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A Case Report of Retroperitoneal Mass: Paraganglioma

Chin-Chou Yang

Division of General Medicine, Department of General Psychiatry, Tsaotun Psychiatric Center, Ministry of Health and Welfare, Nantou, Taiwan

Abstract

Pheochromocytoma and paraganglioma are rarely seen cathecolamine-secreting neuroendocrine tumors. We reported a retroperitoneal tumor with a diameter of 5 cm in a 71 years old man. After admission, he was found loss of consciousness suddenly. Myocarditis related to pheochromocytoma was the most possible reason of sudden collapse. The diagnosis of paraganglioma was made later in ICU (intensive care unit). Pheochromocytoma or paraganglioma may present with life-threatening cardiovascular manifestations, such as acute myocarditis, myocardial infarction, and hypertensive emergency. It should never been neglected in the differential diagnosis of retroperitoneal mass lesions. (J Intern Med Taiwan 2018; 29: 169-174)

Key Words: Paraganglioma, Pheochromocytoma, Adrenal tumor

Present Illness

A 71 years old man presented with acute palpitation, abdominal pain, headache and dizziness in ER (emergency room). He had history of hypertension for 4 years, and his condition remained stable while taking daily combination therapy with Amlodipine, Irbesartan, and Propranolol. At ER, his body temperature was 36.4°C, heart rate was 96 beats per minute, and blood pressure was 87/47mmHg. The ECG (Electrocardiogram) revealed normal sinus rhythm. The chest X-ray revealed negative findings except atherosclerosis and tortuosity of thoracic aorta. Abdominal CT was performed in ER. It revealed a 5cm retroperitoneal mass, considered suspicious of a pancreatic tumor (Figure 1). He was then admitted on Aug 20, 2014. On Aug 22 early morning, he was found loss of consciousness sud-

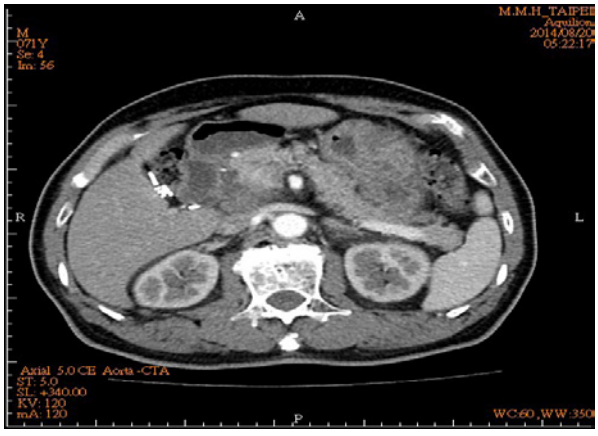

Figure 1. A 5cm heterogeneous soft tissue mass occupied at right para-aortic region with anterior displacement of duodenum and pancreas head.

Reprint requests and correspondence：Dr. Chin-Chou Yang
Address：Division of General Medicine, Department of General Psychiatry, Tsaotun Psychiatric Center, Ministry of Health and Welfare, Nantou, Taiwan, No. 161, Yuping Rd., Caotun Township, Nantou County 542, Taiwan (R.O.C.)

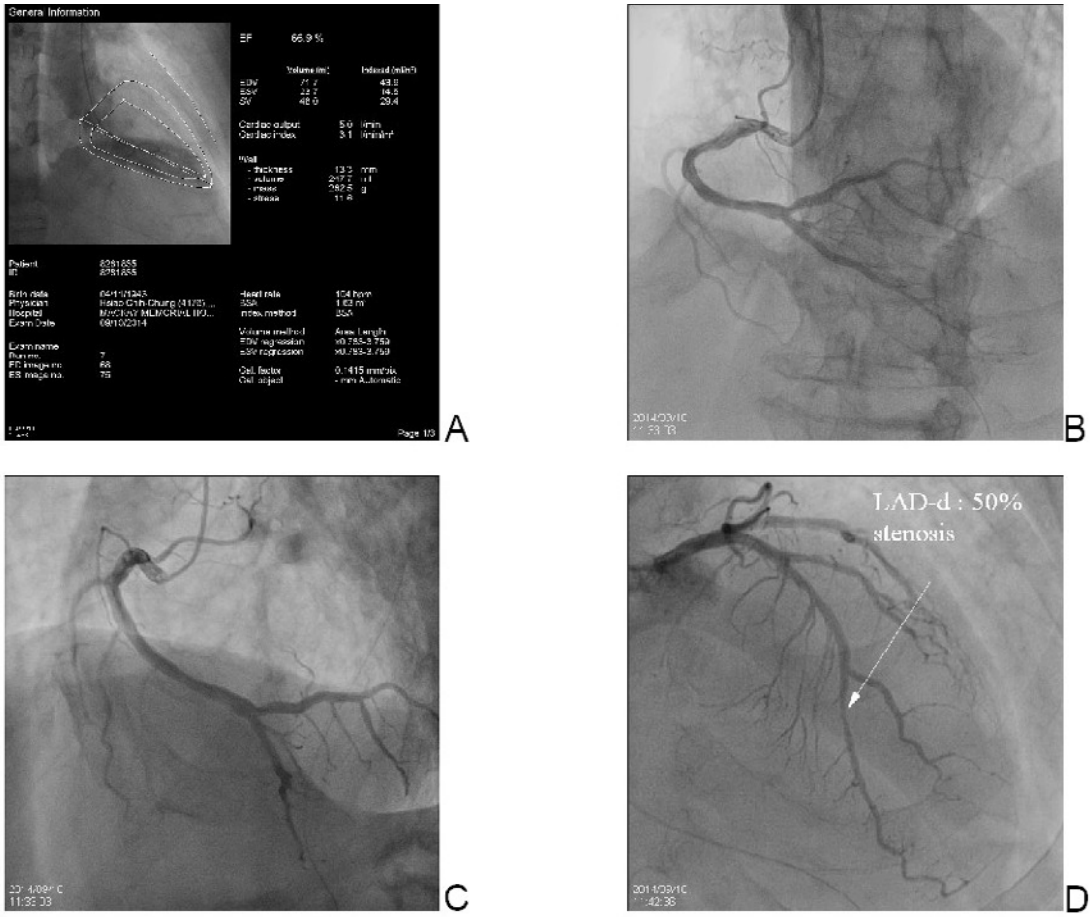

Figure 2. The EKG revealed QS pattern and disappearance of R wave over anterior wall.

denly. After cardiopulmonary resuscitation and intubation, his vital sign recovered. He was then transferred to ICU (intensive care unit). Elevation of cardiac enzyme was noted. (Troponin-I: 12.67ng/mL, CK: 966IU/L, CKMB: 74.5ng/mL). ECG revealed QS pattern and disappearance of R wave over anterior wall (Figure 2). Diagnostic cardiac catheterization was performed on Sep 10 and it revealed one vessel CAD (coronary artery disease) (Figure 3). Fluctuating blood pressure was noted in ICU. We checked 24hr-Urine VMA (Vanillylmandelic acid) and catecholamine for 2 days. The data revealed elevation of both VMA and catecholamine (Table 1). Correlated the clinical condition with laboratory data and CT findings, functional paraganglioma was highly suspected. Surgery is indicated. However, considering the patient's age, his family refused operation after discussion. The patient was discharged on Sep 27 with Terazosin and Labetalol for blood pressure control.

Discussion

The terms of "pheochromocytomas" and "catecholamine-secreting paragangliomas" are referred to as catecholamine-secreting tumors. Pheochromocytomas arise from adrenal medulla whereas paragangliomas arise from extra-adrenal paraganglia. The histology of paragangliomas and pheochromocytomas are similar. Sometimes, pheochromocytomas may be considered as intra-adrenal sympathetic paragangliomas. Because of similar clinical presentations and treatments, some clinicians might use "pheochromocytoma" to refer to both of them. The classic triad of pheochromocytomas or paragan-

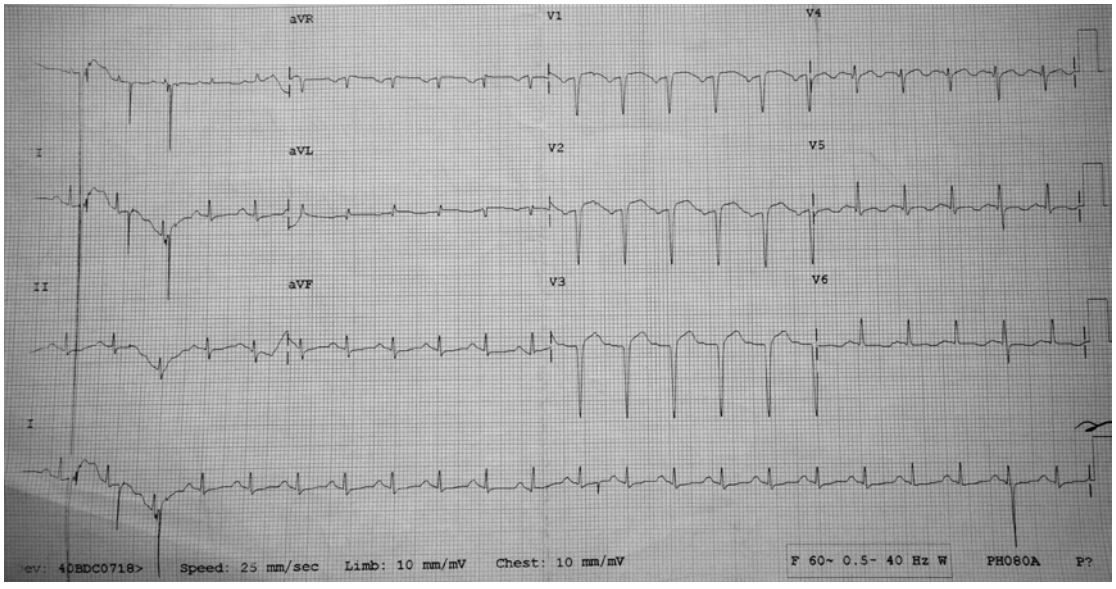

Figure 3. Cardiac catheterization data: (A) Left ventriculography demonstrated preserved LV systolic function (EF 65.9%); (B) (C) RCA patent; (D) LAD-D 50% stenosis; LCX hypoplasia.

gliomas includes symptoms of episodic headache, sweating, and tachycardia. The pathogenesis of secondary hypertension and the role of catecholamines in the pathophysiology of pheochromocytomas or paragangliomas have been well documented[1]. Indeed, sustained or paroxysmal hypertension is common, but about 5 to 15 percent of patients present with normal blood pressure[2]. With high sensitivity, plasma metanephrines remains the first-line test as diagnosis of pheochromocytomas or paragangliomas. However, it is not available in some hospital. Another reliable method for diagnosis is measuring metanephrines and catecholamines in a 24-hour urine collection, both of them have high sensitivity and specificity. Adrenalectomy by an experienced surgeon is suggested as treatment of pheochromocytomas or paragangliomas. However, surgical intervention does not always lead to long-term cure. Long-term monitoring is indicated in all patients, even those apparently cured[3].

The laboratory diagnostic cutoffs for pheochromocytomas or paragangliomas are based on normal reference of general population. However, false-positive results are common. According to the UpToDate(Clinical presentation and diagnosis of pheochromocytoma, section on initial biochemical tests), the diagnosis of pheochromocytomas or paragangliomas should be highly suspected if one or more of the following findings are found in 24hr-urine studies:

Table 1. LAB Data

Items	Day 1	Day 2	Normal range
Catecholamines			
--Norepinephrine	205.6	168.7	11.1~85.5 (ug/dL)
--Epinephrine	16.2	15.1	< 22.4 (ug/dL)
--Dopamine	157.8	191.3	50~450 (ug/dL)
VMA	11.8	13	1.0~7.5 (mg/day)

1. Normetanephrine >900 ug/24 hours or metanephrine >400 ug/24 hours
2. Norepinephrine >170 ug/24 hours
3. Epinephrine >35 ug/24 hours
4. Dopamine >700 ug/24 hours

If positive laboratory evidence is noted, image studies should be initiated to locate the tumor. In this case, a 5cm heterogeneous retroperitoneal mass occupied at right para-aortic region was found in CT scan. In the differential diagnosis of retroperitoneal

mass lesions, pheochromocytomas or paragangliomas should never been neglected. The laboratory data revealed high levels of catecholamines and VMA in 24hr-urine studies (Table 1). Correlated with the finding of CT scan, a paraganglioma is the most likely diagnosis.

In fact, unusual presentation of pheochromocytomas or paragangliomas as acute myocarditis or cardiogenic shock has ever been reported[4,5,6]. The clinical presentation of myocarditis is highly variable. In patients who have elevated cardiac biomarkers, ECG changes, arrhythmia, or other unexplained cardiac abnormalities, myocarditis should never be neglected. Although Troponin is the most common marker to diagnose acute myocardial infarction, it is not specific for acute thrombotic occlusion of a coronary artery. Troponin also increase in a variety of other diseases, such as sepsis, atrial fibrillation, heart failure, pulmonary embolism, myocarditis, myocardial contusion, and renal failure. When a patient presents with clinical manifestations of acute myocardial infarction but the coronary angiogram is normal, myocarditis should be considered. A definitive diagnosis of myocarditis is based on endomyocardial biopsy. In this case, the diagnostic cardiac catheterization revealed one vessel CAD. Echocardiogram demonstrated preserved LV systolic function (Table 2). Although it has not been proven by tissue biopsy, the combination of clinical presentation and noninvasive and invasive diagnostic findings suggest the diagnosis of myocarditis related to paraganglioma. Also, the pathophysiology of car-

Table 2. Echocardiogram report

	Data	Normal range
AO (aortic root dimension)	31	20-38 mm
AV (aortic valve)	19	16-26 mm
LA (left atrium)	26	19-40 mm
RV (right ventricle)	10	5-21 mm
EF slope	43	80-150 mm/sec
IVS (Interventricular septum)	10	7-11 mm
LVPW (Left ventricle posterior wall)	11	9-11 mm
IVRT (Isovolumic relaxation time)	110	70-90 msec
LVIDs (Left ventricular internal diameter end systole)	29	24-36 mm
LVIDd (Left ventricular internal diameter end diastole)	43	37-53 mm
LVEDV (Left ventricular end-diastolic volume)	80.8	80-190 ml
LVESV(Left ventricular end-systolic volume)	30.9	16-83 ml
EF (Ejection fraction)	61.8	50-70 %
CO (Cardiac output)	3.4	2.5-7.2 (1/m)
DT (Deceleration time of mitral inflow)	260	160-220 msec
HR (Heart rate)	69	60-100 bpm
E (Early ventricular filling velocity)	64.2cm/s	
A (Late ventricular filling velocity)	93.8cm/s	

Conclusions:
.Normal sizes of cardiac chambers.
.Preserved global contractility of left ventricle.
.LVEF by M-mode is 61.8 %.
.Grade II diastolic dysfunction.
.Mild tricuspid regurgitation.
.Trivial mitral regurgitation.
.Estimated systolic pulmonary artery pressure is 38 mmHg.
.EKG: Sinus rhythm.

diomyopathy due to pheochromocytoma has been tried to be explained[6,7]. High catecholamine level may directly cause membrane damage and death with degeneration and necrosis of contraction band, interstitial fibrosis, myocyte infiltration, vasoconstriction and medial thickening of coronary arteries. Long-term elevation of catecholamines may lead to cardiomyopathy, acute myocarditis, myocardial infarction, or acute hemodynamic collapse(8). Correlation with clinical findings, myocarditis related to paraganglioma was the most possible reason of sudden collapse in this case.

The laparoscopic adrenalectomy is the procedure of choice for pheochromocytoma or paraganglioma treatment. It can be safely performed in more than 90 percent of cases. In this case, however, the patient's family refused operation even though we had already explained the clinical importance and indication to them. All patients should accept alpha-adrenergic blockade before the procedure. Target blood pressure is less than 120/80 mmHg when sedated, and the systolic blood pressure should be greater than 90 mmHg when standing. After adequate blood pressure control, beta-adrenergic blockade should then be given. In this case, we prescribed Terazosin and then Labetalol for the patient.

Although most clinicians could recognize the classic symptoms, the clinical presentation of pheochromocytomas or paragangliomas may mimic severe diseases. If fewer symptoms were presented, early diagnosis may be more difficult. Patients with unrecognized pheochromocytomas or paragangliomas presenting with critical illness such as sepsis, shock, or acute myocardial infarction may have poor prognosis. For clinicians, pheochromocytoma or paraganglioma should always be kept in mind even though initial manifestations are not suggestive for the diagnosis.

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後腹腔腫瘤個案討論：副神經節瘤

楊晉州

衛生福利部草屯療養院

摘　要

嗜鉻細胞瘤(pheochromocytoma)以及副神經節瘤(paraganglioma)是分泌兒茶酚胺(cathecolamine)的罕見神經性腫瘤。一位71歲的男性病患，因腹痛、心悸、頭暈、頭痛就醫，急診檢查發現一個5公分大的後腹腔腫瘤而住院。該病患住院中喪失意識，經心肺復甦急救後轉入加護病房，診斷為副神經節瘤引起的心肌炎所導致。嗜鉻細胞瘤及副神經節瘤可能會以許多危及生命的心血管疾病做為表現，例如：急性心肌炎、心肌梗塞、以及高血壓危症等。在後腹腔腫瘤的鑑別診斷當中，切不可輕忽。