Extra-adrenal Pheochromocytoma Presenting with Life-threatening Ventricular Tachycardia: A Case Report

Wei-Ming Li, Chun-Hsiung Huang, Chin-Ming Su, Chee-Yin Chai,[1]
Wen-Jeng Wu, and Yii-Her Chou
Departments of Urology and [1]Pathology, Kaohsiung Medical University Hospital, Kaohsiung, Taiwan.

Extra-adrenal pheochromocytoma develops in paraganglion chromaffin cells of the sympathetic nervous system. It probably represents at least 15% of adult and 30% of childhood pheochromocytomas. Although electrocardiographic abnormalities occur in up to 75% of patients with pheochromocytoma, there are only three other reported cases of pheochromocytoma that initially presented with ventricular tachycardia. In this report, we describe a 64-year-old woman with an abdominal para-aortic extra-adrenal pheochromocytoma who presented with pulseless ventricular tachycardia and needed electrical defibrillation initially to keep vital signs. The 24-hour urine vanillylmandelic acid was 35.8 mg. Computerized tomography showed a left retroperitoneal mass. The postoperative course was uneventful. Pheochromocytoma was proved by histologic study.

Key Words: pheochromocytoma, ventricular tachycardia, arrhythmia
(*Kaohsiung J Med Sci* 2004;20:612–5)

Extra-adrenal pheochromocytoma develops in paraganglion chromaffin cells of the sympathetic nervous system. It probably represents at least 15% of adult and 30% of childhood pheochromocytomas [1]. It most commonly occurs in the second or third decade of life [2]. Patients with extra-adrenal pheochromocytoma commonly present with the classic triad of headache, palpitation, and sweating [2]. Cardiac arrhythmias are common with pheochromocytoma. Nevertheless, ventricular tachycardia has been reported in only three cases [3]. We report a rare case of pulseless ventricular tachycardia caused by extra-adrenal pheochromocytoma in an adult.

Received: June 17, 2004 Accepted: September 15, 2004
Address correspondence and reprint requests to: Dr. Chun-Hsiung Huang, Department of Urology, Kaohsiung Medical University, 100 Shih-Chuan 1st Road, Kaohsiung 807, Taiwan.
E-mail: urology@kmu.edu.tw

Case Presentation

A 64-year-old female had had hypertension under drug control (amlodipine 2.5 mg/day and bisoprolol 5 mg/day) for 2 years. She was sent to our emergency department due to cold sweating, headache, palpitation, and hypertension (245/119 mmHg). She lost consciousness and had pulseless ventricular tachycardia (Figure 1) after arrival. After electrical defibrillation (200 J), she recovered consciousness. Electrocardiography (ECG) showed sinus tachycardia and inverted T waves. Blood pressure was 237/160 mmHg. Due to unstable hemodynamics, she was admitted and received advanced survey.

A 24-hour Holter monitor revealed sinus rhythm with intermittent junctional escape rhythms. Heart ultrasonography showed mild tricuspid regurgitation, right ventricular dilation, and septal hypertrophy. The patient also underwent coronary angiographic evaluation, which was normal. The 24-hour urine vanillylmandelic acid was 35.8 mg (normal, 2–9 mg/24 hours). Other laboratory studies revealed: normal thyroid function; serum ionized calcium,

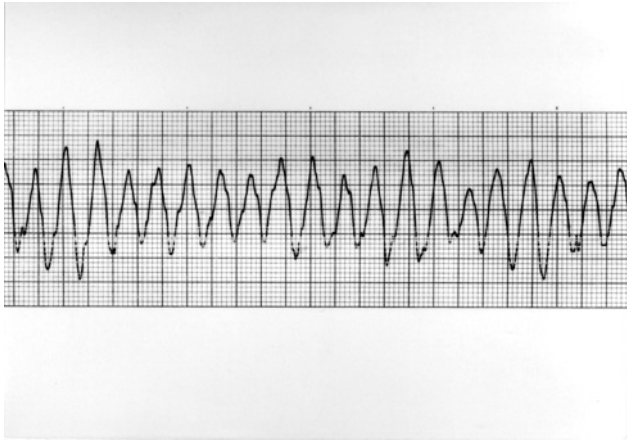

Figure 1. *Ventricular tachycardia on electrocardiography.*

4.9 mg/dL; normal serum electrolyte; blood urea nitrogen, 27 mg/dL; and creatinine, 1.2 mg/dL. Computerized tomography showed a left retroperitoneal mass which compressed the left ureter, resulting in mild obstructive uropathy (Figure 2). ^{131}I-metaiodobenzylguanidine scan revealed abnormal tracer uptake in the left abdomen. After admission, blood pressure and arrhythmia were well controlled with labetalol, a non-selective α- and β-adrenergic blocker. Preoperatively, the patient was taking 200 mg labetalol three times daily, titrated to 200 mg twice daily according to blood pressure when surgery was arranged.

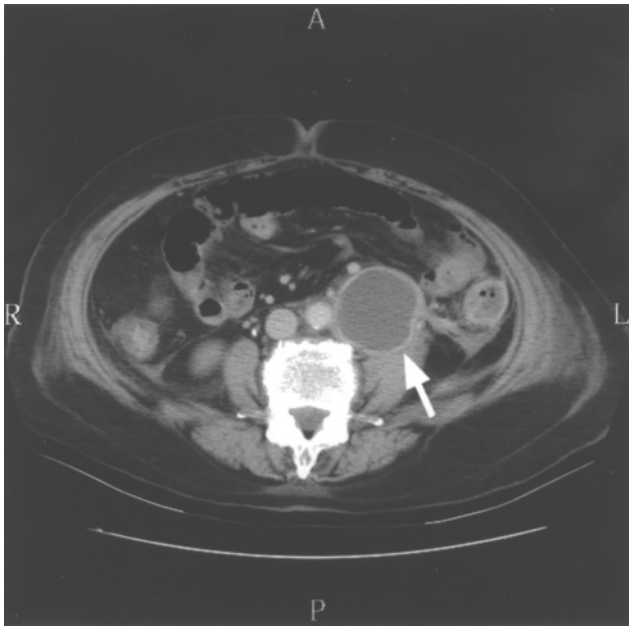

Figure 2. *A left retroperitoneal mass (arrow) on computerized tomography.*

The tumor was totally excised through a para-midline transperitoneal incision. An operative ECG-blood pressure monitor taped several hypertensive and hypotensive episodes without ventricular rhythm disturbance. Histopathologic examination of the tumor confirmed pheochromocytoma (weight, 32 g; 62 × 39 × 28 mm) (Figure 3). Postoperatively, blood pressure remained stable and within the normal range. She was discharged on postoperative day 14.

Discussion

Electrocardiographic abnormalities are common and occur in up to 75% of patients with pheochromocytoma, including sinus tachycardia, premature ventricular complex, T-wave inversion, premature atrial complex, ST elevation, paroxysmal supraventricular tachycardia, atrial fibrillation, prolonged QT interval, painless myocardial infarction, and left ventricular hypertrophy [3]. However, pheochromocytoma that initially presents with ventricular tachycardia is rare and only three cases have been reported in the literature [4]. We report another case of extra-adrenal pheochromocytoma that presented with pulseless ventricular tachycardia and needed electrical defibrillation.

Cardiac arrhythmias are probably due to the sudden release of catecholamines. Ceremuzynski et al found increased plasma catecholamine concentrations in anesthetized dogs during acute myocardial ischemia and a relationship between blood catecholamine concentration and the severity of ventricular arrhythmias [5]. Daugherty et al showed that increases in plasma catecholamine concentrations during coronary artery ligation in pentobarbitone-anesthetized rats were prevented by either acute adrenalectomy or chronic adrenal demedullation, but these procedures did not protect against ventricular arrhythmias [6]. Thus, plasma catecholamines are not obligatory mediators of arrhythmogenesis, but these results do not exclude the possibility that catecholamines released locally to the ischemic myocardium may be important arrhythmogenic mediators.

Other neurohumoral agents may be associated with cardiac arrhythmia. Neuropeptide Y levels are increased in the plasma and tumors of patients with pheochromocytoma. Neuropeptide Y has potent direct and indirect cardiovascular effects [7]. The presence and degree of circulatory disturbance, in particular tachycardia and left heart failure, are strongly related to increased plasma concentrations of neuropeptide Y in coronary care patients [8].

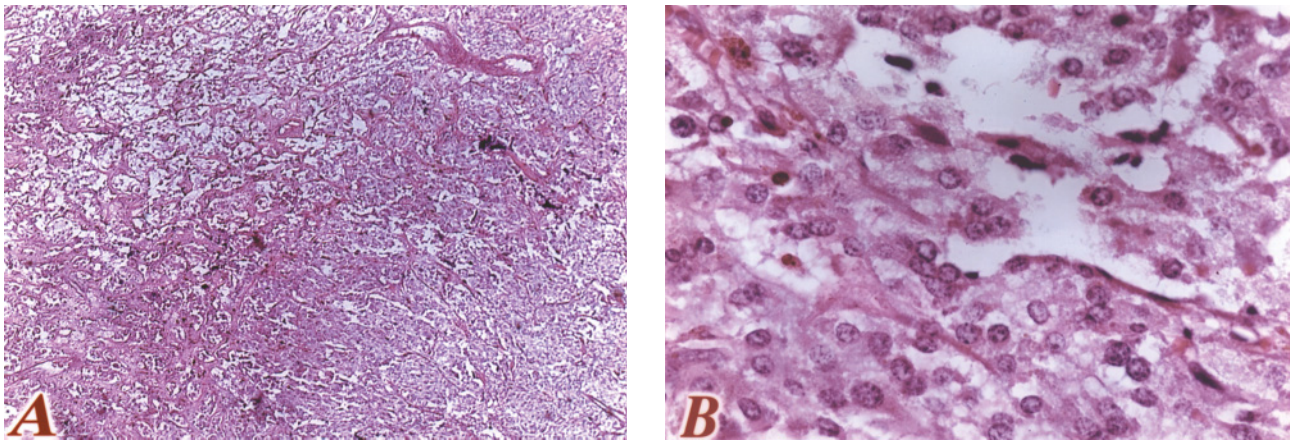

Figure 3. *Microscopic findings of the retroperitoneal mass: (A) tumor cells are arranged in a nest bounded by delicate fibrovascular stroma, the so-called Zellballen (hematoxylin & eosin, × 40); (B) the nuclei are round or oval with finely granular cytoplasm, and there is hyperchromasia with occasional nucleoli (hematoxylin & eosin, × 400).*

In addition, cardiac abnormalities occur commonly in patients with pheochromocytomas, including myocarditis, acute myocardial infarction, cardiac arrhythmias, and cardiomyopathy. Catecholamines released from pheochromocytomas cause myocardial necrosis, focal myofibrillar degeneration, and subsequent fibrous scar formation [9,10].

The preoperative use of phenoxybenzamine, a nonselective α-adrenergic blocker, has a mostly theoretical pharmacologic basis. The β-blocker propranolol is used when tachycardia or catecholamine-induced arrhythmias are present, and should only be started after α-blockade has been established to avoid life-threatening hypertension. Labetalol has the advantage of being a β-blocker with additional α-adrenergic blocking properties. However, it may interfere with diagnostic studies, restricting its use to patients in whom the diagnosis of pheochromocytoma is already firm [1,7]. The optimal length of preoperative adrenergic blockade is variable and primarily depends on the time required for the patient to manifest hemodynamic stability. In our case, the patient was placed on labetalol 2 weeks preoperatively to control hypertension and tachycardia.

Pheochromocytoma should be kept in mind even in an elderly hypertensive patient. In our opinion, while the diagnosis of extra-adrenal pheochromocytoma is established by biochemical examination and image studies, it is best to use continuous ECG monitoring. Although life-threatening cardiac arrhythmias are rare in extra-adrenal pheochromocytoma, cardiopulmonary resuscitative facilities and medications must be easily available to prevent morbidity and cardiac arrest.

References

1. Whalen RK, Althausen AF, Daniels GH. Extra-adrenal pheochromocytoma. *J Urol* 1992;147:1–10.
2. Goldfarb DA, Novick AC, Bravo EL, et al. Experience with extra-adrenal pheochromocytoma. *J Urol* 1989;142:931–6.
3. Michaels RD, Hays JH, O'Brian JT, et al. Pheochromocytoma associated ventricular tachycardia blocked with atenolol. *J Endocrinol Invest* 1990;13:943–7.
4. Petit T, de Lagausie P, Maintenant J, et al. Thoracic pheochromocytoma revealed by ventricular tachycardia. *Eur J Pediatr Surg* 2000;10:142–4.
5. Ceremuzynski L, Staszewska-Barczak J, Herbaczynska-Cedro K. Cardiac rhythm disturbances and the release of catecholamines after acute coronary occlusion in dogs. *Cardiovasc Res* 1969;3:190–7.
6. Daugherty A, Frayn KN, Redfern WS, et al. The role of catecholamines in the production of ischaemia-induced ventricular arrhythmias in the rat in vivo and in vitro. *Br J Pharmacol* 1986;87:265–77.
7. Bravo EL, Tagle R. Pheochromocytoma: state-of-the-art and future prospects. *Endocr Rev* 2003;24:539–53.
8. Hulting J, Sollevi A, Ullman B, et al. Plasma neuropeptide Y on admission to a coronary care unit: raised levels in patients with left heart failure. *Cardiovasc Res* 1990;24:102–8.
9. Van Vliet PD, Burchell HB, Titus JL. Focal myocarditis associated with pheochromocytoma. *N Engl J Med* 1966;274:1102–8.
10. Bravo EL. Pheochromocytoma: new concepts and future trends. *Kidney Int* 1991;40:544–56.

腎上腺外嗜鉻細胞瘤以致命性心室性心搏過速表現 — 病例報告

李威明[1] 黃俊雄[1] 蘇進明[1] 蔡志仁[2] 吳文正[1] 周以和[1]

高雄醫學大學 [1]泌尿科 [2]病理科

腎上腺外嗜鉻性細胞瘤是從交感神經系統的副神經節嗜鉻細胞生長出來的。腎上腺外嗜鉻細胞瘤佔成人嗜鉻細胞瘤的百分之十五；佔小孩的百分之三十。雖然有超過百分之七十五嗜鉻性細胞瘤的病人會出現心電圖上的異常，但一開始即以心室性心搏過速表現並不常見，文獻中只有三個病例。本文中，我們報告一位六十四歲女性罹患腹腔內腎上腺外嗜鉻性細胞瘤的病例，該病人表現出無脈搏性心室性心搏過速，且需心臟電擊以維持生命徵象。二十四小時尿中 vanillylmandelic acid 是 35.8 毫克。電腦斷層發現左側後腹腔有一軟組織腫塊。術後病人恢復狀況良好。病理報告證實為嗜鉻性細胞瘤。

關鍵詞：嗜鉻性細胞瘤，心室性心搏過速，心律不整

（高雄醫誌 2004;20:612–5）

收文日期：93 年 6 月 17 日

接受刊載：93 年 9 月 15 日

通訊作者：黃俊雄醫師

高雄醫學大學泌尿科

高雄市三民區十全一路 100 號