

A 'shark'-masked electrocardiogram: case report of a Tako-Tsubo syndrome

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Background

Triangular ST-segment elevation or 'shark-fin' sign has been described as a specific indicator of acute coronary occlusion and large myocardial ischaemia, translating into poorer prognosis. However, this electrocardiographic presentation has been reported in rare cases of Tako-Tsubo syndrome and associated with more severe physical stressors and neurological involvement.

Case summary

We present a rare case of a 51-year-old woman presenting with incoming epileptic attacks and concomitant pyometra. Despite controlling epilepsy with phenytoin and the surgical treatment of the infection, she developed sepsis requiring vasopressors, and thereafter sustained ventricular tachycardia and diffuse ST-segment elevation with the 'shark-fin' sign. TTC was confirmed by the documentation of normal coronary arteries and the complete recovery of wall motion abnormalities at discharge.

Discussion

Heterogeneous presentation and triggering conditions often challenge the diagnosis of Tako-Tsubo syndrome. The acknowledgement of different electrocardiographic and clinical manifestations can ease the diagnosis and the successful management of these patients, whose prognosis can be extremely severe in the acute phase, if unidentified.

Keywords

Tako Tsubo • Triangular ST-segment elevation • Myocardial infarction • Stress • Case report

Learning points

- 'Shark-fin' sign has been indicated as a negative prognostic marker in acute myocardial infarction.
- Non-ischaemic coved ST-segment elevation is uncommon, as in the present case of Tako-Tsubo syndrome.

Introduction

The Lambda-wave ST-elevation, also known as 'shark-fin pattern',¹ is a particular electrocardiographic presentation of

ST-segment elevation myocardial infarction where the QRS complex, the ST-segment and the T-wave are fused in a unique complex.

Previous studies have linked this pattern with a larger ischaemic area, ventricular arrhythmias and cardiogenic shock, therefore being associated with a more severe outcome.^{2,3}

However, other non-ischaemic causes of coved or down-sloping ST-pattern have been described, including the Brugada syndrome⁴ or Tako-Tsubo cardiomyopathy (TTC), where it represents a rare manifestation, whose prognostic impact is still undetermined.

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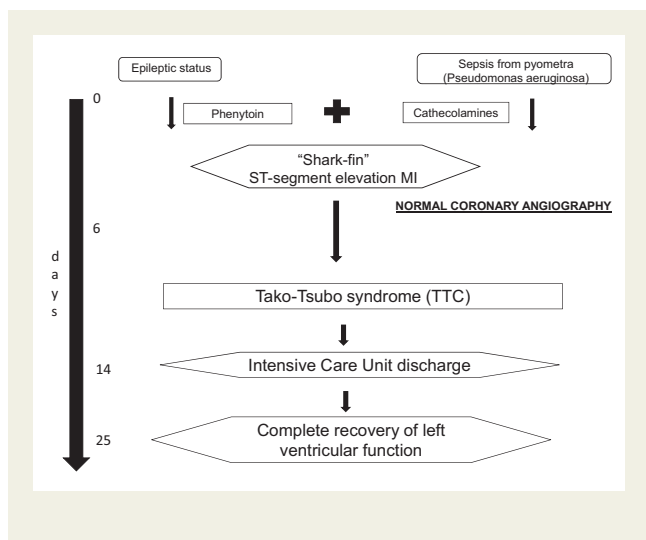
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Timeline



Case presentation

We describe the case of a 51-year-old woman that was found at home presenting general convulsions, lying under a pendulum clock.

After a bolus of diazepam 20 mg, partial vigilance of the patient was restored, and she was carried to the emergency room. Vital signs and physical examination were normal.

Her medical history included hypertension in therapy with ramipril 5 mg daily, diabetes mellitus treated with fast and long-acting insulin, personality disorder in therapy with flufenazide in depot formulation and alcohol abuse.

Laboratory exams are displayed in [Table 1](#), tests for alcohol and drugs were negative.

A whole-body computed tomography (CT) scan was negative for fractures or intracranial haemorrhage, but documented the presence of pyometra due to an extraneous body inside the uterine cavity.

Focal epileptic activity located in the left fronto-parietal portion of the brain was observed at electroencephalographic monitoring, but both infusion of levetiracetam and valproate were ineffective in controlling the incoming attacks that were finally extinguished after the continuous administration of propofol and phenytoin (500 mg in 24 h). Intubation and admission to the intensive care unit (ICU) were then needed.

For the management of the pyometra, endoscopic removal of the extraneous body (plastic plug, [Figure 1](#)) and antibiotic treatment with clindamycin and gentamicin were performed.

Thus, the hospitalization of the patient in the ICU prolonged for over 5 days. Thereafter, progressive worsening of the laboratory and haemodynamic parameters with hypotension was observed, suggesting septic shock, despite an escalation of the antibiotic therapy with anidulafungin and ceftriaxone. Norepinephrine (1 µg/kg/min) and epinephrine (1 µg/kg/min) infusion were then started.

12 h later, the patient suddenly presented a sustained ventricular tachycardia, that was resolved with amiodarone (300 mg bolus only) but immediately relapsed, the second time pulseless. DC-shock

Table 1 Patient's laboratory parameters at admission

Laboratory values	Result	Range
Haemoglobin (mmol/L)	7.01	8.69–11.17
WBC ($10^3/\mu\text{L}$)	10.6	4.00–10.00
Platelets ($10^3/\mu\text{L}$)	228	140–440
Glycaemia (mmol/L)	8.55	3.33–6.11
Creatinine ($\mu\text{mol/L}$)	96.38	50–120
Procalcitonin ($\mu\text{g/L}$)	0.1	<2
C-reactive protein (nmol/L)	447.6	<4.7
LDH (U/L)	278	135–214
CPK (U/L)	1353	26–140
CK-MB (U/L)	24	1–19
AST (U/L)	72	1–32
Arterial blood gas		
pH	7.43	7.34–7.45
pCO ₂ (mmHg)	39.8	41–51
pO ₂ (mmHg)	69.6	83–108
sO ₂ (%)	95	95–99
HCO ₃ ⁻ (mmol/L)	26.4	24–28
Lactic acid (mmol/L)	3.5	0.5–1.6
Calcium (mmol/L)	1.05	1.12–1.27
Sodium (mmol/L)	138	133–140
Potassium (mmol/L)	3.6	3.5–4.9
Chloride (mmol/L)	93	98–106



Figure 1 Abdominal computed tomography displaying the uterine foreign body and pyometra (arrow).

allowed a complete recovery of sinus rhythm without electrocardiographic abnormalities.

After about 5 h, the electrocardiogram (ECG) suddenly modified displaying a diffuse ST-segment elevation, with a triangular pattern known as the 'shark-fin' sign, without mirroring ST-segment depression ([Figure 2](#)).

The echocardiogram showed a severe reduction of left ventricular function (EF around 30%) with akinesia of the apex and periapical segments, suggestive for apical ballooning

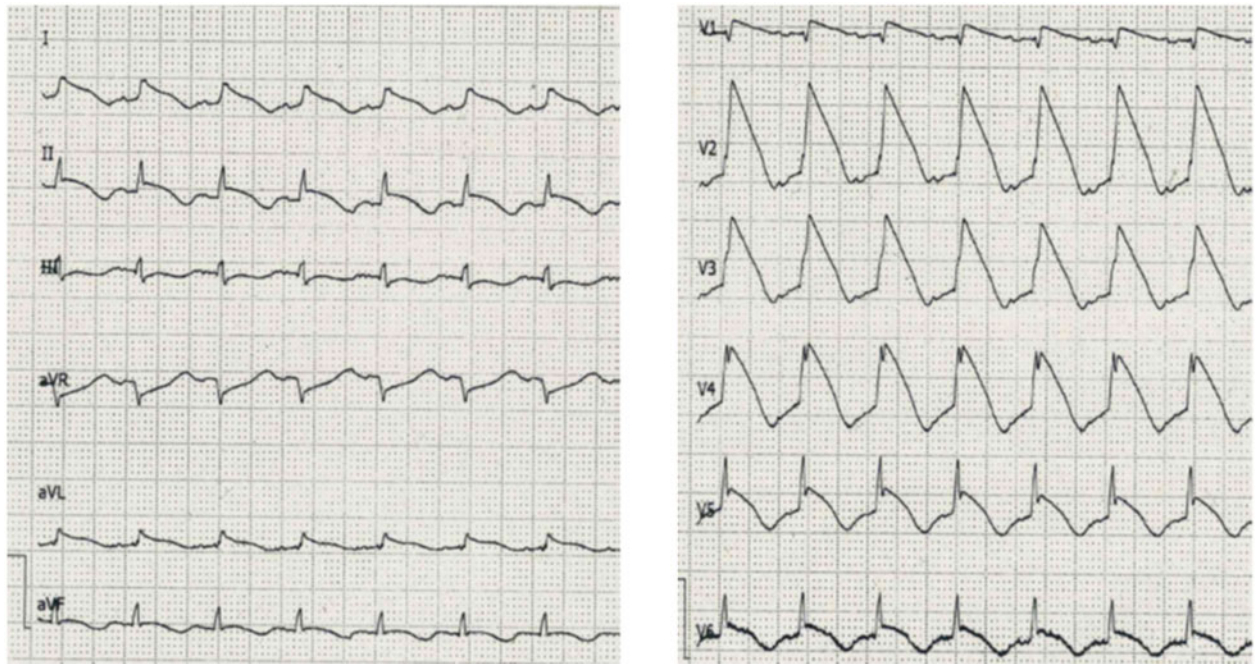


Figure 2 Electrocardiogram displaying a diffuse ST-segment elevation ('shark-fin' sign).

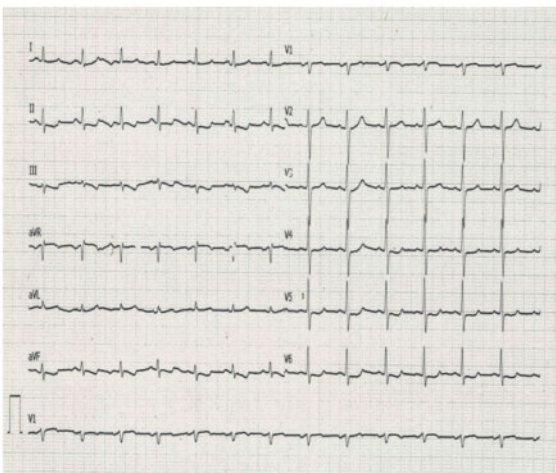


Figure 3 Baseline electrocardiogram at admission.

(Supplementary material online, Video S1). Right ventricle and valve function were normal. Diastolic function evaluation was not performed in the acute phase, being conditioned by tachycardia. Considering the cardiovascular risk factors, the alterations of the baseline ECG at admission (Figure 3, with mild antero-lateral ST depression and partially inverted T-waves) and the hemodynamic instability, coronary angiography was then performed. Coronary tree was normal but for a mild stenosis of

the mid-left anterior descending artery (Videos 1–3), suggesting TTC. Only a mild troponin elevation was observed, which remained stable with no ischaemic curve (1st determination: 0.07 ng/mL, at 6 h 0.07 ng/mL, at 12 h 0.05 ng/mL).

Multiple aetiologies were hypothesized for the TTC, including epileptic status, sepsis, the use of amines, and phenytoin myocardial toxicity.

Thus, phenytoin was substituted again with levetiracetam, without epileptic relapse, and vasopressors were progressively stopped. Single antiplatelet therapy with aspirin was administered, which was not deemed contraindicated from a neurological or gynaecological point of view, and, after the hemodynamic stabilization, beta-blockers were started and ramipril was re-introduced. Patient was extubated and discharged from ICU within 6 days.

A progressive resolution of the ST-segment elevation and recovery of the left-ventricular function were observed in the subsequent days (Figure 4A–C).

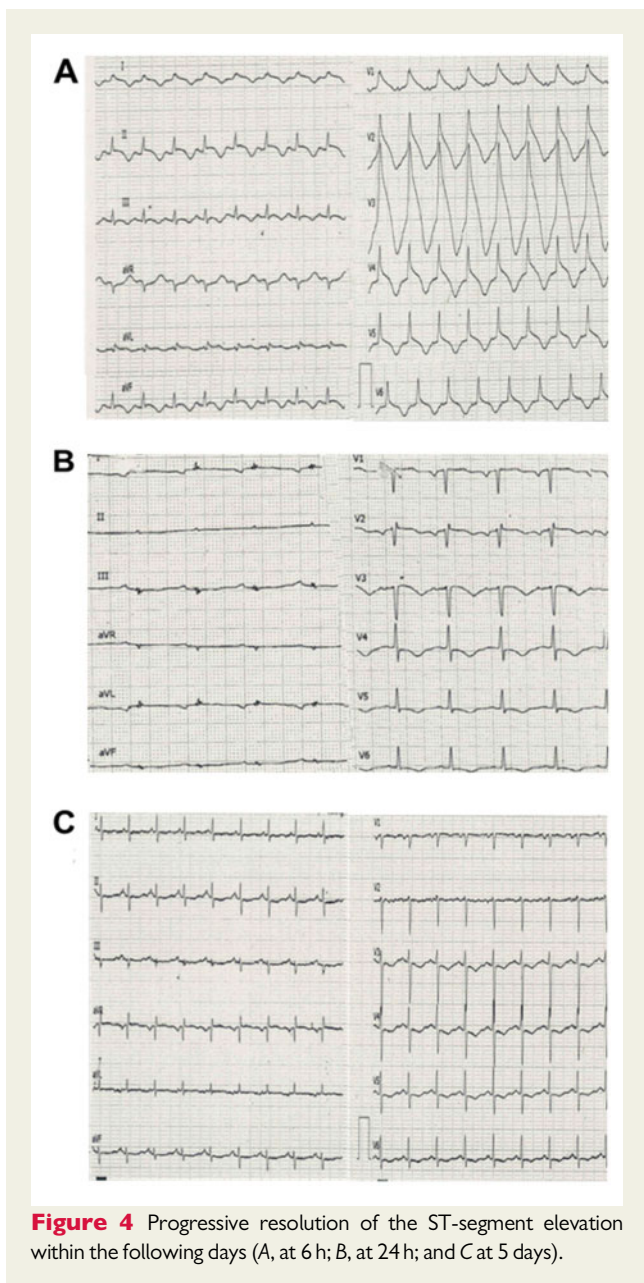
Her echocardiogram at discharge, after 25 days from the acute event, showed the complete recovery of the left ventricular function (EF 60%) with normal kinesis and no valvular dysfunction.

Therapy with aspirin, ramipril, and beta-blockers was maintained at discharge.

At 6 months follow-up, she remains asymptomatic with no further cardiovascular event.

Discussion

We describe an uncommon case of TTC presenting with the electrocardiographic 'shark-fin' sign or triangular ST-segment elevation.

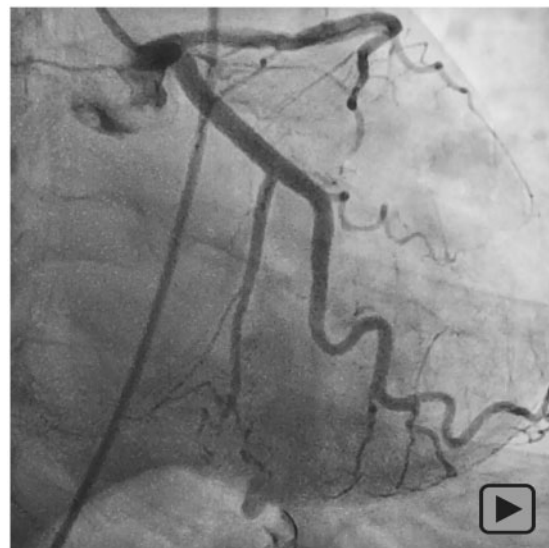


ST-segment elevation has been described in about 56% of TTC cases, although its amplitude is usually lower than in anterior ST-elevation myocardial infarction.^{5,6}

So far, the cases of TTC presenting with lambda-wave ST-segment elevation have been rarely reported.^{7,8} Vasospasm, local intermittent microvascular constriction and catecholamine-induced increase of myocardial metabolic demand can certainly be claimed for the observation of a diffuse ischaemic pattern in these patients.

In addition, transmural systolic wall tension, especially located at the apex, in the TTC could influence action potentials (the mechano-electric feedback⁹) resulting in a coved-type ST modification, like in the Brugada syndrome, mimicking the ischaemic 'shark-fin' sign.

However, the same prognostic implications have been associated to this specific ECG-pattern, either when consequent to coronary obstruction or non-ischaemic causes.



Video 1 Left coronary artery angiography (caudal projection).



Video 2 Left coronary artery angiography (cranial projection).

In a larger study, enrolling 158 consecutive subjects with TTC, Tarantino et al.¹⁰ described the lambda-wave pattern in five patients, being associated with ventricular dysfunction, in-hospital complications and long-term adverse events in 80% of the cases. Of notice, physical stressors were more commonly associated to the 'shark-fin' sign than to psychological causes (100% vs. 49%, $P = 0.04$) and an abnormal neurological status was the alteration more frequently reported before the event, suggesting an implication of autonomic dysregulation, that could favour the sympathetic activation and an abnormal response to a stressor.



Video 3 Right coronary angiography.

Similarly, in a US cohort study, TTC was associated with several acute neurological diseases, the most common being subarachnoid haemorrhage [odds ratio (OR) 11.7; 95% confidence interval (CI) 10.2–13.4], status epilepticus (OR 4.9; 95% CI 3.7–6.3), and seizures (OR 1.3; 95% CI 1.1–1.5).¹¹ In particular, 1/1000 epilepsy-related hospitalizations resulted in TTC occurrence, with a negative prognostic impact.¹²

A similar brain–heart interaction leading to TTC and ‘shark-fin’ sign presentation can be hypothesized in our patients, in consequence of the relapsing epileptic attacks, although the cardiac insult occurred several days after the main convulsive episode. Indeed, despite phenytoin infusion prevented the epileptic manifestations, we cannot exclude an alteration of the neurological autonomic regulation in our case.

Furthermore, sepsis, high-dose catecholamines and phenytoin cardiac toxicity could have played a role in the pathogenesis of a ‘multi-factorial’ TTC, further contributing to the rarity of the case. In fact, severe cardiac adverse effects of phenytoin overdose have been reported with its intravenous administration, including cardiac arrhythmia, blocks, asystole, and hypotension have been described, in consequence of the blockage of sodium channels, impairs myocardial contractility, decreases peripheral vascular resistance and depresses cardiac conduction.^{13–15} Thus, it might be argued that in our patient, phenytoin interaction with Na-channels could have favoured the appearance of a Brugada-like downsloping ST-segment elevation during TTC.

On the contrary, despite the inflammatory milieu, the diagnosis of myocarditis was considered more remote, based on the rarity of a bacterial aetiology, the non-segmental pattern of distribution and the extremely fast onset of ECG and echocardiographic alterations. Moreover, the positive evolution after the removal of the stressing triggers did not render necessary any further invasive evaluation, including endomyocardial biopsy. On the contrary, cardiac MRI (CMR) could have supported our diagnosis, although its technical

feasibility was limited in the acute phase and its diagnostic accuracy significantly decreases with progressing of time after the event.¹⁶

In addition, non-invasive coronary imaging could certainly have represented an option for ruling-out coronary disease in a case with a lower probability of coronary artery disease and increased risk of complications,¹⁷ although in our patient prolonged history of diabetes favoured an invasive approach. Moreover, coronary CT is not routinely available in our centre.

About pharmacological treatment, evidence regarding the best strategy for the management of TTC is still modest. Beta-blockers represent one of the mostly appointed therapeutic options in the acute phase, based on the pathophysiological role of catecholamines,¹⁸ although in our case, it was possible only after weaning-off inotropic support. However, despite beta-blocker therapy after hospital discharge has not demonstrated to prevent TTC recurrence, we preferred to maintain it at long-term, in association with acetylsalicylic acid (ASA), due to the documentation of coronary atherosclerosis.^{19,20} Angiotensin-converting enzyme inhibitors, in our patient, were reintroduced and maintained at discharge, accounting for their potential benefits in left ventricular remodelling and outcomes.²⁰

Indeed, TTC patients still represent a debated category of patients with acute coronary syndromes, considering the multiple aetiologies, the heterogeneous clinical manifestations and comorbidities, challenging the diagnosis and the pharmacological treatment. Therefore, additional studies dedicated to patients with TTC are certainly warranted.

Conclusion

Tako-Tsubo syndrome (TTC) represents a heterogeneous clinical condition, depending potentially on a variety of different triggers. The concomitant summation of multiple stressors in a single patient and the electrocardiographic presentation with the ‘shark-fin’ sign represent rare conditions, which are associated with a poor prognosis. We describe a case of TTC with triangular ST-segment elevation at presentation consequent to epileptic status, sepsis and phenytoin administration, that was successfully managed with a complete resolution.

Lead author biography



Monica Verdoia graduated at Università degli Studi del Piemonte Orientale “A. Avogadro” in Novara and is now Interventional Cardiologist at the Ospedale degli Infermi, Biella. She has focused research on biomarkers and genetics of atherosclerosis within the Novara Atherosclerosis Study Group. She is International Fellow the American College of Cardiology, member of

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Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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.

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