

Case Report

Sheehans syndrome: the hidden nemesis

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ABSTRACT

Sheehan's syndrome is the name given to postpartum hypopituitarism, usually precipitated by massive uterine haemorrhage and hypovolemic shock during or after childbirth resulting into pituitary injury in varying degrees. Most cases are diagnosed very late in life and the first presentation may be a life-threatening situation like shock or cardiorespiratory collapse. This hidden nemesis in women compromises their quality of life and may result into mortality if not suspected early. In this article we present 3 patients who presented in the ICU in a state of complete cardio-respiratory collapse and later discharged in full health.

Keywords: Adrenal crisis, Delayed diagnosis, Sheehans syndrome

INTRODUCTION

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.^{1,2} In developing countries it is an important and a undiagnosed cause of hypopituitarism owing to the lack of effective management of postpartum bleeding and poor obstetric care. When 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. Potential mechanisms considered for the 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 pregnancy.³ Also, autoantibodies detected in many patients against the pituitary gland have

been considered as a contributing factor in the etiopathogenesis of Sheehan's syndrome.⁴ 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.⁵ For patients with Sheehan's syndrome, studied by Gei-Guardia et al, the period of time between the postpartum episode of bleeding and the diagnosis of Sheehan's syndrome is 13 years.⁵⁻⁷ Characteristic manifestations 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. It can present with hyponatremia seen in 59.0%; hypokalaemia in 26.9%, hypocalcaemia in 35.9%, hypomagnesemia in 47.4%, and hypophosphatemia in 23.1%.^{7,8} Sudden unexpected death and cardiovascular complications of hypopituitarism like dilated cardiomyopathy and congestive heart failure have been described previously but reports of lethal cardiac arrhythmia are very rare as are electrocardiographic

changes in hypopituitarism.⁷⁻⁹ The electrocardiographic changes that are considered to be associated with hypopituitarism are QT prolongation, giant T inversion, and ST changes.⁹⁻¹¹ The diagnosis of Sheehan's syndrome is based on a suggestive obstetric history, the features of hormone deficiency and decreased basal hormone levels. The diagnostic radiological finding of Sheehan's syndrome is the image of an empty sella (around 70% of patients) or partially empty sella (30%).¹² Treatment involves lifelong hormone replacement therapy, and it is essential to replace the hormones that the pituitary gland fails to produce.

CASE REPORT

Case 1

Among 44 year old female with a progressive history of apathy and procrastination, slowness of activity and diminishing mental facilities was presented to the trauma in an unresponsive condition. Preliminary examinations revealed a pulse less patient with the monitor showing an atrial fibrillation with a fast ventricular rate, confirmed by an ECG (Figure 1).

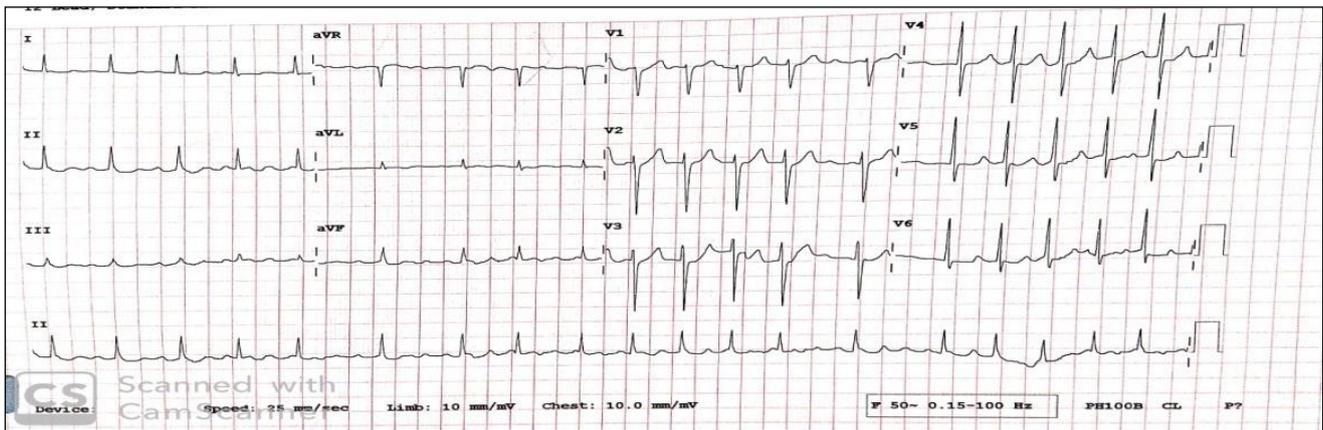


Figure 1: ECG of AF with fast ventricular rate.

The reading changed frequently to ill sustained Ventricular tachycardia and then back to atrial fibrillation without any external interventions. We assessed her initial condition and CPR was initiated as she was pulseless and simultaneous reports were sent bedside. Patient responded to two cycles of CPR but the stay in the ICU was eventful with the patient's cardiac rhythm reverting back to AF with occasional runs of unprovoked V-tach. An initial bedside Random blood sugar showed a very low reading of 41 mg/dl which was later verified by venous blood sampling. ABG surprisingly revealed metabolic alkalosis with a HCO₃ of 36 and both hyponatremia and hypokalemia. General examination showed complete loss of axillary and pubic hair, puffy face and pallor. Although she responded to CPR, she responded very poorly to fluids. Even her hypoglycaemia failed to respond optimally with dextrose. After stabilising her vitals, we corrected her electrolytes and sent her pituitary hormone assay. A detailed history from her brother revealed that the patient was apparently healthy 20 years back. During the birth of her last child, she had massive blood loss and had to be transfused (exact number of blood components not known). Later on, in the course of next 2 years she developed awkwardness in speech, actions and decision making. She stopped taking care of her child. The patient,

otherwise, a robust and educated female, slowly lost interest in her family and she was labelled with depression and then psychosis for which she was treated in various hospitals. With no improvement in her condition, she was abandoned by her family and she had taken up begging for last 10 years. During this time, she was admitted twice in a condition of collapse and was discharged with basic treatment and investigations. Her abnormal metabolic alkalosis, electrolyte imbalance and unresponsiveness to fluids and dextrose along with vivid general examination features prompted us to consider the diagnosis of Sheehan's syndrome. Laboratory investigations suggested anterior pituitary hypo functioning. Patient responded to corticosteroids and L-thyroxine. She was discharged in normal health. She has given up begging and has returned to her house after 10 years of being homeless.

Case 2

Among 52 year old female presented in a pulseless condition in the trauma with a history of being found unconscious in the kitchen. She had a history of having giddiness and fainting spells since last 2 months which was attributed to fatigue. Due to her primary presentation, she was given CPR to which she responded

immediately. She responded marginally to fluids and we could wean her off ventilator support within a short time. A detailed history suggested that the patient had a bad obstetric history with the birth of the last child followed by post-partum haemorrhage requiring an urgent hysterectomy. This was followed by a failure of lactation. Patient became progressively procrastinated and constantly complained of fatigue and listlessness. After 23 years of these events she started having fainting spells which were largely ignored because she was found in relative normal health conditions when seen by the local doctors.

Table 1: Tabulation of investigations.

Investigations	Results
Hb	7.4 gm/dl
PT/INR	10 SEC/ 0.9
Serum electrolyte - Na/K/Cl	125/3.4/1.11
Serum creatinine	0.96
PH	7.52
Free T3	2.02
Free T4	1.80
Serum TSH	3.90
Serum cortisol-AM	1.66 mcg/dl
Serum cortisol -PM	1.4 mcg/dl
Serum FSH	4.67 mIU/ml
Serum LH	1.75 mIU/ml
Serum ACTH	<5 pg/ml
Serum Prolactin	2.22
Urine-routine micro	NAD



Figure 2: Faces, general examination and radiographic findings.

Evidently, the husband of the patient had all her previous recent records of her visits to the doctor which showed a persistent feature of hypotension. A record of her last admission in a tertiary care centre showed abnormal metabolic alkalosis, hyponatremia, anemia and hypoglycaemia. A general examination showed puffy

face, spare axillary and pubic hair, extreme skin ichthyosis and pallor (Figure 2). With a positive obstetric history, suggestive additional laboratory tests (Table 1) and general examination we put forth a working diagnosis of Sheehan’s syndrome.

Pituitary hormonal assay shows anterior hypopituitarism. MRI showed empty sella turcica (Figure 2). Patient was started on corticosteroids followed by L thyroxin. She recovered fully on day 3 with improving mentation, normotensive and euglycemic. She has been seen once post discharge and now leads an active life.

DISCUSSION

Sheehan’s syndrome is a neurological and endocrinological emergency and is potentially lethal.⁵ For patients with Sheehan’s syndrome, studied by Gei-Guardia et al, the period of time between the postpartum episode of bleeding and the diagnosis of Sheehan’s syndrome is 13 years.⁵⁻⁷ In this cases the diagnosis was made almost after two decades of initiation of symptoms. Both cases although sporadic in presentation had uncanny resemblances in their first presentation. Both were middle aged females who had a long, although smothering history of chronic and easy fatiguability, worsening procrastination and a general worsening of quality of life. Lactation failure, 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 are common features.⁷ It can present with hyponatremia seen in 59.0%; hypokalaemia in 26.9%, hypocalcaemia in 35.9%, hypomagnesemia in 47.4%, and hypophosphatemia in 23.1%.^{7,8} Electrocardiographic changes hypopituitarism are rare.⁹⁻¹¹ One of our cases presented to the trauma with atrial fibrillation. A poor quality of life and deteriorating social status results into a social stigma. Although in both the cases the patients were repeatedly seen by clinicians the symptoms were dismissed as “normal” for women belonging to their age. This is more or less a social stigma now a days and it seems that it needs extreme conditions like a complete cardio-respiratory collapse to even consider the diagnosis of pituitary insufficiency. In both the cases the patients lost more than a decade of their lives before they were diagnosed and thankfully saved.

CONCLUSION

In conclusion, SS may present as acute emergency or as chronic ailment either to obstetrician, internist or endocrinologist because of varied presenting features. Moreover, the clinical features of SS are often subtle, leading to delay in diagnosis. History of PPH, lactational failure and amenorrhea are important clues.

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