

# A rare endocrine cause of electrical storm - a case report

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## Abstract

Sheehan's syndrome, also called Simmond's syndrome, postpartum apoplexy, postpartum pituitary necrosis, and postpartum panhypopituitarism, is the name given to postpartum hypopituitarism. The syndrome is caused by an infarction in the adenohypophysis, usually precipitated by massive uterine haemorrhage and hypovolemic shock during or after childbirth. Extensive destruction of pituitary cells results in varying degree of hypopituitarism. Acute loss of adenohypophysis function can be fatal without glucocorticoid and thyroid replacement therapy and survivors will require life time hormonal replacement therapy. Most cases present in the postpartum period with lactation failure or after months to years after the delivery. In many affected women, anterior pituitary dysfunction is not diagnosed for many years. Dyselectrolytemia is one of a common presentation in Sheehan's syndrome. Herein, we report a case of a 35-year old female with a history of obstetric hysterectomy 2 years ago in view of severe postpartum haemorrhage with history of failure of lactation and three episodes of syncope since last 1½ years and now presented with polymorphic ventricular tachycardia which required DC cardioversion. She was referred as a case of long QT syndrome. On investigating further, she had hypokalaemia secondary to hypopituitarism due to Sheehan's syndrome.

## Keywords

Sheehan's syndrome • Ventricular tachycardia • Hypokalaemia • Hypopituitarism

## Learning point

- Ventricular tachycardia is not always due to a primary cardiac pathology, a wide array of other causes especially endocrinal causes should be entertained.

## Patient information

A 35-year-old female presented to the emergency department of a nearby hospital with nausea, vomiting, and giddiness since 3 days. She was dry and cold to touch and the pulse and blood pressure were not measurable. Initial assessment was remarkable for gross clinical features of hypothyroidism with hoarse voice and myxoedema. A rhythm strip (long lead II) ECG (*Figure 1*) revealed broad complex polymorphic ventricular tachycardia (PMVT). Successful cardioversion was performed with 150 J asynchronous DC shock. Baseline sinus rhythm showed a prolonged corrected QT interval (QTc) of 760 ms with T-wave inversion in V1–V6, I, avL, II, III, and avf (*Figure 2*). Post-cardioversion she had

normal pulse of good volume, force, and tension with BP of 110/70 mmHg. On auscultation she had normal S1S2 with no added sounds.

Further course in ICCU was turbulent with two recurrent episodes of haemodynamically unstable PMVT requiring cardioversion over the next 24 h. She was referred to our tertiary centre as a case of long QT syndrome. Patient gave history of normal full term vaginal delivery 2 years ago which was complicated by severe postpartum haemorrhage for which she received six units of blood and had to undergo an obstetric hysterectomy for the same. She gives a history of lactation failure for that period but otherwise she was asymptomatic for her postpartum period. However, during the past 1½ years she gives history of three episodes of syncope lasting for 5–10 s.

There was no family history for any heart disease or sudden cardiac death in her family. On gross clinical examination, she was dry and cold to touch and features of hypothyroidism with hoarse voice and myxoedema. Lab investigations as in the following table were suggestive of hypokalaemia and secondary hypothyroidism.

Channelopathies causing long QT syndrome and peripartum cardiomyopathy with secondary ventricular tachycardia were some

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differentials which were strong on the list considering her history and presentation. With this history and laboratory investigations (Table 1), MRI brain (Figure 3) was done suggestive of empty sella turcica confirming diagnosis of Sheehan's syndrome.

Cardiac MRI was done in the same setting which was normal. Patient was monitored in ICCU. Required deficit potassium of 400 meq was administered intravenously with close monitoring of serum potassium levels over 2 days. Steadily her Sr potassium levels were within normal limits in a couple of days. Thyroid replacement was done with Tb Levothyroxine 75 µg 1/2 OD for 5 days and then with 75 µg OD to continue. Steroid supplementation was given with Inj Hydrocortisone 100 mg i.v. stat followed by Tb prednisolone 5 mg OD to continue. Further ward course was stable with gradual normalisation of QTc, persistent T wave inversion in V1-V6 and improvement in LVEF with predischarge LVEF of 50%. Patient was discharged after 2 week onTb levothyroxine 75 µg OD and Tb Prednisolone 5 mg o.d. and with a plan to supplement oestrogen and progesterone on follow-up on OPD basis with endocrinology follow-up. Patient was not started on any antiarrhythmics.

EP study and coronary angiography was not performed in our patient despite being a lethal arrhythmia. The cause of PMVT due to hypokalaemia as a result of Sheehan's syndrome was striking based on laboratory investigations and imaging which was supported by an appropriate response to potassium and required hormonal supplementation.

## Timeline

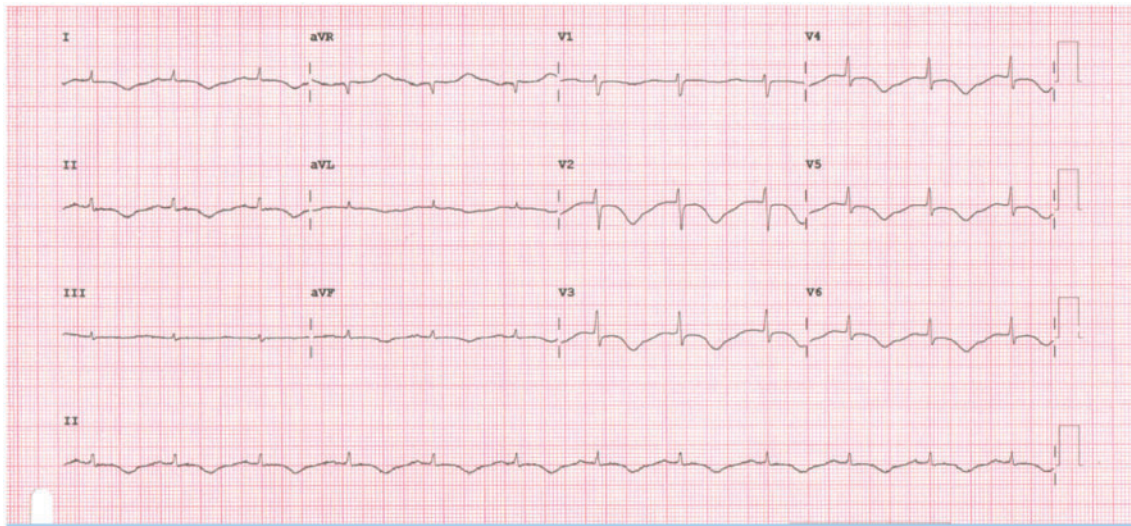
Day	Events
2 years ago	Obstetric hysterectomy due to postpartum haemorrhage with history of blood transfusion and postpartum lactation failure
Over last 1 1/2 years	History of three episodes of syncope
Day 1	Presented unstable haemodynamics with rhythm(II) strip suggestive of PMVT. Successful cardioversion with 150 J DC shock done. Baseline ECG shows sinus rhythm with QTc of 760 ms Shifted to ICCU, developed two recurrent episodes of haemodynamically unstable PMVT which required cardioversion over 24 h
Day 2	Referred as a case of long QT syndrome
Day 3	Clinical features suggestive of hypothyroidism with laboratory findings suggestive of hypokalaemia with secondary hypothyroidism.2DEchocardiography—global LV hypokinesia EF 35%
Day 4	Cardiac MRI—normalMRI brain—empty sella turcica suggestive of Sheehan's syndromeStarted on potassium supplementation and hormone replacement

## Discussion

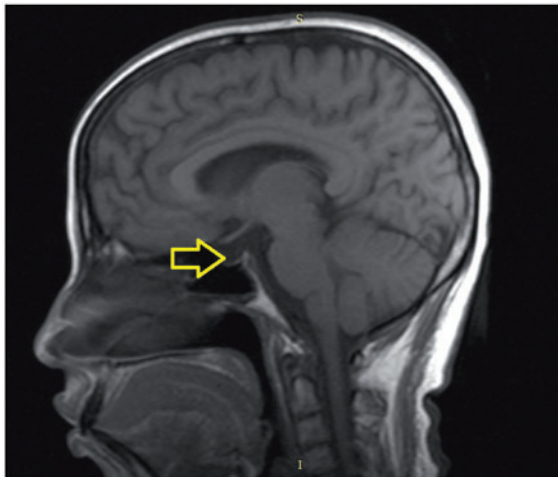
Sheehan's syndrome is a serious and rare postpartum complication and was first described in 1937 by Harold Leeming Sheehan. It was named as Simmond's disease in 1939.<sup>1</sup> Sheehan's syndrome refers to a state of variable degree of hypopituitarism due to severe postpartum haemorrhage and hypovolumic shock during or after childbirth, which results in ischaemic necrosis of anterior pituitary. The prevalence of Sheehan's syndrome is 100–200/100 000 womans.<sup>2</sup> The frequency of this syndrome is decreasing worldwide and is now one of the rare causes of hypopituitarism in the developed countries owing to recent advances in obstetric care. However in developing countries it is still an important and a undiagnosed cause of hypopituitarism owing to the lack of effective management of postpartum bleeding and poor obstetric care, although decreasing in frequency in recent years. During pregnancy, hyperplasia and hypertrophy of the lactotrophs leads to the enlargement of anterior pituitary, without corresponding increase in the blood supply. Secondly, the anterior pituitary gland has its blood supply from a low-pressure portal venous system. When such vulnerable pituitary is affected by hypotension or a major Haemorrhage during peripartum period, it can lead to ischemia of affected pituitary regions thereby giving rise to necrosis. The posterior pituitary gland is generally unaffected because of its direct arterial supply. The cause for the interruption in the blood flow, however, is not clear. Potential mechanisms considered for the same are arterial thrombosis similar to that seen in stroke, arterial spasm due to severe hypotension as a result of massive uterine bleeding, or compression of pituitary vessels due to a very relatively small sella turcica volume associated with enlargement of the pituitary during



**Figure 1** Long lead II (rhythm strip) showing Broad complex polymorphic ventricular tachycardia (PMVT).



**Figure 2** Baseline 12-lead ECG showing sinus rhythm with prolonged QT interval (QTc) of 760 ms with T-wave inversion in V1–V6, avL, II, III, and avf.



**Figure 3** MRI brain with T1 weighted saggital view showing empty sella.

pregnancy.<sup>3</sup> Also, autoantibodies detected in many patients against the pituitary gland have been considered as a contributing factor in the etiopathogenesis of Sheehan's syndrome.<sup>4</sup> Very few patients with Sheehan's syndrome develop acute postpartum hypopituitarism after postpartum haemorrhage. The most common clinical scenario is a woman presenting years later with amenorrhea, with the diagnosis of Sheehan's syndrome being made retrospectively. Though, it is important to emphasize that Sheehan's syndrome is a neurological and endocrinological emergency and is potentially lethal.<sup>5</sup> For patients with Sheehan's syndrome, studied by Gei-Guardia *et al.*,<sup>6</sup> the period of time between the postpartum episode of bleeding and the diagnosis of Sheehan's syndrome is 13 years.<sup>5</sup> Characteristic manifestations

**Table I** Table showing the required hormonal and electrolyte work up done

Laboratory parameters	Observed values	Reference range
Serum creatinine	0.78 mg/dL	0.35–1.25 mg/dL
Serum sodium	133 mmol/L	135–146 mmol/L
<b>Serum potassium</b>	<b>2.6 mmol/L</b>	3.6–5.3 mmol/L
Serum phosphorus	2.7 mg/dL	2.5–4.7 mg/dL
Serum calcium	8.9 mg/dL	8.5–10.3 mg/dL
Serum magnesium	1.96 mg/dL	1.32–2.44 mg/dL
<b>Serum free T4</b>	<b>0.17 ng/dL</b>	0.70–1.80 ng/dL
<b>Serum free T3</b>	<b>0.9 pg/mL</b>	1.70–4.20 pg/mL
<b>Serum TSH</b>	<b>0.10 uIU/mL</b>	0.24–5.50 uIU/mL
<b>Serum cortisol</b>	<b>3.81 µg/dL</b>	4.3–22.40 µg/dL
FSH	7.10 mIU/mL	1.6–10.2 mIU/mL
LH	3.82 mIU/mL	2.40–13.8 mIU/mL

Bold values indicates significant values in the patient.

include lactation failure or failure to resume menses, asthenia and weakness, genital and axillary hair loss, fine wrinkles around the eyes and lips, dry skin, signs of premature aging, hypopigmentation, and other evidence of hypopituitarism. The presence of postpartum lactation or the absence of amenorrhea; however, does not rule out the diagnosis. It can present, uncommonly, acutely with circulatory collapse, severe hyponatremia, hypoglycaemia, diabetes insipidus, psychosis, or congestive cardiac failure.<sup>7–13</sup> The extent of dysfunction of anterior pituitary dysfunction varies in different series. The most common involvement was the secretion of growth hormone and prolactin seen in 90–100%, while deficiencies in cortisol secretion, gonadotropin, and thyroid stimulating hormone ranged from 50% to 100%. Before clinical manifestations become evident at least 75% of pituitary must be destroyed. In Sheehan's syndrome growth

hormone deficiency is very common because somatotrophs are located in the lateral and lower regions of the pituitary gland and are most likely to be damaged by ischaemic necrosis of the pituitary.<sup>14–18</sup> Cardiac involvement in Sheehan's syndrome is rare. Sheehan's syndrome may present as dyselectrolytaemia with hyponatremia seen in 59.0%; hypokalaemia in 26.9%, hypocalcaemia in 35.9%, hypomagnesaemia in 47.4%, and hypophosphatemia in 23.1%.<sup>19</sup> Sudden unexpected death<sup>20</sup> and cardiovascular complications of hypopituitarism like dilated cardiomyopathy<sup>21</sup> and congestive heart failure<sup>8</sup> have been described previously but reports of lethal cardiac arrhythmia are very rare<sup>22</sup> as are electrocardiographic changes in hypopituitarism.<sup>23,24</sup> The electrocardiographic changes that are considered to be associated with hypopituitarism are QT prolongation, giant T inversion, and ST changes. The exact cause of electrocardiographic changes in such cases is not defined but hypoglycaemia, catecholamine surge secondary to hypoglycaemia and hypomagnesaemia, hypokalaemia are considered as possible causes.<sup>20</sup> Hypokalaemia-induced arrhythmogenicity is attributed to prolonged ventricular repolarization, slowed conduction, and abnormal pacemaker activity. The prolongation of ventricular repolarization in hypokalaemic setting is caused by inhibition of outward potassium currents and often associated with increased propensity for early afterdepolarizations. Slowed conduction is attributed to membrane hyperpolarization and increased excitation threshold. Abnormal pacemaker activity is attributed to increased slope of diastolic depolarization in Purkinje fibres, as well as delayed afterdepolarizations caused by Ca<sup>2+</sup> overload secondary to inhibition of Na<sup>+</sup> –K<sup>+</sup> pump and stimulation of the reverse mode of the Na<sup>+</sup> –Ca<sup>2+</sup> exchange. In hypokalaemic heart preparations, the prolongation of action potential may be associated with shortening of effective refractory period, thus increasing the propensity for ventricular re-excitation over late phase of repolarization.<sup>25</sup> The diagnosis of Sheehan's syndrome is based on a suggestive obstetric history, the features of hormone deficiency and decreased basal hormone levels (freeT3, free T4, TSH, cortisol, ACTH, FSH, LH, oestrogen, prolactin, and insulin-like growth factor-1). Findings on magnetic resonance imaging characterize Sheehan's syndrome and provide early confirmation of the clinical diagnosis. The diagnostic radiological finding of Sheehan's syndrome is the image of an empty sella (around 70% of patients) or partially empty sella (30%). Time-dependent evolution of the findings on magnetic resonance imaging in Sheehan's syndrome begins acutely with non-haemorrhagic changes in the signal intensity consistent with central infarction, along with a heterogeneous central and peripheral enhancement in an enlarged pituitary gland. These changes are due to patchy central ischaemic necrosis in an enlarged gland and are followed by pituitary gland atrophy and an empty sella.<sup>26</sup> Treatment involves lifelong hormone replacement therapy, and it is essential to replace the hormones that the pituitary gland fails to produce. Hypothyroidism and adrenal insufficiency have been reported to result in ventricular arrhythmias and, with appropriate replacement of deficient hormones, reversal has been seen to be delayed or immediate.<sup>27–29</sup>

## Conclusion

This case demonstrates an uncommon cause of electrical storm in an eminently correctable backdrop of dyselectrolytemia in

Sheehan syndrome. Sudden death and cardiovascular complications of hypopituitarism like dilated cardiomyopathy and heart failure have been described but reports of lethal cardiac arrhythmia are very rare as are electrocardiographic changes in hypopituitarism. The electrocardiographic changes considered to be associated with hypopituitarism are QT prolongation, giant T-wave inversion, and ST changes. Hypoglycaemia with its associated catecholamine surge, hypomagnesaemia, and hypokalaemia are speculated as possible causes.

**Consent:** Informed consent was obtained from this patient for publication of this case history and associated images in line with COPE recommendations.

**Conflict of interest:** none declared.

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