

Utility of Alpha-Blockade in a Hypotensive Pheochromocytoma Patient with Myocardial Infarction

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Key Words

Pheochromocytoma · Cardiomyopathy ·
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Abstract

Objective: The aim of this case study is to emphasize the importance of α -blockade in managing a rare complication of an untreated pheochromocytoma. **Clinical Presentation and Intervention:** A 41-year-old man with previous bilateral pheochromocytoma presented with chest pain. He was suffering from cardiac failure and persistent hypotension requiring an inotrope. Cardiac markers, an electrocardiogram and an echocardiogram confirmed acute myocardial infarction with poor ejection fraction and global hypokinesia. An ¹⁸F-fluorodeoxyglucose PET/CT scan showed progressive left suprarenal and organ of Zuckerkandl pheochromocytomas. Blood pressure stabilisation proved challenging but was achieved by titrating an incremental dose of α -blocker against a tapering inotropic dose. **Conclusion:** This case showed the efficacy of an α -blocker despite persistent hypotension in a patient with pheochromocytoma-induced cardiomyopathy.

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Introduction

Pheochromocytoma is a neuroendocrine tumour of the adrenal gland which secretes high levels of catecholamine. Classically, patients with pheochromocytoma present with the triad of episodic headaches, palpitations and increased sweating [1]. Other presenting symptoms related to the excessive release of catecholamines include hypertension, weight loss, hyperglycaemia, abdominal pain and fever. In addition to hypertension, other cardiovascular complications, such as acute myocardial infarction (MI), ischaemic heart disease and heart failure, are rarely seen in patients with pheochromocytoma [2]. Hence, we are reporting this case to illustrate the challenges involved in the management and the usefulness of an α -blocker in pheochromocytoma-induced cardiomyopathy.

Case Report

A 41-year-old man was admitted with sudden-onset chest pain which was associated with shortness of breath and sweating. On examination, his blood pressure (BP) was 80/50 mm Hg and his pulse was 100 bpm. He was tachypneic with an oxygen saturation

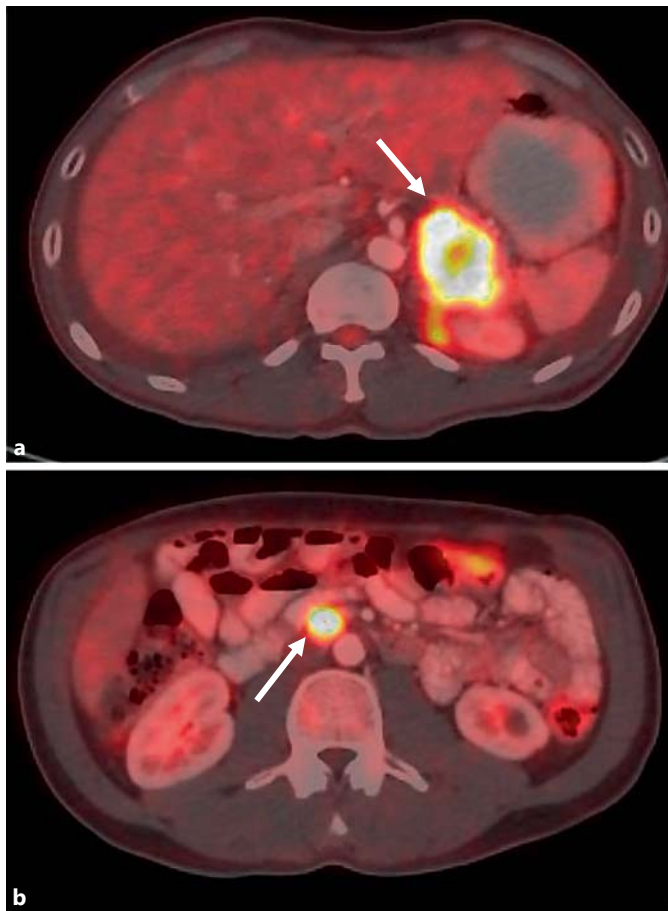


Fig. 1. Axial slices of fusion PET/CT images demonstrating hypermetabolic masses in the left suprarenal region (a) and anterolateral to the abdominal aorta at the level of the inferior mesenteric artery (b).

of 88% in room air. His electrocardiogram showed sinus tachycardia with ST-segment elevation in leads I, aVL and V4–V5. His troponin T was 6.1 µg/l (normal is <0.1 µg/l) and his creatine kinase-MB was 566 IU/l. The chest radiograph showed cardiomegaly with pulmonary congestion. He was treated for extensive anterolateral MI leading to cardiac failure (Killips III) and was thrombolysed.

A transthoracic echocardiography revealed severe left ventricular dysfunction with an estimated ejection fraction of 38.2% and global hypokinesia of the left ventricle. He was dependent on inotropic support for 5 days and remained in hospital for 2 weeks because his cardiac failure was unresolved for another 3 days. Less than 48 h after being discharged home, he was readmitted for unstable angina with left ventricular failure.

The patient had presented with a presumed acute coronary event in the past. The first presentation had been approximately 12 months previously, when he was treated for non-ST elevation MI. At that time, an echocardiogram had shown a normal ejection fraction of 70% with no hypokinesia. Further cardiac assessment was planned but the patient defaulted on his follow-up. Importantly,

his past medical history included a diagnosis of bilateral pheochromocytoma in 2002. Although he was scheduled for bilateral adrenalectomy, only the right adrenal could be successfully removed as the surgery was complicated by intraoperative bleeding. Upon recovery, he had declined further intervention. He then defaulted on the follow-up and resorted to buying over-the-counter antihypertensive medications. He only agreed to further treatment after these recent presumed acute coronary events. Apart from newly diagnosed diabetes and smoking, he did not have additional cardiac risk factors.

During a subsequent admission for angiography, he had an acute deterioration with chest pain, shortness of breath and hypotension, which continued to occur intermittently. Consequently, the angiography had to be postponed. Fluid resuscitation had to be performed judiciously due to the left ventricular failure. He remained hypotensive despite the up-titration of noradrenaline as inotropic support. His BP started to improve, from a systolic blood pressure of 80–85 mm Hg and a diastolic blood pressure of 40–49 mm Hg to a blood pressure above 110/65 mm Hg, only after the gradual introduction of prazosin, a low-dose oral α-blocker. Prazosin was introduced orally at 0.5 mg daily and up-titrated by 0.5 mg every other day. At a prazosin dose of 1.0 mg twice daily, his noradrenaline was successfully kept within an acceptable range. Meanwhile, his 24-hour urinary noradrenaline was markedly elevated at 5,455 nmol/24 h (normal is 40–780 nmol/24 h); his adrenaline was 32 nmol/24 h (normal is 5–80 nmol/24 h) and dopamine was 1,100 nmol/24 h (normal is 200–3,500 nmol/24 h). An ¹⁸F-fluorodeoxyglucose PET/CT scan showed a hypermetabolic left suprarenal mass with central necrosis measuring 5.1 × 3.8 × 5.5 cm, and another mass measuring 1.3 × 1.6 cm was anterolateral to the abdominal aorta at the level of the inferior mesenteric artery (fig. 1a, b). These findings were consistent with pheochromocytoma of the left adrenal and organ of Zuckerkandl.

Unfortunately, on day 10 after admission, he had another acute deterioration with a suspected further MI and cardiogenic shock requiring intubation. Despite triple inotropic support, his BP remained low. He developed ventricular arrhythmias and died on the same day. A post-mortem was planned but refused by the next-of-kin on religious grounds.

Discussion

This case highlights the difficulties involved in the management of cardiac complications due to long-standing untreated pheochromocytoma as has been previously reported [3, 4]. Approximately 10% of patients with pheochromocytoma have catecholamine-induced cardiomyopathy [1]. Excessive circulating catecholamines cause injury to myocytes, resulting in systolic dysfunction and subsequent dilated cardiomyopathy [2, 5].

The difficulty in distinguishing a catecholamine-induced myocardial ischaemia from a true MI due to coronary blockages in a patient with pheochromocytoma in a hypotensive crisis poses a dilemma for the attending physician in choosing between an α- or β-blocker as the treat-

ment. The use of a β -blocker in cardiomyopathy secondary to coronary artery disease would improve cardiac function. Conversely, the use of β -blockade before sufficient α -blockade would precipitate or worsen a pheochromocytoma crisis.

The previous few reports of pheochromocytoma suggestive of MI were mostly not accompanied with cardiac regional wall abnormalities or significant coronary blockages [5]. The high levels of catecholamine in pheochromocytoma exert a direct toxic effect on the myocardium and cause vasoconstriction or coronary spasm, leading to an imbalance between oxygen supply and demand [6]. Concurrently, excess catecholamine increases the permeability of pulmonary capillaries, leading to pulmonary oedema [5]. A catecholamine surge in these patients may also lead to myocardial hibernation or myocardial stunning [2, 3, 5].

The patient could not be treated with fluid resuscitation that might have exacerbated the underlying left ventricular failure; hence, we chose α -blockade in spite of the hypotension. The response of his BP to inotropes was limited as this was a patient who had chronically high levels of noradrenaline and dopamine. We chose prazosin over phenoxybenzamine in view of its short-acting property, which made it a safer option. The establishment of mechanical circulatory support is recommended to enable the initiation of α -blockade in hypotension [7]. Nev-

ertheless, the potential benefit of this procedure could not be offered to our patient as we lack the required expertise.

The improvement of our patient's BP with α -blockade strongly suggests that his cardiac complications were due to the effects of catecholamines rather than traditional cardiac risk factors, given that his diabetes was only diagnosed in the past year. It is difficult to conclusively say that this patient had purely catecholamine-induced cardiomyopathy without angiographic evidence of normal coronaries. His history of exposure to high catecholamines, mainly noradrenaline, could be traced as far back as 10 years prior to his first cardiac presentation. This chronic exposure was most likely responsible for the cardiomyopathy and irreversible hypotension, which proved to be fatal. The presence of paraganglioma in the organ of Zuckerkandl in addition to the left suprarenal mass points to an underlying genetic cause, which is substantiated clinically as one of his children has histologically proven bilateral pheochromocytoma. However, genetic phenotyping was not performed as we lack the facility.

Conclusion

This case showed the efficacy of α -blockers despite persistent hypotension in a patient with pheochromocytoma-induced cardiomyopathy.

References

- 1 Park JH, Kim KS, Sul JY, et al: Prevalence and patterns of left ventricular dysfunction in patients with pheochromocytoma. *J Cardiovasc Ultrasound* 2011;19:76–82.
- 2 Galetta F, Franzoni F, Bernini G, et al: Cardiovascular complications in patients with pheochromocytoma: a mini-review. *Biomed Pharmacother* 2010;64:505–509.
- 3 Gujja KR, Aslam AF, Privman V, et al: Initial presentation of pheochromocytoma with Takotsubo cardiomyopathy: a brief review of literature. *J Cardiovasc Med (Hagerstown)* 2010;11:49–52.
- 4 Darzé ES, von Sohsten RL: Pheochromocytoma-induced segmental myocardial dysfunction mimicking an acute myocardial infarction in a patient with normal coronary arteries. *Arq Bras Cardiol* 2004;82:175–177.
- 5 Prejbisz A, Lenders JWM, Eisenhofer G, et al: Cardiovascular manifestations of pheochromocytoma. *J Hypertens* 2011;29:2049–2060.
- 6 Menke-van der Houven van Oordt CW, Twickler TB, van Asperdt FGMH, et al: Pheochromocytoma mimicking an acute myocardial infarction. *Neth Heart J* 2007;15:248–251.
- 7 Whitelaw BC, Prague JK, Mustafa OG, et al: Pheochromocytoma crisis. *Clin Endocrinol (Oxf)* 2014;80:13–22.