Hypertensive emergency due to pheochromocytoma crisis complicated with refractory hemodynamic collapse

Refrakter hemodinamik çökmeyle komplike olan feokromasitomaya bağlı hipertansif acil

Mert İlker Hayıroğlu, M.D., Özlem Yıldırımtürk, M.D., Mehmet Bozbay, M.D., Mehmet Eren, M.D., Seçkin Pehlivanoğlu, M.D.

Department of Cardiology, Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul

Summary– Hypertensive emergency usually appears in older patients with previous recurrent episodes, and is among the most frequent admissions to emergency departments. A 29-year-old woman was referred to our clinic with the diagnosis of hypertensive emergency. The patient complained of severe headache, dyspnea, palpitation, diaphoresis, and confusion due to hypertensive encephalopathy. Her blood pressure was 250/150 mmHg on admission. At the referral hospital, the patient had undergone cranial CT because of her confused state and this excluded acute cerebral hemorrhage. Also at that hospital, thoracoabdominal CT for differential diagnosis depicted an adrenal mass with a necrotic core. After admission to our clinic, initial control of excessive blood pressure was not achieved despite high dose intravenous nitrate therapy. Thereafter intravenous esmolol treatment was initiated simultaneously with oral alpha blocker therapy in order to counterbalance the unopposed alpha adrenergic activity with beta blocker therapy. After 12 hours, sudden onset of hypotension developed and deepened despite IV saline, inotropic and vasopressor agents such as IV dopamine, noradrenaline and adrenaline. The patient died at the 24th hour due to hemodynamic collapse as a result of hyperadrenergic state due to possible pheochromocytoma crisis. This case is an exceptional example of hypertensive emergency secondary to fulminant pheochromocytoma crisis failing to respond to intensive antihypertensive treatment, and in which patient death was unavoidable due to uncontrolled excessive adrenergic activity which led to profound cardiogenic shock.

Özet– Hipertansif acil durumlar, acil servise başvuruların en sık sebeplerindendir. Genellikle ileri yaşta ve tekrarlayan şekilde ortaya çıkar. Yirmi dokuz yaşında kadın hasta acil servise hipertansif acil tanısı ile getirildi. Hastanın başvuru sırasında baş ağrısı, nefes darlığı, çarpıntı, terleme ve 250/150 mmHg olan tansiyonu sebebiyle hipertansif ensefalopatiye bağlı konfüzyonu vardı. Hastayı hastanemize sevk eden hastane tarafından çekilen beyin bilgisayarlı tomografisi (BT) akut serebral kanamayı dışlamıştı ve ayırıcı tanı için çekilen torakoabdominal BT'de nekrotik merkezli adrenal kitle saptanmıştı. Kliniğimize kabulünden sonra yüksek doz intravenöz nitrat tedavisine rağmen aşırı olan kan basıncı kontrolü sağlanamadı. Bu nedenle intravenöz esmolol tedavisi yanında eş zamanlı oral alfa bloker tedavisi beta bloker ile ortaya çıkacak alfa adrenerjik aktiviteyi baskılamak için başlandı. On iki saat sonra intravenöz sıvı, dopamin, adrenalin ve noradrenalin gibi inotropik ajanlar verilmesine rağmen ani hipotansiyon gelişti. Muhtemel feokromasitoma krizine bağlı hiperadrenerjik duruma ikincil hemodinamik çökme sebebiyle hasta 24. saatte kaybedildi. Bu olgu ciddi antihipertansif tedaviye cevap vermeyen fulminan feokromasitoma krizine sekonder istisnai bir hipertansif acil olgusudur. Hastanın aşırı adrenerjik aktivitesi derin kardiyojenik şoka yol açmış ve hasta kaybedilmiştir.

Hypertensive emergencies include a spectrum of clinical conditions which present with uncontrolled blood pressure leading to progressive end-organ dysfunction. Approximately 0.02%–0.5% of patients are diagnosed with pheochromocytoma on hypertensive work-up, while 10% of pheochromocytomas are found incidentally.[1]

Abbreviations:

CT	*Computerized tomography*
ECG	*Electrocardiography*
TTE	*Transthoracic echocardiography*

Received: May 02, 2015 *Accepted:* July 06, 2015

Correspondence: Dr. Mert İlker Hayıroğlu. Dr. Siyami Ersek Göğüs ve Kalp Damar Cerrahisi Eğitim ve Araştırma Hastanesi, Kardiyoloji Kliniği, İstanbul.
Tel: +90 216 - 542 44 44 e-mail: mertilkerh@yahoo.com

© 2015 Turkish Society of Cardiology

This case report describes a case of hypertensive emergency with preliminary diagnosis pheochromocytoma crisis. The patient was followed in the coronary care unit for a day and eventually lost due to a profound hypotensive state which was thought to have occurred following sudden adrenal depletion.

CASE REPORT

A 29-year-old woman with no history of chronic disease was admitted to emergency department with severe headache, dyspnea, palpitation, diaphoresis and confusion. On physical examination she was tachypneic, had blurred vision, disturbed orientation and an unstable condition. Her blood pressure was 250/150 mmHg, oxygen saturation 82% with pulse oximetry, and temperature 37.9 ºC. Auscultation of the patient revealed loud S1 and S2, S3-S4 gallop rhythm, and bilateral diffuse rales were heard on pulmonary auscultation. Electrocardiography (ECG) revealed sinus tachycardia. Heart rate was 170 beat/min, without any significant ST segment changes.

Before admission to our clinic cranial computerized tomography (CT) and thoracoabdominal CT were performed at the referral hospital, in order to reveal the underlying etiology of disturbed consciousness and unstable clinical condition. There was no acute pathology in the cranial CT. However, in the thoracoabdominal CT, a mass of dimensions 4.5x4 cm with a necrotic core and compatible with malignancy was detected in the left adrenal gland region.

The patient was immediately transferred to intensive care, where she was intubated due to acute respiratory failure secondary to acute pulmonary edema depicted with worsening of arterial blood gas analysis and disturbed consciousness. Bedside transthoracic echocardiography (TTE) revealed severe depressed biventricular dysfunction with global hypokinesia (left ventricular ejection fraction 30–35%). Both chambers were of normal dimensions and there was no accompanying valvular heart disease. The patient's cardiac troponin I value was over the normal limits, but coronary angiography was not performed since this elevation was thought to be related to catecholamine-induced myonecrosis.

Her preliminary diagnosis was hypertensive emergency due to pheochromocytoma crisis resulting in catecholamine-induced cardiomyopathy and hypertensive encephalopathy. IV saline, IV nitrate and IV esmolol infusion was started with alpha blocker (doxazosin) in order to assuage the state. Her blood pressure and heart rhythm were significantly above the normal limits for several hours. After twelve hours, severe hypotension suddenly developed, and thus all antihypertensive treatment was discontinued. Transthoracic echocardiography was repeated, but no cardiac complication was detected. The diameter of the inferior vena cava was 2.1 cm, invasive hemodynamic measurements were not used. The hypotension did not recover despite administration of high dose IV inotropic and vasopressor agents such as dopamine, noradrenaline and adrenaline along with IV saline infusion. As a result patient died because of hemodynamic collapse due to adrenergic crisis.

DISCUSSION

One of the most common systemic diseases, hypertension is due to secondary causes in 4–5% of patients. Of hypertension cases, 0.1–0.2% are caused by pheochromocytomas, or catecholamine-producing tumors derived from chromaffin tissue. Secondary hypertension should be particularly considered in younger patients (<30 years), in whom clinically-resistant and sudden onset is also remarkable. Sustained or paroxysmal hypertension is the cardinal feature of pheochromocytoma. The classical triad of headache, palpitations and diaphoresis provide strong clues for this diagnosis.[2]

The late course of pheochromocytoma may be devastating, as was the case with this patient. These patients may be presenting with severe labile hypertension and accompanying chest pain, recent onset cardiomyopathy, pulmonary edema, fever and metabolic acidosis.[3] Chest pain may also be seen due to severe vasospasm, and result in cardiomyopathy related to both vasospasm-induced ischemia and inflammatory infiltration, and should not be confused with coronary artery disease.[4,5] However, adrenergic crisis may also be cause to acute myocardial infarction, even Takatsubo cardiomyopathy.[6–9] In our case, troponin level was high, but TTE did not show segmental motion dysfunction. ECG showed no ST segment changes, thus excluding acute coronary syndrome due to coronary artery disease. Echocardiographic features were consistent with catecholamine-induced cardiomyopathy.

While the ultimate treatment of pheochromocytoma is surgical resection of the tumor, pharmacological therapy still remains of vital significance in pre-operative and operative control of blood pressure. Treatment should be undertaken in intensive care with intravenous short half life IV drugs such as labetolol, esmolol, fenoldopam, nicardipine and sodium nitroprusside. In adrenergic hypertensive crisis, differently from other hypertensive crises, alpha blockers must be used.[10] IV phentolamine (alpha-adrenergic antagonist) is the preferred agent.[11] In our case, the lack of labetolol and other drugs meant the use of doxazosin , IV esmolol and IV nitrate.

In our case, profound hypotension developed suddenly, and was unresponsive to high dose inotropic and vasopressor agents. There are some case reports in which intra aortic balloon pumping and percutaneous cardiopulmonary bypass systems were successfully used in the treatment of medically unresponsive patients. Catastrophic shock can be completely reversible without leading to further damage after recovery from the acute phase.[12,13] The sudden onset of deep hypotension continued in asystole in our case, and therefore we were unable to use mechanical support devices. It is significant that the hemodynamic change in pheochromocytoma patients is so rapid, and thus invasive hemodynamic measurements (cardiac output, pulmonary wedge pressure) should be kept in mind and properly used. The explanation for this situation was thought to be secondary to adrenal hormonal depletion or down-regulation of adrenal hormonal receptors.

This case is an important example of pheochromocytoma crisis with devastating consequences. Early recognition and treatment is critical for management of these patients. While planning surgery, pharmacological treatment should include termination of catecholamine effects and expansion of plasma volume.

Conflict-of-interest issues regarding the authorship or article: None declared.

REFERENCES

1. Beard CM, Sheps SG, Kurland LT, Carney JA, Lie JT. Occurrence of pheochromocytoma in Rochester, Minnesota, 1950 through 1979. Mayo Clin Proc 1983;58:802–4.
2. Werbel SS, Ober KP. Pheochromocytoma. Update on diagnosis, localization, and management. Med Clin North Am 1995;79:131–53.
3. Kizer JR, Koniaris LS, Edelman JD, St John Sutton MG. Pheochromocytoma crisis, cardiomyopathy, and hemodynamic collapse. Chest 2000;118:1221–3. CrossRef
4. Szakacs JE, Cannon A. L-Norepinephrine myocarditis. Am J Clin Pathol 1958;30:425–34.
5. Van Vliet PD, Burchell HB, Titus JL. Focal myocarditis associated with pheochromocytoma. N Engl J Med 1966;274(:1102–8.
6. Ortiz-Vázquez IC, Ramos-García MA, Maza-Juárez G, Clavellina-Rosas JM, Moreno-Vázquez A, Calderón-Abbo M. Acute myocardial infarction due to pheochromocytoma. [Article in Spanish] Rev Med Inst Mex Seguro Soc 2012;50:559–63. [Abstract]
7. Boulkina LS, Newton CA, Drake AJ 3rd, Tanenberg RJ. Acute myocardial infarction attributable to adrenergic crises in a patient with pheochromocytoma and neurofibromatosis 1. Endocr Pract 2007;13:269–73. CrossRef
8. Radtke WE, Kazmier FJ, Rutherford BD, Sheps SG. Cardiovascular complications of pheochromocytoma crisis. Am J Cardiol 1975;35:701–5. CrossRef
9. Celebi H, Erdim R, Karabay KO, Yildirimturk O, Aytekin V. Recurrent left ventricular apical ballooning syndrome in a patient with pheochromocytoma. Int J Angiol 2012;21:63–8.
10. Knapp HR, Fitzgerald GA. Hypertensive crisis in prazosin-treated pheochromocytoma. South Med J 1984;77:535–6.
11. Varon J. Treatment of acute severe hypertension: current and newer agents. Drugs 2008;68:283–97. CrossRef
12. Khan MH, Corbett BJ1, Hollenberg SM1. Mechanical circulatory support in acute cardiogenic shock. F1000Prime Rep 2014;6:91. CrossRef
13. Zhou X, Liu D, Su L, Long Y, Du W, Miao Q, et al. Pheochromocytoma crisis with severe cyclic blood pressure fluctuations in a cardiac pheochromocytoma patient successfully resuscitated by extracorporeal membrane oxygenation: a case report. Medicine (Baltimore) 2015;94:790. CrossRef

Keywords: Hypertension; hypertensive emergency; pheochromocytoma/complications.

Anahtar sözcükler: Hipertansiyon; hipertansif acil; feokromasitoma/komplikasyonlar.