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Case Report

A case of paroxysmal pheochromocytoma concurrent with coronary artery aneurysm presenting as acute coronary syndrome

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ABSTRACT

A 51-year-old man recently diagnosed with preclinical Cushing's syndrome complained of chest oppression concomitant with back pain. Following contrast-enhanced computed tomography (CT) to rule out acute aortic dissection, he developed chest symptoms accompanied by elevation of blood pressure to 240/120 mmHg and ischemic electrocardiographic change. Urgent coronary angiography revealed a coronary artery aneurysm (15 mm × 6 mm) in the distal portion of the left anterior descending artery concomitant with coronary flow delay. Re-analysis of the blood sample taken at admission showed elevated plasma catecholamine concentrations, leading to a diagnosis of paroxysmal pheochromocytoma. An adrenal tumor was excised laparoscopically and histologically shown to be a pheochromocytoma. These findings show that coronary artery aneurysm may be a rare complication of pheochromocytoma, and indicate that monitoring of blood pressure or analysis of stored blood samples, if necessary, is essential to detect pheochromocytoma when using contrast medium or glucagon in patients known to have an adrenal incidentaloma. It should be noted that pre-treatment with an α-blocker is necessary when patients who are likely to have pheochromocytoma need to undergo contrast-enhanced CT.

<**Learning objective:** Coronary artery aneurysm may be a rare complication of pheochromocytoma. Monitoring of blood pressure or analysis of stored blood samples, if necessary, is essential to detect pheochromocytoma when using contrast medium or glucagon in patients known to have an adrenal incidentaloma. It should be noted that pre-treatment with an α-blocker is necessary when patients who are likely to have pheochromocytoma need to undergo contrast-enhanced CT.>

Introduction

Pheochromocytomas are relatively rare, generally benign tumors, usually located in the adrenal medulla, and related to catecholamine-producing paragangliomas. Typical presentations include headaches, palpitations, sweating, pallor, and sustained or paroxysmal hypertension due to excess catecholamine secretion. Clinical expression of pheochromocytoma can include several acute cardiovascular manifestations, such as hypertensive crisis, shock, acute coronary syndrome, acute heart failure, Tako-tsubo cardiomyopathy, lethal arrhythmia, cerebrovascular events, and aortic dissection [1].

Coronary artery aneurysms are often encountered in clinical cardiology and are usually associated with atherosclerosis, or with inflammatory, infectious, or iatrogenic disease, and are usually located in the proximal coronary artery. The incidence of coronary artery aneurysm has been reported to be 1.4% in autopsy cases and 4.9% in patients with suspected coronary artery disease who undergo coronary angiography [2].

We describe a patient with paroxysmal pheochromocytoma concurrent with coronary artery aneurysm that presented as acute coronary syndrome.

Case report

A 51-year-old man was recently diagnosed with preclinical Cushing's syndrome because of (a) the presence of a right adrenal tumor, an incidentaloma, (b) the lack of overt signs of Cushing's syndrome, and (c) autonomic cortisol secretion as confirmed by

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low-dose (1 mg) and high-dose (8 mg) dexamethasone suppression tests. At that time, however, he showed no definite findings suggestive of another hypothalamo-pituitary adrenal disorder, including pheochromocytoma. He had experienced paroxysmal nocturnal palpitations over the past 5 years. Although he was slightly obese (height, 162.5 cm; body weight, 70.1 kg; body mass index, 26.5 kg/m^2), annual routine medical check-ups did not confirm a diagnosis of arrhythmia, hypertension, diabetes, or dyslipidemia. In addition, he had no history of ischemic heart disease, Kawasaki disease, syphilis, or systemic vasculitis.

Five months after the initial endocrinological examinations, the patient presented at the Emergency Service department of our hospital complaining of chest oppression concomitant with back pain, palpitations, non-bilious vomiting, and periodic sweating lasting for 6 h. At the time of arrival, however, his symptoms were somewhat alleviated. A thorough physical examination showed a trace of heavy sweating, despite his being afebrile. He had a regular pulse rate of 82 beats per minute, dual heart sounds with no cardiac murmur, clear lung fields, and no notable findings in the abdomen. Alternatively measured blood pressure showed slight elevation in his right arm (140/88 mmHg) compared with his left arm (127/80 mmHg). Initial electrocardiography (ECG) findings are shown in Fig. 1A. Plain chest roentgenography showed neither cardiomegaly nor pulmonary congestion, and trans-thoracic echocardiography showed no evidence of a wall motion abnormality of the left ventricle, significant valvular dysfunction, pericardial effusion, or left ventricular hypertrophy. Initial blood analysis showed abnormally elevated creatine kinase (CK) (283 IU/l), CK-MB (41 IU/l), and troponin I (6.45 ng/ml) levels. Emergency contrast-enhanced computed tomography (CT) showed no evidence of aortic dissection. Subsequently, however, he complained of chest oppression concomitant with back pain, palpitations, and heavy sweating. A second ECG showed T inversion in the V4-6 leads (Fig. 1B). His blood pressure rose to 240/120 mmHg. Within 5 min after sublingual nitrate spray, his symptoms vanished completely, and his blood pressure normalized (130/70 mmHg). Because of a preliminary diagnosis of probable acute coronary syndrome, we performed emergency coronary angiography, which showed a coronary artery aneurysm, measuring 15 mm × 6 mm, in the distal portion of his left anterior descending artery, along with coronary flow delay (thrombolysis in myocardial infarction grade 2) caused by stasis of blood flow at the coronary aneurysm (Fig. 2A and B). No significant coronary stenosis was found at either the inflow or the outflow tract of the coronary artery aneurysm. No notable findings were observed in the right coronary artery. During coronary angiography, his blood pressure also rose periodically concomitant with the same symptoms and ECG changes. His condition stabilized in the intensive care unit with treatment with low-dose aspirin, intravenous unfractionated heparin, and intravenous diltiazem.

Re-analysis of the blood sample taken in the Emergency Service department showed that the patient's plasma catecholamine concentrations were elevated (epinephrine, 2577 pg/ml; norepinephrine, 923 pg/ml; dopamine, 49 pg/ml). Glucagon test was performed while monitoring blood pressure. Following injection of 1 mg glucagon, the patient's blood pressure rose from 120/70 mmHg to 140/90 mmHg, and his plasma catecholamine concentrations were significantly elevated. A 24-h urine specimen also showed a large quantity of secreted catecholamine metabolites (Table 1). Re-evaluation of the previous CT images revealed that the right adrenal tumor had increased in size, from 26 mm × 20 mm to 29 mm × 21 mm over 7 months (Fig. 3A and B). ^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy revealed positive radioactive uptake by this tumor, but distant metastases were not observed (data not shown). Paroxysmal pheochromocytoma concurrent with coronary artery aneurysm was finally diagnosed. The patient's symptoms subsequently stabilized with doxazosin (4 mg/day), low-dose aspirin (100 mg/day), and sustained-release diltiazem (200 mg/day).

Approximately 1 month later, the patient underwent laparoscopic excision of the right adrenal tumor. He recovered uneventfully following surgery. Histologic examination of the specimen showed tumor cells in the adrenal gland containing both amphophilic granule-containing cytoplasm and a clear nucleus proliferating in a nodular pattern, and compressing the normal

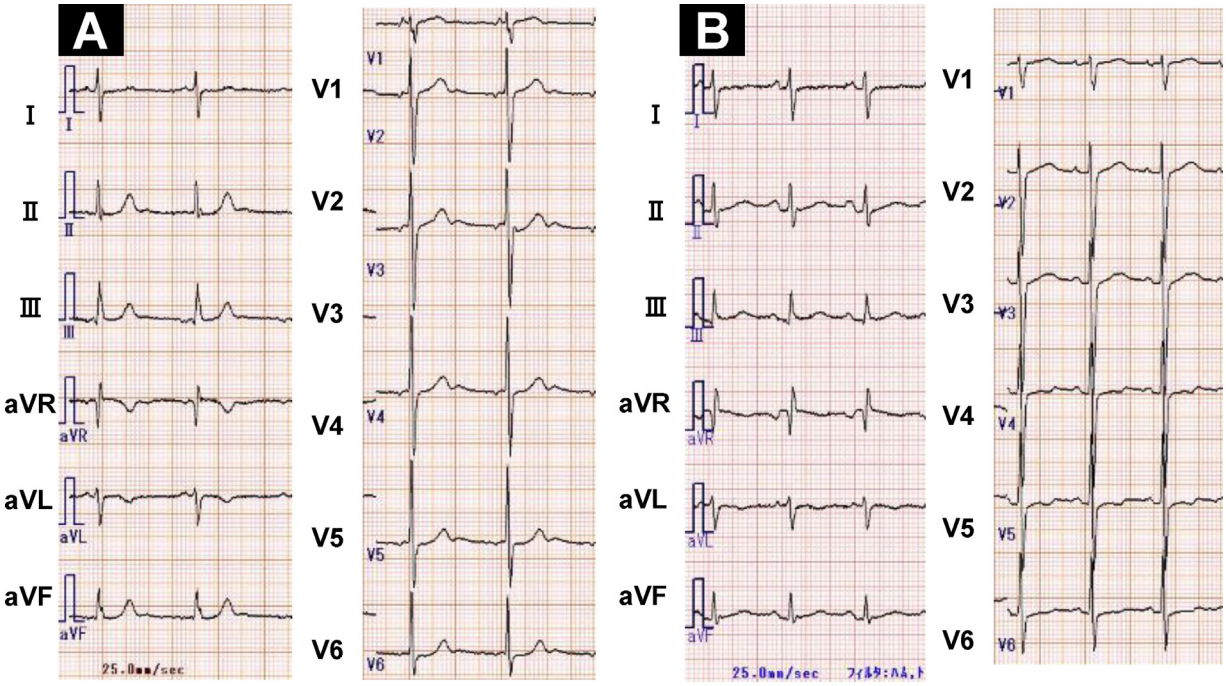

Fig. 1. (A) Initial electrocardiography (ECG) showed no remarkable findings except for ectopic atrial rhythm. (B) The second ECG when he complained of chest symptom soon after contrast-enhanced computed tomography showed T inversion in the V4-6 leads.