Sheehans syndrome with reversible cardiomyopathy

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Abstract

Introduction
Diagnosing hypopituitarism resulting from Sheehans syndrome in the early postpartum period is a challenging clinical condition. Earlier diagnosis and initiation of appropriate hormone replacement reverses the clinical condition. Sheehans syndrome has various clinical presentations from partial to panhypopituitarism. It is usually diagnosed very late. Acute presentation and diagnosis of Sheehan’s syndrome is very rare. High degree of suspicion is required to diagnose Sheehan in the immediate postpartum period.

Case report

Our patient in the immediate postpartum period presented with signs of cardiac failure and left ventricular systolic dysfunction. She was diagnosed with peripartum cardiomyopathy and treated with antifailure measures. Later she presented with secondary infertility, amenorrhea since her last child birth, absence of axillary and pubic hair, suspected of hypopituitarism due to Sheehans syndrome, which was confirmed biochemically and by imaging of pituitary. After initiating the hormonal replacement her left ventricular function improved.

Conclusion
In any patient with intra or postpartum cardiac circulatory failure, hypopituitarism secondary to Sheehan syndrome should be ruled out.

Introduction
Sheehans syndrome is caused by acute pituitary necrosis following postpartum haemorrhage.

It presents with lactation failure, amenorrhea, secondary infertility. Predominantly it is diagnosed very late. Acute presentation of Sheehan could be in the form of hyponatremia (hypocortisolemia), hypoglycaemia, and cardiac failure. Sheehan syndrome presenting with cardiac failure is very rarely reported. Here we report a case of Sheehan syndrome with cardiomyopathy which normalised following hormone replacement.

Case report

31 year old lady presented to our outpatient department with the history of secondary amenorrhea for 5 years since her last delivery. She delivered a live IUGR baby by caesarian section. Intraoperatively she was desaturated, intubated and shifted to the Intensive care unit. She had persistent breathlessness with signs of cardiac failure (Figure 1). At that time her echocardiography showed left ventricular dysfunction, global hypokinesia with ejection fraction of 44%. She was diagnosed with peripartum cardiomyopathy and she was treated with antifailure medications and improved. One unit of blood was transfused post operatively. She was discharged with Tab.lasix, digoxin, ramipril and carvedilol. Following this delivery she did not breast feed her baby.

As she wanted another child, she was evaluated and found that she has been amenorrheic for the last 5 years. On examination, her blood pressure was 80/40 mmHg (sitting), absence of axillary and pubic hair, atrophy of breast, normal fundus and no goitre.

Her investigation, shown in table 1, revealed central hypothyroidism of low free T4, borderline elevated TSH (FT4-0.4ng/dl, TSH-10.23μIU/ml) and hypocortisolemia. With the clinical features of postpartum lactation failure, amenorrhea, requirement of blood transfusion in previous delivery, hypotension and bio chemical evidence of partial hypopituitarism, sheehans syndrome was considered.

Her echocardiography showed persistent left ventricular systolic dysfunction. MRI of pituitary gland showed small sized pituitary with CSF filled sella (Figure 2). She was started on low dose thyroxine and glucocorticoid replacement.

All cardiac drugs were continued. After eight months of hormonal replacement her total T4, free T4 was normalised (T4- 9.3 μg%, Free T4-1.69 ng%). She underwent repeat echocardiography which showed normal left ventricular systolic function with EF-54% (Figure 3).

Discussion

Acute pituitary necrosis following postpartum haemorrhage has been first described by Sheehan in 1939. During pregnancy the pituitary gland is very much enlarged and there is increased risk of haemorrhage and infarction following acute insult. Sheehan syndrome is predominantly diagnosed during the later part of life with features related to deficient pituitary function. In our patient, the diagnosis was made in the early postpartum period.

Figure 1: x-ray chest showing signs of cardiac failure.
hormones, hypothyroidism, amenorrhea, hypocortisolemic crisis and GH deficiency. It can be either pan or partial hypopituitarism.

Sheehan’s syndrome can present with secondary amenorrhea, infertility, lactation failure, loss of libido, psychiatric disturbance, change of body composition, fatigability and diabetes insipidus. Haematologic manifestations of sheehans are anaemia, pancytopenia, acquired factor VIII and von Willebrand factor (aFVIII–VWF) deficiency which are reversible with hormone replacement. Hypocotisolemia (low Na), hypoglycaemia, features of diabetes insipidus are the acute presentation of Sheehan’s syndrome.

Sheehan presenting with cardiac failure and its reversibility with hormone replacement is one of the rarest acute manifestations.

Other endocrine causes of cardiomyopathy are hypothyroidism, hypoparathyroidism, acromegaly, GH deficiency, hyperthyroidism, pheochromacytoma and carcinoid syndrome.

Hypothyroidism both subclinical and overt state is associated with structural and functional cardiovascular alteration with predominant impairment of left ventricular function more in diastole. Echocardiographic features of hypothyroidism are pericardial effusion, decrease in wall thickness and cardiac output. Long standing untreated congenital hypothyroidism can present with cardiomyopathy, which reverse back to normal after hormonal replacement.

Wani et al. has reported cardiomyopathy secondary to hypocalcaemia and hypocortisolism as part of autoimmune polyglanular syndrome type 1 which was reverted to normal after appropriate correction.

GH deficiency also lead to subclinical left ventricular dysfunction, which is unmasked by delivery and postpartum state. It is associated with decreased left ventricular ejection fraction at rest and on effort, decreased diastolic filling at rest, and myocardial wall thickness. Congenital GH deficiency presenting as a cardiomyopathy and its reversibility with appropriate replacement therapy has been reported.

In our case cardiomopathy has improved with glucocorticoids and thyroxine, without GH replacement. It shows the role and actual need of glucocorticoids and thyroxine on cardiac function is more than growth hormone.

In our case, in the immediate postpartum period she has been diagnosed and treated as peripartum cardiomyopathy with successful measures. Even after 4 years of treatment with cardiac drugs, she had persistent left ventricular systolic dysfunction.

Peripartum cardiomyopathy is usually diagnosed with symptoms of cardiac failure in the last trimester and upto 5-6 month after delivery, without any other demonstrable cause of heart failure, absence of heart disease before pregnancy. As the cardiac function improved after hormonal replacement it confirms sheehans syndrome with reversible cardiomyopathy against peripartum cardiomyopathy. Even in the diagnosed case of peripartum cardiomyopathy, we should rule out sheehans syndrome.

Diagnosing and suspicion of sheehans syndrome in the immediate postpartum period is very rare. Patients with postpartum haemorrhage or unexplained hypotension- hypopituitarism due to acute pituitary necrosis should be suspected. Because proper hormonal replacement at that time improves the clinical condition.

Other conditions similar to Sheehan, should be considered as lymphocytic hypophysities, which can present in the postpartum period with isolated ACTH/adrenal axis involvement.

In our case the patient was evaluated for secondary amenorrhea, diagnosed as subclinical hypothyroidism [TSH-10.23(µIU/ml)]. Further detailed examination revealed hypotension, paucity of secondary sexual characters. So we suspected central
hypothyroidism, which was confirmed (FT4: 0.4ng%).

Central hypothyroidism can present as normal or borderline elevated TSH with low freeT4. This high level of TSH is due to increased sialylation (a form of glycosylation) which reduces its metabolic clearance leading to increased half-life. However, this glycosylated TSH has reduced biological activity.

Abnormal circadian rhythm and increased total TSH secretion due to increased tonic non-pulsatile TSH secretion also explains high TSH level. So at the primary physician level evaluating hypothyroidism should include both TSH and freeT4, especially if there is strong suspicion of central hypothyroidism/hypopituitarism.

**Conclusion**

Diagnosing sheehan's syndrome in the immediate postpartum period is very rare unless there is high degree of suspicion. Any patient with intra- postpartum cardiac, or circulatory failure, hypopituitarism is one of the differential diagnosis, which should be ruled out by detailed obstetric and endocrine history and examination. Appropriate diagnosis and timely replacement of hormones will normalise the cardiac function.

As chance of borderline elevated TSH can mislead us, combined TSH and freeT4 should be part of evaluation of hypothyroidism at the primary physician level.

**Consent**

Written informed consent was obtained from patient for publication of this case study and accompanying images for scientific purposes.

**References**


**Table 1: Biochemical parameters at base line before initiating treatment.**

<table>
<thead>
<tr>
<th>Hormone Levels</th>
<th>Before Treatment</th>
<th>Normal Range</th>
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<tbody>
<tr>
<td>TOTAL T4 (ug%)</td>
<td>3.5</td>
<td>4.5-12.5</td>
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<tr>
<td>FREE T4 (ug%)</td>
<td>0.48</td>
<td>0.8-2</td>
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<tr>
<td>TSH (µIU/ml)</td>
<td>10.23</td>
<td>0.3-4.5</td>
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<tr>
<td>FSH (Miu/ml)</td>
<td>4.26</td>
<td>2.8-11</td>
</tr>
<tr>
<td>LH (Miu/ml)</td>
<td>3.06</td>
<td>1.1-11</td>
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<tr>
<td>S.CORTISOL(8AM) (µg%)</td>
<td>0.69</td>
<td>5-25</td>
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<tr>
<td>IGF 1 (ng/ml)</td>
<td>97.7</td>
<td>115-307</td>
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<td>GROWTH HORMONE (ng/ml )</td>
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<td>UPTO 5</td>
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<tr>
<td>PROLACTIN (ng/ml )</td>
<td>6.96</td>
<td>1.9 -25</td>
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</tbody>
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