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Electrical storm – a rare manifestation of paraganglioma triggering catecholamine crisis

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Abstract

An electrical storm is a life-threatening emergency characterized by recurrent and sustained ventricular tachycardia (VT) or ventricular fibrillation (VF). The clinical presentation of electrical storms can vary widely, requiring a systematic evaluation and personalized management approach. The management of electrical storms should be individualized based on the specific clinical presentation and underlying heart disease. We present a rare case of an electrical storm triggered by a catecholamine crisis induced by a paraganglioma, highlighting the critical role of early diagnosis and management of paraganglioma-related complications.

Keywords: electrical storm, ventricular arrhythmias, catecholamine-crisis, paraganglioma.

Introduction

Despite significant advancements in cardiovascular care over the past three decades, sudden cardiac death (SCD) and malignant arrhythmias continue to represent substantial public health challenges [1].

Electrical storms (ES) are characterized by recurrent and sustained episodes of ventricular tachycardia (VT) or ventricular fibrillation (VF). The term "electrical storm" is used to describe a condition where three or more distinct episodes of ventricular tachycardia (VT) or ventricular fibrillation (VF) occur within 24 hours. Electrical storms are becoming more prevalent and are considered life-threatening emergencies that demand hospitalization and intensive medical intervention [2, 3]. The clinical presentation of arrhythmias during an electrical storm can vary widely. In rare cases, patients may experience repetitive and hemodynamically destabilizing polymorphic VT or VF episodes that require adequate anti-arrhythmic medical therapy.

Early identification of the underlying cause of the electrical storm, such as acute ischemia (in acute myocardial infarction), structural heart disease (both non-ischemic and ischemic cardiomyopathies), or an inherited channelopathy, may be

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life-saving since it may help to guide management decisions. A systematic approach, starting with early diagnosis of the underlying cardiac disease, may be crucial to evaluate and manage electrical storms effectively [4].

The current management of electrical storms aims to terminate the arrhythmia, stabilize the patient, and prevent a recurrence. Treatment options may include medications (antiarrhythmic drugs), ICD therapy (shocks or anti-tachycardia pacing), catheter ablation (to eliminate the arrhythmogenic focus), or revascularization procedures (such as percutaneous coronary intervention or coronary artery bypass grafting) [4, 5]. ICD implantation is contraindicated during an arrhythmia storm, as it does not address the underlying cause. ICD placement is typically reserved for cases where arrhythmias have been stopped with medication, revascularization (if applicable), ablation, and in some patients, sympathectomy [2, 3, 5]. It is, however, important to note that the management of electrical storms should be individualized based on the specific clinical presentation, underlying cardiac disease, and available resources. Close collaboration between cardiologists, electrophysiologists, and critical care specialists is often necessary to optimize patient outcomes.

Here we present an uncommon and rare case illustrating the occurrence of a malignant arrhythmia and electrical storm triggered by a catecholamine crisis induced by a paraganglioma.

The medical management of catecholamine-producing tumors involves a combination of pharmacological therapy to control symptoms, preoperative preparation to minimize the risk of intraoperative hypertensive crises, and postoperative care. Surgical treatment with optimal preoperative preparation is required in certain situations, such as in the presence of underlying causes or triggering factors like catecholamine hypersecretion from tumors (such as pheochromocytoma/paraganglioma) [5, 6].

This article highlights the critical role of early identification and management of paraganglioma-related complications.

Case presentation

A 69-year-old male known with hypertension, permanent atrial fibrillation, and secondary pulmonary hypertension presented to the emergency department

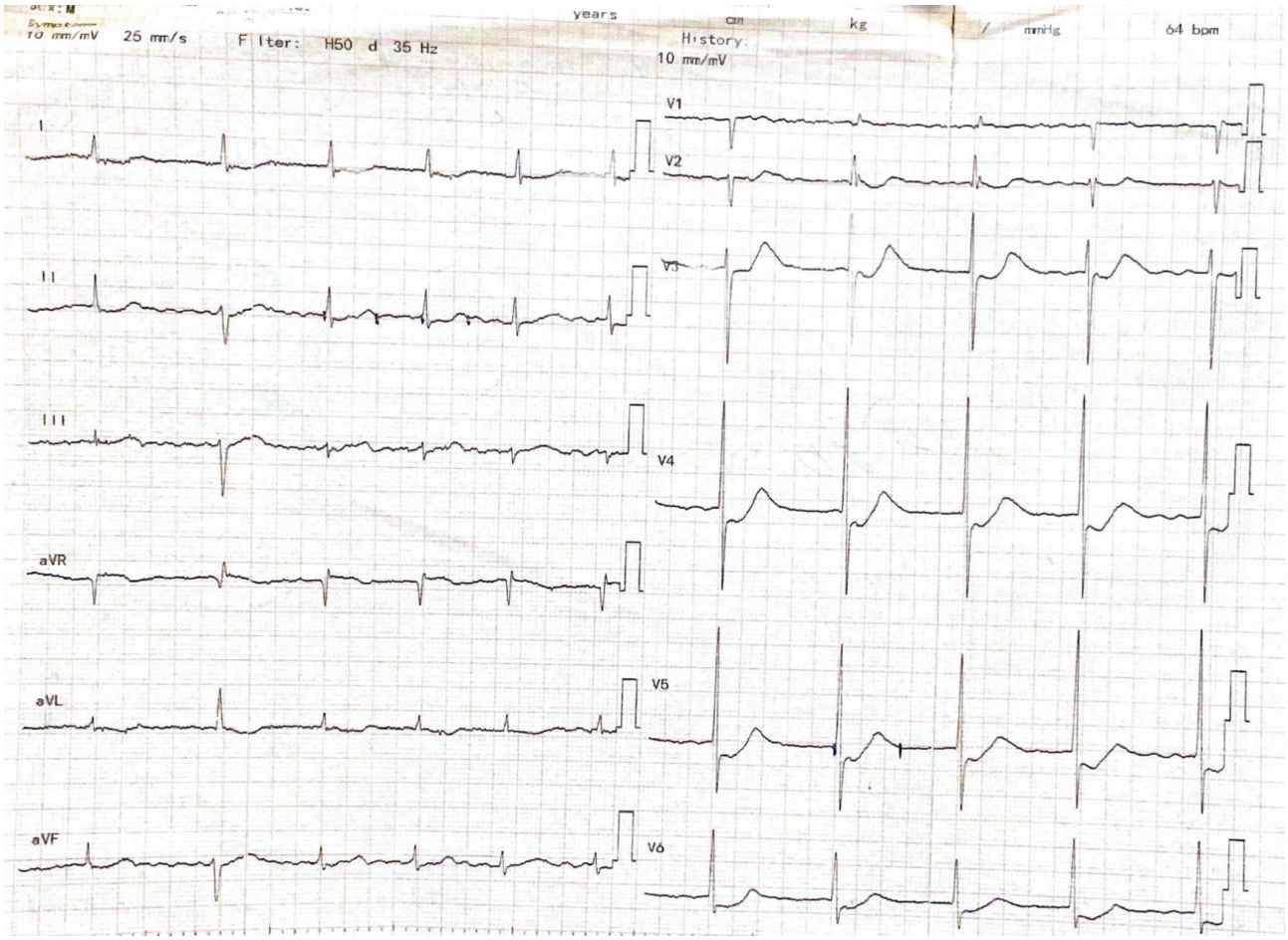

Figure 1. The 12 leads ECG showed ST-segment depression in leads V3 to V6.

with constrictive chest pain with radiation in the interscapulovertebral region. In addition, these symptoms were associated with dyspnea, sweating, two syncopal episodes, and one monomorphic non-sustained ventricular tachycardia (NSVT) episode. The 12 leads ECG showed depression of segment ST over leads V3 to V6 (Figure 1). The patient's serum creatinine and leucocytes were increased, with liver and thyroid function tests within normal limits.

The echocardiographic evaluation revealed an ejection fraction of 50%, indicating preserved left ventricular systolic function. In addition, the thickness of the interventricular septum was 14 mm, suggesting the presence of mild interventricular septal hypertrophy with a medioventricular gradient was 15 mmHg. Emergency coronary angiography did not reveal any significant coronary abnormalities.

During hospitalization, his blood pressure was severely unstable, with remarkable blood pressure fluctuation from 260/130 mmHg to 60/40 mmHg, which was extremely sensitive to vasoactive substances. Moreover, the hypertension crises were associated with electrical storms with multiple episodes of NSVT and VT (Figure 2), making cardioversion necessary with 200 J DC shock.

Contrast CT abdomen revealed an inhomogeneous, cystic, polylobate mass in the retroperitoneal, paraaortic, infrarenal region, with a size of 7,3/7,0/7,2, suggestive of extra-adrenal paraganglioma (Figure 3).

Laboratory tests showed elevated levels of 24 h urinary metanephrines: 726 µg (normal range: 59-394 µg), elevated normetanephrine: 15476 µg (normal range: 129-937 µg), noradrenalin 834,88 µg (normal values <97 µg), while adrenaline and dopamine levels were within normal limits (Table 1).

Following appropriate medication preparation with alpha and beta blockers, the patient underwent exploratory surgery, during which the tumor described by CT imaging was excised (Figure 4). Repetitive arrhythmias were successfully terminated after tumor removal, and the histopathological examination of the excised tumor confirmed the diagnosis of catecholamine-producing paraganglioma.

Discussion

An electrical storm (ES) is characterized by the occurrence of at least three sustained episodes of ventricular tachycardia (VT), ventricular fibrillation (VF), or appropriate shocks from an implantable

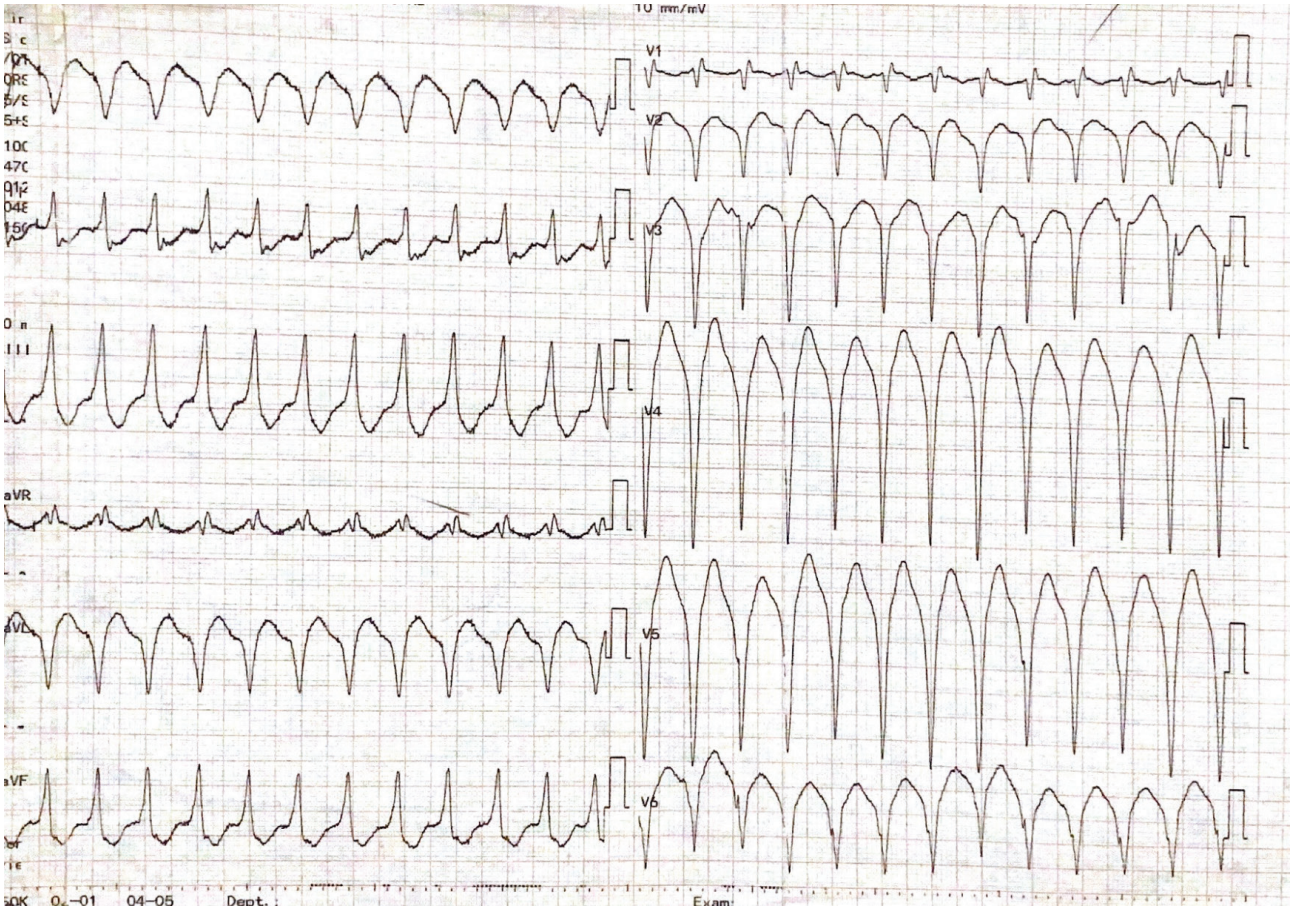

Figure 2. ECG during an episode of electrical storm accompanied with hypertensive crisis.

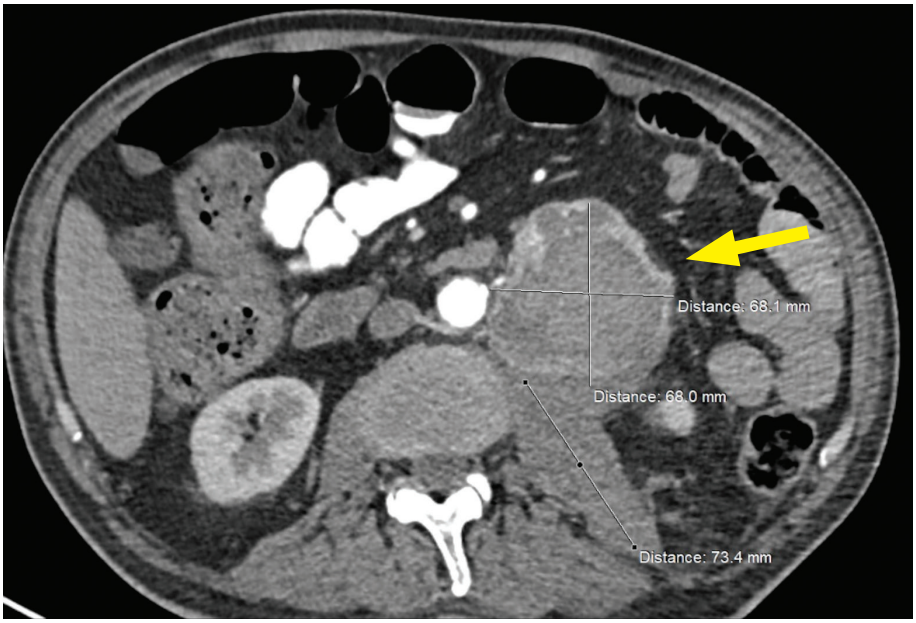

Figure 3. Contrast CT abdomen revealed an inhomogeneous, cystic, polylobate 7.3 cm x 6.8 cm x 6.8 cm mass (yellow arrow) in the retroperitoneal, paraaortic, infrarenal region, suggestive for extra-adrenal paraganglioma.

cardioverter-defibrillator (ICD) within 24 hours [7]. Various case reports have documented the cardiac manifestations associated with pheochromocytomas and paragangliomas, which include acute coronary syndrome, VT, and Takotsubo cardiomyopathy. Interestingly, some cases have even presented with VT as the initial symptom of pheochromocytomas or paragangliomas [8–14].

Coronary angiography is a gold standard procedure for visualizing the coronary arteries and the presence of coronary abnormalities, and it plays a crucial role in determining the etiology of tachyarrhythmias. In our case, there were no significant stenoses on coronary angiography, suggesting the need for further investigations in order to determine the underlying cause of the patient's symptoms.

Throughout the patient's hospital stay, we observed severe blood pressure instability, notable fluctuations ranging from hypertensive crisis to hypotension, extreme sensitivity to vasoactive substances, and associated arrhythmias suggesting a prominent catecholamine efflux. In addition, this case highlights the intricate interplay between catecholamine release, blood pressure dysregulation, and arrhythmogenic potential.

Pheochromocytomas are rare tumors developed from chromaffin cells found in the adrenal medulla and extra-renal sympathetic paraganglia, named paragangliomas [15]. They can continuously or episodically produce a surplus of catecholamines, such as epinephrine and norepinephrine [16].

Although pheochromocytoma and paraganglioma can present with various clinical manifestations, arrhythmias and hypertensive crises account for 71% of the mortality of these critical cases [17]. At the level of the cardiovascular system, it can cause

Table 1. Elevated levels of urinary and serum biomarkers suggestive for catecholamine crisis.

	Patient values	Normal range
Urinary metanephrines	**726,18 µg/24h**	(59,00–394,00 µg/24h)
Urinary normetanephrines	15476,20 µg/24h	(128,00–934,00 µg/24h)
Urinary adrenaline	22,30 µg/24h	(≤27,00 µg/24h)
Urinary noradrenaline	834,88 µg/24h	(≤97,00 µg/24h)
Urinary dopamine	394,80 µg/24h	(≤500,00 µg/24h)
Acid vanilmandelic	**38,55 mg/24h**	(1,60–7,30 µg/24h)
Acid homovanilic	**8,03 mg/24h**	(1,80–6,90 µg/24h)

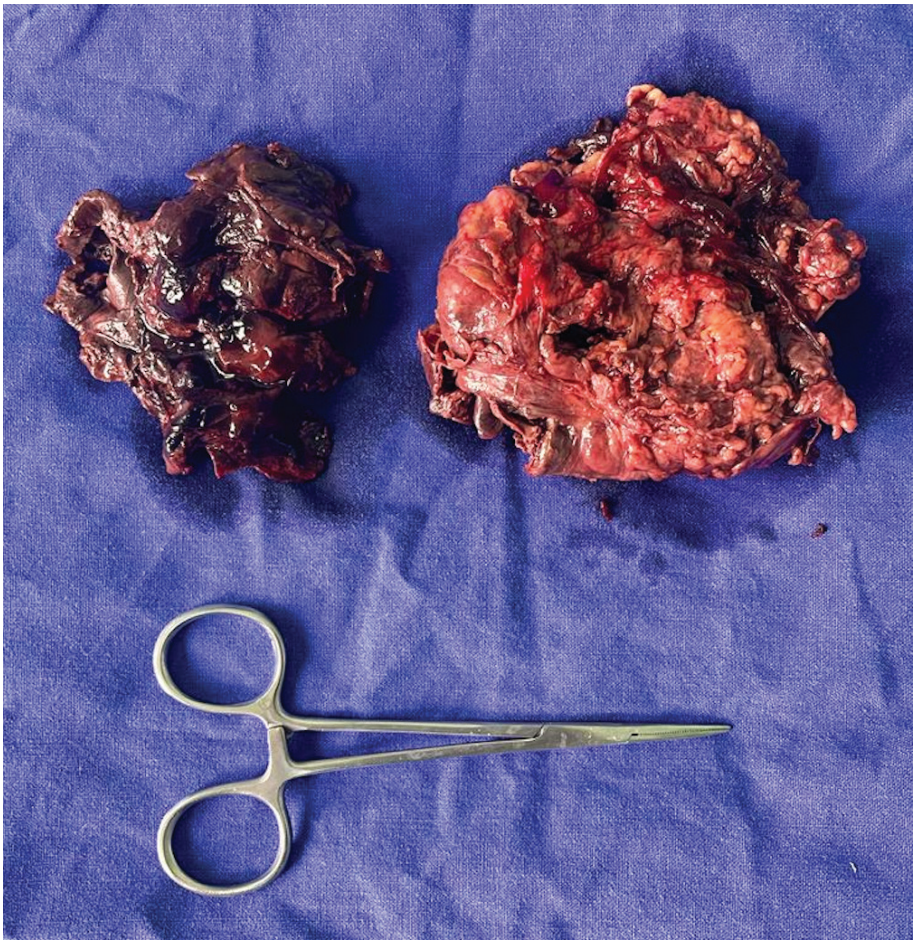

Figure 4. Fragments from the tumor removed during exploratory surgery, which was confirmed by histopathology as catecholamine-producing paraganglioma.

cardiac complications such as heart failure, myocardial infarction, cardiomyopathy, and severe, potentially lethal arrhythmias. Arrhythmias are found in 10-20 % of patients with pheochromocytomas and may consist of sinus tachycardia, atrial fibrillation in 60% of cases, bradyarrhythmia in 20 % of cases, or ventricular tachycardia in 13% of cases [18–20].

Managing catecholamine-induced tachyarrhythmias may represent a complex challenge as they frequently resist to conventional medical treatments. Without intervention, a catecholamine crisis can lead to severe complications such as hypertensive crises, hypotensive shock, and death [21]. Given the intricate nature of these cardiovascular complications and the evolving treatment protocols for arrhythmias, the involvement of a cardiologist becomes crucial in ensuring optimal care in these critical patients [22].

Identifying a pheochromocytoma's or paraganglioma's specific biochemical phenotype is crucial to assess catecholamine-induced tachyarrhythmias. These phenotypes are characterized by the production and release of catecholamines, which elicit cardiovascular responses. Therefore, understanding these phenotypes, categorized as noradrenergic, adrenergic, or mixed, is vital as they directly determine the frequency of arrhythmias, the level of blood pressure disturbances, and the overall risk of the patient.

Catecholamines affect the heart by activating adrenergic receptors in myocytes and the conduction system. The surge of catecholamines (characterized by an excessive release of catecholamines such as epinephrine and norepinephrine) can lead to alterations in intracellular calcium, resulting in abnormal electrical impulses and disturbances in myocardial repolarization. Additionally, enhanced sympathetic stimulation can modulate ion channel activity, including the sodium, potassium, and calcium channels, significantly influencing cardiac excitability and impulse propagation. These molecular and cellular changes contribute to the initiation and perpetuation of arrhythmias [23].

Catecholamine excess can result from developing various types of VT, including monomorphic, bidirectional, and polymorphic VT. While the exact underlying mechanism remains unclear, it is believed that calcium overload, primarily mediated by cAMP-induced effects on calcium influx, storage, and spontaneous release, plays a significant role in most cases. The elevated levels of catecholamines lead to increased myocardial oxygen consumption,

primarily driven by tachycardia, augmented inotropy, and heightened wall stress resulting from high blood pressure. This heightened oxygen demand can contribute to pro-arrhythmic ischemia [22].

While surgical removal remains the primary treatment modality for pheochromocytoma and paraganglioma, medical therapies have become invaluable in managing this rare neuroendocrine tumor. Alpha-beta-blockers are crucial in controlling symptoms and preparing patients for surgery.

Our report presents a rare case of paraganglioma-induced hypertensive crisis accompanied by an electrical storm, which was successfully treated by tumor removal. However, VT remains an uncommon consequence of these tumors, and understanding the mechanism of ventricular tachycardias in paraganglioma needs further research.

Conclusions

Pheochromocytomas and paragangliomas are uncommon tumors, and the intermittent nature of these symptoms often leads to a delay in the diagnosis. Therefore, if patients present with symptoms of heart failure, hypertensive crisis, and arrhythmia in the absence of any coronary or valvular heart disease, a paraganglioma-triggering catecholamine crisis should be suspected.

Pheochromocytoma that initially presents with VT in the form of the electrical storm is a very rare condition, being reported only in a few isolated cases. Our case presents the rare association of electrical storm, hypertensive crisis, and paraganglioma as a result of excessive catecholamine stimulation triggered by the paraganglioma.

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Conflict of interests

The authors declare no conflict of interest.

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