia associated with catecholamines secreted into the circulation after the rupture of the tumor may be observed, but when the tumor becomes necrotic, catecholamine secretion decreases gradually. The cardiovascular system is not able to adapt quickly to this rapid decrease in sympathic stimulus (3). Furthermore, intravascular volume is decreased because of hypertension and chronic vasoconstriction and there is a fluid loss to the interstitial space. Consequently, serious hypotension and shock develop rapidly in a previously hypertensive patient. At this stage, the clinical

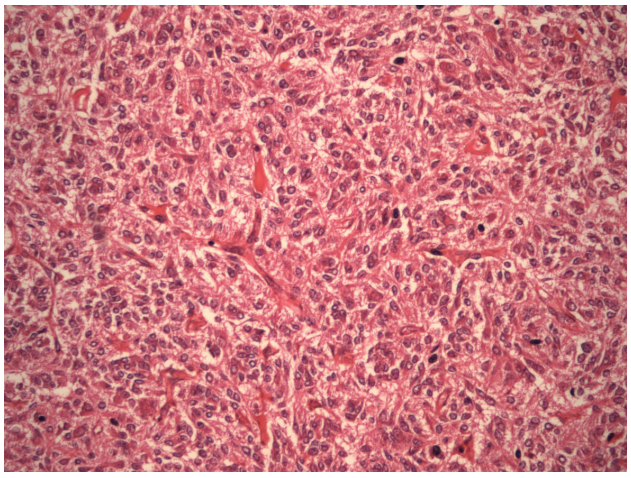

Figure 3. Histological findings of the tumor. Pheochromocytoma cells located around the branching capillaries have round-oval nuclei and finely eosinophilic granular cytoplasm (hematoxylin-eosin, x200).

picture can mimic leaked abdominal aortic aneurysm; computerized axial tomography is the most useful diagnostic method to distinguish between these two entities (4). While these patients do not respond to inotropic treatment, they recover quickly with fluid replacement therapy.

Even though tumor rupture occurs spontaneously, it is reported that rupture and necrosis can appear after alpha-blocker usage (e.g. phentolamine) (3). This situation may be due to the reduction in blood supply after the decrease in high BP due to alpha-blocker treatment and intramural progressive interstitial hemorrhage occurring because of increased blood flow to necrotic areas through vasodilatation inside the tumoral tissue. Another hypothesis is that increased intracapsular pressure as a result of malignant growth predisposes avascular necrosis of the tumoral tissue (5). Furthermore, it is reported that anticoagulants such as warfarin (6) and prochlorperazine (7) may precipitate hemorrhagic necrosis. Occasionally, as a result of capsular invasion, rupture or tearing of the capsule may also appear (8).

For the ruptured pheochromocytoma, the choice of treatment is surgery. The mortality rate was 31% for the 54 cases reported in the literature (9,10). The most important factor affecting the results of the surgery is the resuscitative treatment, which is done at the time of diagnosis and perioperatively (9). Phentolamine is the agent of choice for the treatment of hypertension in this setting. It should be known that a hypotensive stage will follow the excision of the tumor, and this condition should be treated aggressively with fluid replacement and phenylephrine infusion.

In conclusion, ruptured pheochromocytoma should be considered in the differential diagnosis of patients with acute abdominal syndrome concomitantly presenting with uncontrolled hypertension or shock. It should be known that the mortality of the urgent surgery is high in these cases; a proper perioperative approach by a multidisciplinary team consisting of an internist, surgeon, anesthetist, and an intensive care specialist will increase the success of the operation.

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