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## **► To cite this version:**

Nicolas Lanot, Jérôme Adda, François Roubille, Mariama Akodad. Pheochromocytoma in a patient presenting with ventricular fibrillation and carotid dissection: a case report. *European Heart Journal. Case Reports*, 2021, 5 (2), <10.1093/ehjcr/ytab012>. <hal-03140068>

**HAL Id: hal-03140068**

**<https://hal.science/hal-03140068v1>**

Submitted on 12 Feb 2021

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# Pheochromocytoma in a patient presenting with ventricular fibrillation and carotid dissection: a case report

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## Background

Pheochromocytoma is an endocrine tumour secreting catecholamines, most often revealed by clinical symptoms (headache, palpitations, diaphoresis, or resistant hypertension). Some cases of ventricular arrhythmias were described in the literature, without any formal link between arrhythmia and pheochromocytoma.

## Case summary

We report a case of pheochromocytoma discovered after cardiac arrest due to ventricular fibrillation in a 46-year-old patient. The diagnosis was suggested by clinical symptoms (headache, palpitation, and diaphoresis) and suspected on the abdominal computed tomography scan. The diagnosis was corroborated by metaiodobenzylguanidine scintigraphy and finally confirmed by anatomopathological analysis of the operative specimen. The cerebral imaging showed a dissection of the left internal carotid artery and an intraparenchymal haematoma that might be secondary to a catecholaminergic discharge of phaeochromocytoma and severe hypertension.

## Discussion

Since pheochromocytoma is accessible to curative treatment, its detection in case of cardiac arrest is essential to decrease the risk of arrhythmic recurrence.

## Keywords

Case report • Pheochromocytoma • Ventricular fibrillation • Sudden cardiac death • Carotid dissection

## Learning points

- Pheochromocytoma is a rare but classical aetiology of cardiac arrest.
- Its detection is essential to decrease the risk of recurrence.
- From clinical suspicion, diagnosis is allowed by imaging, biology, and confirmed by pathology.

## Introduction

Pheochromocytoma is a rare endocrine tumour (annual incidence 1–4 per million) developing at the expense of adrenal medullary chromaffin cells, leading to an excess of catecholamines release.<sup>1</sup> This tumour is most often revealed by clinical symptoms such as headache, palpitations, and diaphoresis, or during the aetiological assessment of a resistant hypertension.<sup>2</sup> Some cases of ventricular arrhythmias

were described in the literature,<sup>3–5</sup> without any formal link between arrhythmia and pheochromocytoma. We present the case of a pheochromocytoma with ventricular fibrillation (VF), preceded by clinical symptoms suggestive of an adrenergic thrust, in a young patient.

ranging from 118 mmHg to 160 mmHg). The electrocardiogram showed regular sinus rhythm with regression of the QT interval prolongation (440 ms at Day 12; 360 ms at Day 68).

Transthoracic and transoesophageal echocardiography did not highlight any elements in favour of embologenic cardiopathy. LVEF

## Timeline

One month before the event	Asthenia and professional overwork
The event	Palpitations and intense headache, followed by cardiac arrest Resuscitation after three electric shocks
The event	Coronary angiography: healthy coronary network Ventriculography: inferobasal and inferoseptal segmental hypokinesia, with a preserved left ventricular ejection fraction (LVEF)
The event	Abdominal computed tomography (CT) scan: large retroperitoneal haematoma Cerebral CT scan: left fronto-parietal haematoma + focal dissection of intrapetrous portion of left carotid artery Cerebral magnetic resonance imaging (MRI): occipital lesions with a FLAIR hypersignal
12 days after	Transthoracic echocardiography: preserved LVEF, hypokinesia of medial inferoseptal and inferior territories. Normal contractility of the apex and loss of contractility of the basal segments on speckle-tracking
14 days after	Metaiodobenzylguanidine scintigraphy: extensive hyperfixation involving the fleshy part of the hypertrophied left adrenal gland, compatible with a pheochromocytoma
15 days after	Cardiac MRI: normal, no late gadolinium enhancement
22 days after	Subcutaneous cardioverter defibrillator implantation
34 days after	Surgery: laparoscopic left adrenalectomy
39 days after	Anatomopathology: left adrenal pheochromocytoma, with 45 mm long axis intra-tumoural haematoma
11 months later	<sup>18</sup> F-fluorodeoxyglucose-positron emission tomography scan: non-recurrence of pheochromocytoma
11 months after	Cardiological consultation: absence of arrhythmia recurrence in the defibrillator's memories

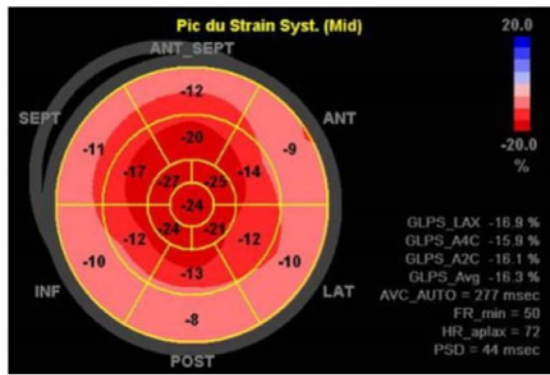
## Case presentation

A 46-year-old patient without significant medical history was admitted with VF leading to cardiac arrest following palpitations and intense headache in a context of asthenia and professional overwork. The no-flow time was estimated to 0 min (immediate cardiopulmonary resuscitation by bystanders), and the low-flow time (active cardiopulmonary resuscitation) to 15 min. Resuscitation was successfully obtained after three electric cardioversions. The electrocardiogram showed regular sinus rhythm with long QT interval (>440 ms) immediately after resuscitation.

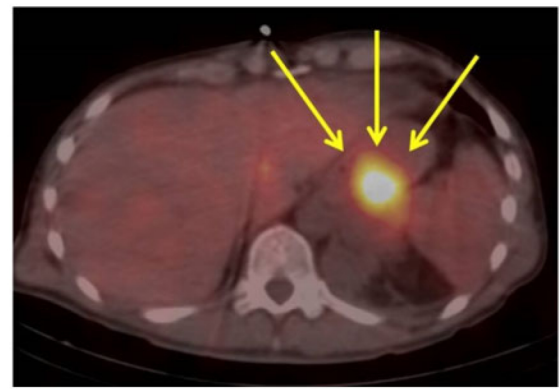
He was initially referred to the intensive care unit for haemodynamic instability with severe systolic left ventricular dysfunction and need of haemodynamic support with dobutamine 5 µg/kg/min (weaned at Day 2), norepinephrine 5 mg/h (weaned at Day 3), propofol (stopped at Day 1), sufentanil (stopped at Day 1), and amoxicillin—clavulanate (stopped at Day 5) were also administered. Coronary angiography performed at Day 1 revealed a normal coronary network, ventriculography revealed the presence of inferobasal and inferoseptal hypokinesia, with a preserved left ventricular ejection fraction (LVEF). The evolution was favourable allowing inotropic and vasopressor weaning at Day 2. The patient had no physical signs except moderate arterial hypertension (systolic blood pressure

was preserved (LVEF 63%, global longitudinal strain 16.3%) with a hypokinesia in medial inferoseptal and inferior territories. Speckle-tracking analysis showed a normal contractility of the apical segments, and a loss of contractility of the basal segments, compatible with a reverse Tako-Tsubo cardiomyopathy (Figure 1). Cardiovascular magnetic resonance imaging was normal without significant late gadolinium enhancement. Brain magnetic resonance imaging (MRI) performed as part of the cardiac arrest assessment found a left parieto-frontal intraparenchymal haematoma with posterior lesions.

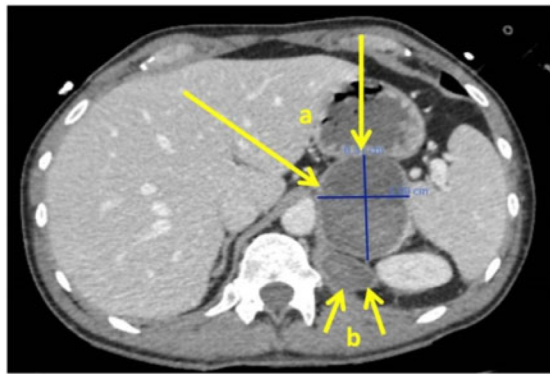
Abdominal computed tomography (CT) scan showed a left retroperitoneal haematoma associated with a left suprarenal haematoma of 64 mm, with no active bleeding or visible vascular abnormality (Figure 2). The ionogram was strictly normal throughout the hospitalization and moderate biological inflammatory syndrome appeared 10 days after the cardiac arrest (C-reactive protein peak 80 mg/L, thrombocytosis 700 giga/L) with stable moderate anaemia (haemoglobin 10 g/dL). The high-sensitive troponin did not exceed 81 ng/L (normal value < 14 ng/L). An elevation of plasma metacholines (metanephrin 379 ng/L for a normal value < 181 ng/L; normetanephrin 927 ng/L for a normal value < 236 ng/L) was noticed. In view of clinical signs and abdominal CT scan results, a metaiodobenzylguanidine (MIBG) scintigraphy was performed which confirmed the appearance of a left adrenal pheochromocytoma by fixation (Figure 3). During



**Figure 1** Two-dimensional global longitudinal speckle-tracking echocardiography analysis showing a normal apical contractility, with a loss of contraction of the basal segments, compatible with a catecholaminergic cardiomyopathy.



**Figure 3** Extensive hyperfixation involving the fleshy part of the hypertrophied left adrenal gland in I123-metaiodobenzylguanidine whole body scintigraphy.



**Figure 2** Abdominal computed tomography scan showing a left adrenal haematoma of 64 mm long-axis dissecting the adrenal parenchyma (a), associated with retroperitoneal haematoma (b).

hospital stay, nimotidine, bisoprolol, and ramipril were first administered. The patient remained stable along the hospitalization, without any recurrence of arrhythmia. Bisoprolol was substituted by an alpha-beta-blocker therapy (labetalol 200 mg q.d.), after scintigraphy and prior to the surgical resection of the tumour. Pheochromocytoma was confirmed by histological analysis of the surgical specimen. A subcutaneous defibrillator was implanted before the end of hospitalization in intensive care unit. The discharge treatment was as follows: labetalol 200 mg q.d., ramipril 2.5 mg q.d. Within the 11 months after the event, no symptomatic ventricular arrhythmia occurred in the defibrillator's memories.

## Discussion

The diagnosis of pheochromocytoma is generally based on the demonstration of an excessive production of catecholamines or their

degradation products, the methoxylated derivatives. The anamnesis was in this case the key to evoke the diagnosis (symptomatic triad headache, palpitations, sweating; arterial hypertension). The abdominal CT scan allowed to guide the diagnosis with the discovery of a left adrenal mass. Likewise, the brain imaging highlighted a left carotid dissection and deep intraparenchymal lesions. This location evokes a hypertensive pressure-related encephalopathy, falling within the framework of the pheochromocytoma physiopathology. The dosage of methoxylated derivatives is a reliable test to orientate the diagnosis, with a sensitivity<sup>3,6</sup> of 90% and a specificity of 98%.<sup>6,7</sup> However, its interpretation at initial phase may be disturbed by the adrenergic discharge secondary to the cardiac arrest or the administration of vaso-pressor amines. We measured plasma methoxylated derivatives (10 days after the end of amines administration) in order to remove this bias. MIBG scintigraphy confirmed the diagnosis by showing a fixation on the left adrenal mass, indicating the presence of an increased catecholamine secretion. Pheochromocytoma was known to give a high rate of cardiovascular complications, as well as ventricular arrhythmia and Tako-Tsubo-like cardiomyopathy.<sup>4</sup> Indeed, catecholamine secretion causes changes in repolarization that may be responsible for ventricular arrhythmias.<sup>8</sup> As well, patients with large tumours and high levels of biochemical markers were more likely to develop cardiac injury.<sup>9</sup> Patients with cardiac complications harboured larger tumours, exhibited higher biochemical marker levels, longer corrected QT interval, and lower ejection fraction.<sup>9</sup> Complete removal of the tumour may prevent the risk of recurrent ventricular arrhythmia.<sup>10,11</sup> Its multidisciplinary management is medico-surgical and involves cardiologists, endocrinologists, radiologists, visceral surgeons, anaesthesiologists, and pathologists. The radical cure is a delicate surgery usually preceded by a medical care mainly based on symptomatic treatment preventing the negative effects of bursts of catecholamines.<sup>12</sup> As the cardiovascular complications of pheochromocytoma can be life-threatening, several studies suggest to screen all patients with manifestations suggesting excessive catecholamine secretion.<sup>9,13</sup> This screening could avoid unnecessary cardiac interventions and allow a fast cardiac recovery.<sup>9,14</sup> Nonetheless, there is no evidence to support the link between VF and the discovery of

pheochromocytoma in this case. On MRI, no evidence for acute myocarditis or ischaemic disease was found. Given the clinical signs and the presence of brain lesions, the hypothesis of VF secondary to a catecholaminergic discharge remains the most probable. On the other hand, an idiopathic VF or an early dilated cardiomyopathy could not be formally discarded. Therefore, after multidisciplinary discussion, a subcutaneous defibrillator was implanted to prevent ventricular arrhythmias recurrence related to an unknown primitive cardiomyopathy or to recurrence of pheochromocytoma, as previously described in patients with multiple endocrine neoplasia.<sup>15,16</sup>

## Conclusion

This clinical case reveals a rare and serious potential complication of pheochromocytoma, a VF. In case of resuscitated VF in a young patient without obvious aetiology, abdominal CT scan, and methoxy-lated derivatives assessment should be performed to eliminate the diagnosis of pheochromocytoma particularly in case of suggestive symptoms. Indeed, since pheochromocytoma is accessible to curative treatment, its detection is essential to decrease the risk of arrhythmia recurrence.

## Lead author biography



Nicolas Lanot was born in Paris, France, in 1992. He received his medical training at the University of Montpellier-Nîmes and is currently in his final year of residency. His fields of interest are heart failure, paediatric and congenital cardiology, and sports cardiology. He has been involved in research at the Arnaud de Villeneuve hospital, University of Montpellier in France.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** None declared.

**Funding:** None declared.

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