

# Catecholamine-induced cardiomyopathy and paraganglioma in a pediatric patient

*Bir çocuk hastada katekolamine bağlı kardiomyopati ve paraganglionöroma*

**Nazmi Narin, Ali Baykan, Sadettin Sezer, Sertaç Hanedan Onan, Kazım Üzüm, Mustafa Küçükaydın\***  
From Departments of Child Health and Disease, Pediatric Cardiology and \*Pediatric Surgery, Faculty of Medicine, Erciyes University, Kayseri-Turkey

## Introduction

Catecholamine-induced cardiomyopathy is characterized by left ventricle apex and mid ventricle wall-motion abnormalities in the absence of coronary artery stenosis. Catecholamine-induced cardiomyopathy is very rare in children. Pheochromocytoma is one of the most important causes of this contractile pattern.

Herein we report the case of catecholamine-induced cardiomyopathy in a child with paraganglioma.

## Case Report

A 16-year-old boy with history of bicuspid aortic valve and mild aortic regurgitation was admitted to emergency department with abdominal pain, nausea, dizziness, sweating, and lightheadedness after football playing. Physical examination at the time of admission showed alert consciousness, with body temperature of 36°C, pale skin and sweating, blood pressure of 80/60 mmHg, pulse 160/min. There were fine crackles, and I/VI systolic murmur on lower left sternal border in auscultation. The electrocardiogram revealed ST-T changes and sinus tachycardia (160/min) with long QT (0.50sc) (Fig. 1, 2). Cardiomegaly and pulmonary congestion were detected on chest X-ray. Echocardiographic examination revealed dilated left ventricle (LV) (LV diastolic dimension 5.66cm, LV systolic dimension 4.83 cm) with reduced ejection fraction (38%), mild mitral regurgitation and aortic regurgitation. The LV showed diffuse, severe hypokinesia with slight dominance in its lateral and posterior wall (Fig. 3).

The white blood cell count was 24780/ml and the serum analyses revealed: GOT 35 IU/L, GPT 69 IU/L, CK 644 IU/L, CK-MB 121 IU/L and blood glucose 52 mg/dl. Troponin I (3.11 ng/ml) was elevated. Despite saline hydration and continuous infusion of inotropic agents, his blood pressure was unstable. While we were treating hypotension and shock the clinical presentation changed and we detected hypertension and



Figure 1. ST-T changes during acute attack

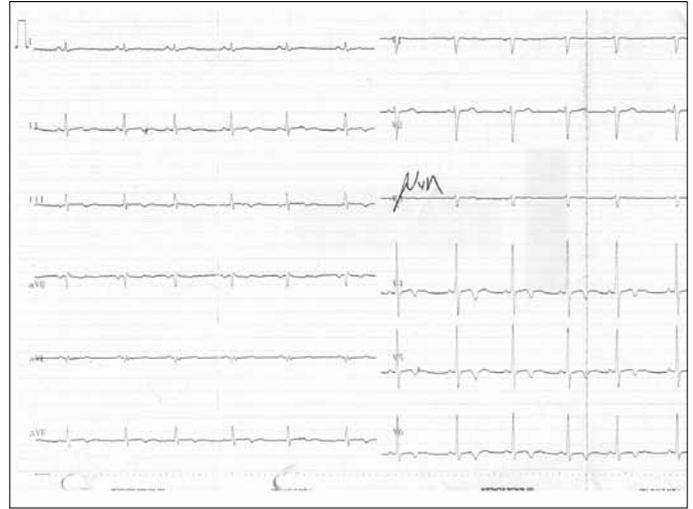


Figure 2. Prolonged QT (0.50 sc)

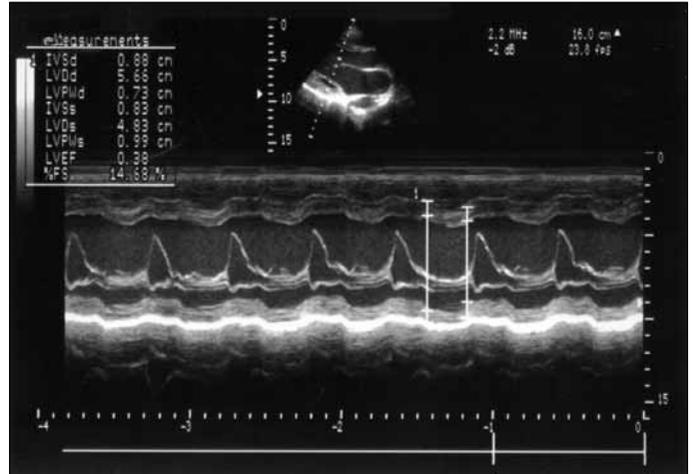


Figure 3. Dilated left ventricle and paradoxical LV movement on M-mode echocardiography

LV - left ventricle

long QT, we started metoprolol. Then blood pressure and QT distance measured at normal ranges (QT: 0.42 sc).

On day 3 at hospital, because of the episodic headache, severe abdominal pain attack, especially in left abdominal region, palpitation and fluctuating blood pressure, pheochromocytoma was suspected and relevant investigations performed: Abdominal ultrasonography revealed a solid mass on left side. Magnetic resonance imaging scan of the abdomen revealed left abdominal mass,  $\alpha$ -adrenergic blocker; prazosin was started with the presumptive diagnosis of pheochromocytoma. Urinary catecholamine levels were: metanephrine 151  $\mu$ g/day (normal range <320  $\mu$ g/day), and normetanephrine 854  $\mu$ g/day (normal range 390  $\mu$ g/day).

Pathological findings of excised mass were compatible with pheochromocytoma (paraganglioma with hemorrhagic necrosis) (Fig. 4). Histological and immunohistochemical analyses confirmed the initial diagnosis. From these findings, catecholamine-induced cardiomyopathy with acute heart failure due to pheochromocytoma was final diagnosis.

The postoperative recovery was quick and uneventful. The postoperative urinary catecholamine levels were as follows: metanephrine 279  $\mu$ g/24 day, normetanephrine 131  $\mu$ g/day. Echocardiography repeated two weeks after the surgery revealed improvement in left ventricular ejection fraction of 64%. The patient remains asymptomatic at 12<sup>th</sup> months of the operation.

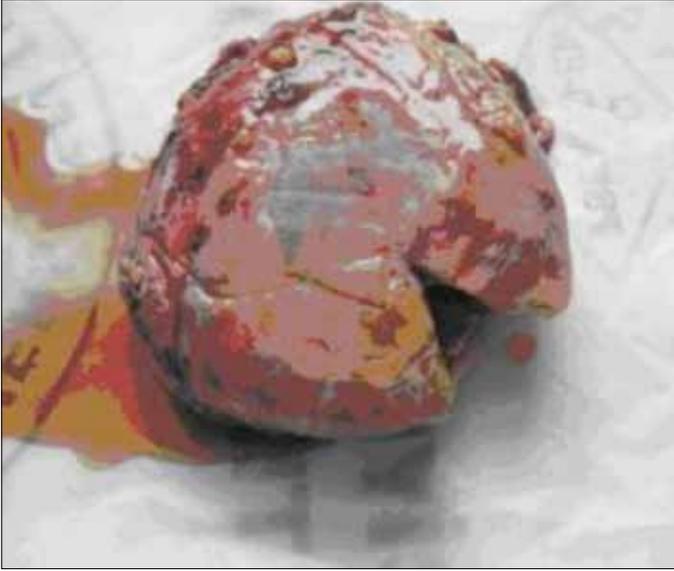


Figure 4. Excised mass: paraganglioma with hemorrhagic necrosis

### Discussion

Paraganglioma or extra-adrenal pheochromocytoma is rare neuroendocrine tumor of the autonomic nervous system. Paraganglioma often present with signs of catecholamine excess. The most secreted catecholamine is norepinephrine, and classical symptomatologic triad due to catecholamine excess is headache, sweating and palpitations (1, 2). But sometimes patients may be asymptomatic or symptoms may be vague: psychiatric disorders, anxiety, facial pallor, weight loss, polyuria/polydipsia, hyperglycemia, secondary erythrocytosis, stroke, and cardiomyopathy (1). Hypertension, cardiogenic shock, pulmonary congestion and long QT were present in our case.

When we investigated the etiology of symptoms in our patient, paraganglioma was detected. High circulating catecholamines have been found during episodes of catecholamine cardiomyopathy (3, 4). Catecholamine levels of our patient were also high.

The cause of left ventricular dysfunction in catecholamine cardiomyopathy is still controversial. In catecholamine cardiomyopathy, physical stress is recognized as an important predisposing factor (5). The suspected pathophysiology for catecholamine cardiomyopathy are coronary vasospasm, increased vascular resistance, tachycardia or direct catecholamine-mediated myocyte injury, which results in wall motion abnormalities and biventricular dilation (6, 7).

Acute catecholamine cardiomyopathy of two cases related with pheochromocytoma was presented in the literature and cardiomyopathy resolved by the resection of adrenal tumor in these cases (4, 6, 8).

In our case firstly patient was in shock than improved immediately and clinical signs changed to catecholamine excess. After detection and resection of paraganglioma patient improved completely as conjunction with literature.

### Conclusion

Cardiogenic shock caused by catecholamine crisis is less common recognized manifestation of catecholamine secreting tumors. To the best of our knowledge, this is a rare case presented as cardiogenic shock caused by atypical transient LV dysfunction related with catecholamine excess which has been successfully treated by early diagnosis and treatment. Improvement or reversal of catecholamine cardiomyopathy weeks after surgical resection is an important phenomenon.

### References

1. Yeo H, Roman S. Pheochromocytoma and functional paraganglioma. *Curr Opin Oncol* 2005; 17: 13-8. [CrossRef]
2. Prys-Roberts C. Pheochromocytoma: recent progress in its management. *Br J Anaesth* 2000; 85: 44-57. [CrossRef]
3. Gianni M, Dentali F, Grandi AM, Sumner G, Hiralal R, Lonn E. Apical ballooning syndrome or Takotsubo cardiomyopathy: a systematic review. *Eur Heart J* 2006; 27: 1523-9. [CrossRef]
4. Sanchez-Recalde A, Costero O, Oliver JM, Iborra C, Ruiz E, Sobrino JA. Images in cardiovascular medicine. Pheochromocytoma-related cardiomyopathy: inverted Takotsubo contractile pattern. *Circulation* 2006; 113: 738-9. [CrossRef]
5. Bybee KA, Kara T, Prasad A, Lerman A, Barsness GW, Wright RS, et al. Systemic review: transient left ventricular apical ballooning: a syndrome that mimics ST-segment elevation myocardial infarction. *Ann Intern Med* 2004; 141: 858-65.
6. Mann D, Kent RL, Parsons B, Cooper G 4th. Adrenergic effects on the biology of the adult mammalian cardiocyte. *Circulation* 1992; 85: 790-804.
7. Yoshinaga K, Torii H, Tahara M. A serial echocardiographic observation of acute heart injury associated with pheochromocytoma crisis. *Int J Cardiol* 1998; 66: 199-202. [CrossRef]
8. Poyraz P, Açıl M, Arıboğan A. Takotsubo sendromu olarak değerlendirilen bir hastada saptanan feokromositoma. *Türkiye Klinikleri J Anest Reanim* 2009; 7: 44- 8.

**Address for Correspondence/Yazışma Adresi:** Dr. Nazmi Narin  
Erciyes Üniversitesi Tıp Fakültesi, Çocuk Sağlığı ve Hastalıkları Anabilim Dalı,  
Çocuk Kardiyolojisi Bilim Dalı, Kayseri-*Türkiye*  
Phone: +90 352 437 49 37 E-mail: nnarin@erciyes.edu.tr  
**Available Online Date/Çevrimiçi Yayın Tarihi:** 03.12.2011

©Telif Hakkı 2011 AVES Yayıncılık Ltd. Şti. - Makale metnine [www.anakarder.com](http://www.anakarder.com) web sayfasından ulaşılabilir.

©Copyright 2011 by AVES Yayıncılık Ltd. - Available on-line at [www.anakarder.com](http://www.anakarder.com)  
doi:10.5152/akd.2011.200