A Case of Sheehan Syndrome Presenting with Pancytopenia and Dilated Cardiomyopathy: A Rare Combination

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ABSTRACT

Sheehan syndrome is a rare complication of postpartum haemorrhage resulting in pituitary necrosis and panhypopituitarism. We report a case of a 52-year-old female who presented with easy fatigability, asthenia and loss of appetite for 8-10 months. She was found to have alopecia, hypotension, hypoglycaemia, pancytopenia, and a history of failure to lactate after the last childbirth (17 years back) with premature menopause. The hormone profile revealed panhypopituitarism and magnetic resonance imaging of the brain was suggestive of partially empty sella. Her echocardiogram showed dilated cardiomyopathy with an ejection fraction of 35%. A diagnosis of Sheehan syndrome presenting with pancytopenia and dilated cardiomyopathy was made. The patient was started on injectable hydrocortisone and later, on thyroid hormone supplementation after which she improved. Sheehan syndrome is responsive to hormone replacement therapy and therefore should always be ruled out in such patients.

Keywords: Sheehan Syndrome, Pancytopenia, Dilated Cardiomyopathy, Panhypopituitarism

Introduction

Sheehan syndrome is a rare complication of post-partum haemorrhage resulting in pituitary hypofunction. Pituitary increases in size during pregnancy, putting it at risk of infarction due to hypovolemic shock.¹ It may be diagnosed in the immediate postpartum when the patient has a failure of lactation and non-resolution of menstrual cycle, but more often it is diagnosed very late due to subtle symptoms and social constraints in developing countries.

Case Report

We report a case of a 52-year-old female who presented to the Medicine department with a history of easy fatiguability, decreased appetite, and asthenia for 8 to 10 months. There was no history of fever, headache, jaundice, or pain in abdomen. There was a past history of pulmonary tuberculosis treated 25 years back. There was a history of increased bleeding during her last childbirth (17 years back) requiring multiple blood transfusions. She also gave a history of inability to lactate and of premature menopause after the delivery.

On physical examination, she was conscious and oriented, with a pulse rate of 90 per minute, blood pressure of 84/56 mm Hg and respiratory rate of 18 per minute.
Her skin was dry and pale, and she had loss of axillary and pubic hair as well as alopecia (Figure 1). There was pallor without icterus or pedal oedema. The patient appeared to be older than her chronological age. Systemic examination was unremarkable. Her blood sugar levels were less than 100 on multiple occasions and blood investigations were suggestive of pancytopenia with haemoglobin of 7.2 g/dl, total leukocyte count of 3060 /cumm, and platelet count of 1.06 lakhs per cumm. Peripheral smear was suggestive of pancytopenia with normocytic normochromic anaemia. Her serum sodium was 112 mmol/L and potassium was 4 mmol/L. Her liver and kidney functions were normal. Arterial blood gas analysis revealed no abnormality. Chest X-ray and abdominal ultrasound were normal. Her electrocardiogram was suggestive of poor R wave progression and echocardiography revealed dilated cardiomyopathy with 30-35% ejection fraction.

Her hormone profile was suggestive of low serum cortisol, free T3, free T4, LH, FSH and serum prolactin. Her thyroid stimulating hormone was in the lower range of normal despite low levels of free T3 and T4. The adrenocorticotropic hormone was also in the low normal range, and glycated haemoglobin was normal. The values of the hormone profile have been provided in Table 1. Brain magnetic resonance imaging revealed partially empty sella turcica (Figures 2a and 2b).

In view of hypotension, hypoglycaemia, hyponatremia, and pancytopenia on the background of her obstetric history, a diagnosis of Sheehan syndrome was made which was supported by the hormonal profile and radiological investigation.

The patient was managed with dextrose solution and inotropes initially and then was started on injectable hydrocortisone. Later, she was also started on thyroxine tablets based on her body weight. In due course of time, the patient was shifted to oral prednisolone in the replacement dose. She showed clinical improvement and was discharged on hormone replacement.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Patient’s Value</th>
<th>Normal Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glycated haemoglobin</td>
<td>5.6%</td>
<td>4-5.6%</td>
</tr>
<tr>
<td>Serum cortisol</td>
<td>27.75 nmol/L</td>
<td>123-626 nmol/L</td>
</tr>
<tr>
<td>Adrenocorticotropic hormone</td>
<td>27.8 pg/mL</td>
<td>7.2-63.3 pg/mL</td>
</tr>
<tr>
<td>Thyroid-stimulating hormone</td>
<td>1.99 mIU/L</td>
<td>0.465-4.680 mIU/L</td>
</tr>
<tr>
<td>Free triiodothyronine</td>
<td>0.77 pmol/L</td>
<td>4.25-8.10 pmol/L</td>
</tr>
<tr>
<td>Free thyroxine</td>
<td>0.88 pmol/L</td>
<td>10.0-28.20 pmol/L</td>
</tr>
<tr>
<td>Follicle-stimulating hormone</td>
<td>5.84 mIU/L</td>
<td>23-116.3 mIU/L</td>
</tr>
<tr>
<td>Leuteinising hormone</td>
<td>2.22 mIU/L</td>
<td>15.9-54.0 mIU/L</td>
</tr>
<tr>
<td>Serum prolactin</td>
<td>&lt; 30 mIU/L</td>
<td>64-395 mIU/L</td>
</tr>
</tbody>
</table>

Figure 2(a,b). Sagittal Section of Gadolinium-enhanced Magnetic Resonance Imaging of Brain Showing Partially Empty Sella Turcica
Discussion

Panhypopituitarism is a condition where there is a deficiency of all anterior pituitary gland hormones. Any pathological state involving the hypothalamus or pituitary gland can result in this condition. Conditions like tuberculous meningitis, stroke, and traumatic brain injury along with tumours like craniopharyngiomas as well as metastasis from lung and breast cancers can all affect the hypothalamus. Infarction, infections, pituitary surgery, radiation therapy of pituitary adenoma, and pituitary adenomas are a few diseases of the pituitary gland.3

Sheehan syndrome or postpartum hypopituitarism is a rare cause of panhypopituitarism. This diagnosis is made very late in most cases. The delay in diagnosis ranges from a year to almost 20-25 years in various studies. The average delay in diagnosis in multiple studies has been 9.0 ± 9.7 years.3 In our case, it was 17 years after the last pregnancy. The disease typically presents in the early postpartum period as failure to lactate and failure to resume menses. The features of hypothyroidism, gonadotropin deficiency, and acute adrenal insufficiency present later on.4 There are case reports on patients presenting with hypoglycaemia years after the last pregnancy and being diagnosed with Sheehan syndrome.5 In most cases, there is a history of asthenia and premature wrinkling of the perioral and periorbital areas. In a study conducted in the University of China on patients diagnosed with Sheehan syndrome, the presenting features were amenorrhea (82.5%), agalactia (74.2%), loss of axillary and pubic hair (85.6%), hypothyroidism (82.8%), cortisol deficiency (76.6%), insufficient adrenocorticotropic hormone (51.4%), hypo-prolactinemia (57.3%), hyponatremia (33.7%), hypoglycaemia (26.4%), and 74% of patients had an empty sella.6 Our patient also presented with subtle symptoms like easy fatigability, asthenia, alopecia, amenorrhea, and failure to lactate after the last childbirth. She was in a state of adrenal crisis with hypotension, hypoglycaemia, and hyponatremia. Further investigations showed pan-hypopituitarism with hypothyroidism and cortisol deficiency.

Most cases present with loss of axillary and pubic hair but there are a few case reports where patients have also presented with alopecia universalis, a severe form of alopecia areata.7 Our patient also had a loss of scalp hair along with axillary and pubic hair.

A few cases have also reported haematological abnormalities due to unknown mechanisms, especially anaemia and pancytopenia.8 Anaemia in these cases could be explained as the effect of pituitary hormone deficiencies and lower oxygen demands, resulting in decreased production of erythropoietin. The mechanism of pancytopenia is unknown due to the rarity of reported cases and a lack of studies, but in the cases studied, other causes were ruled out and the patients responded to hormone replacement.9 Our patient also had pancytopenia with normocytic normochromic anaemia and her parameters showed a response within a week of starting hormone replacement therapy.

Cardiac disorders including dilated cardiomyopathy have been previously reported in a few cases of Sheehan syndrome. The likely causes may be multifactorial including hypothyroidism, growth hormone, and cortisol deficiency. These cases showed a response of dilated cardiomyopathy to hormone replacement therapy.10 Our case also had dilated cardiomyopathy which we attributed to Sheehan syndrome.

Our patient had a good response to steroid and thyroid hormone replacement and is continuing with her normal lifestyle and work.

Conclusion

It often requires a sharp clinical acumen to diagnose Sheehan syndrome. Despite the delay in diagnosis, this disease has shown a good response to hormone replacement therapy which considerably improves the patient’s outcome. This case emphasises the need to take a detailed history, especially obstetric and menstrual history, and corroboration with clinical presentation.

Conflict of Interest: None

