An Interesting case of Partial Sheehan's Syndrome with Primary Hypothyroidism

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ABSTRACT

Introduction: Sheehan's syndrome (SS) is a state of hypopituitarism occurring in postpartum period caused by necrosis of the pituitary gland. Shock resulting from severe haemorrhage in the peripartum or postpartum period leads to pituitary necrosis. Sheehan's syndrome often evolves slowly and hence is diagnosed late. Patients with Sheehan's Syndrome can have varying levels of hormone deficiency, with secondary hypothyroidism. But here, we report a case of partial Sheehan's syndrome with Primary Hypothyroidism which is not common.

Case Report: 35 years old female patient with a history of postpartum hemorrhage and with primary hypothyroidism presented with features of hypopituitarism. Investigations revealed partial loss of anterior pituitary hormones. She was started on thyroxine and hydrocortisone supplementation.

Conclusion: Sheehan's syndrome can be partial or complete. Patients showing features of hypopituitarism at any age with a history of hemorrhage during childbirth must be investigated for Sheehan's syndrome. Prompt treatment prevents mortality and morbidity of the patients.

Keywords: Hypopituitarism, Hypothyroidism, Sheehan's Syndrome

INTRODUCTION

Sheehan's syndrome is the hypopituitarism resulting from an infarct of the pituitary gland following postpartum haemorrhage or shock. Injury to the anterior pituitary causes deficiency in thyroid, adreno-cortical, and gonadal functions (figure 1). Very rarely pituitary infarction occurs with diabetic vasculitis, sickle cell anaemia, and idiopathic disease (most often called in these conditions as the Simmonds syndrome).[1] Harold Leeming Sheehan, an English pathologist, in 1937 reported Sheehan a series of cases of hypopituitarism in the pregnant patients following severe hypotension due to massive peripartum hemorrhage which led to pituitary gland infarction.

Anterior pituitary necrosis was seen in autopsy of many of the reported cases. A case of diabetes insipidus has shown posterior pituitary damage in postmortem examination.[2] Incidence is estimated at 1 to 2 :10,000 pregnancies in the 1960s. Sheehan's syndrome is found to be rare in developed countries because of advanced obstetric care whereas it is still frequent in developing and underdeveloped countries.

The syndrome shows features of hypopituitarism such as loss of lactation, amenorrhea, genital and axillary hair loss, wrinkled and dry skin. In emergency cases, it presents with circulatory failure, severe hyponatremia, diabetes insipidus, hypoglycemia, and congestive heart failure.[3] Sheehan's Syndrome can be partial or complete with variable levels of hormone deficiency presenting at any stage. Sheehan's Syndrome often develops slowly and hence is diagnosed late. Timely intervention during the intrapartum or postpartum period can reduce the incidence of Sheehan's syndrome. Early diagnosis and prompt treatment can reduce the morbidity and mortality

CASE STUDY

Mrs. K, a 35 years old female presented with complaints of slurring of speech and secondary amenorrhea for 10 years followed by weight loss, easy fatiguability and generalized weakness for 2 years. She had history of hypothyroidism for 11 years and was not on any medications.

On Examination, she was thin built with dry and wrinkled skin, pale looking, alopecia with receding hair line, loss of axillary and pubic hair was present (figure 2, 3). Voice was hoarse. Breast atrophy present. Pulse rate- 74/min, BP- 90/60 mmHg with postural hypotension. CNS examination revealed diminished reflexes, B/L plantar – no response. Other systems were normal.

On meticulous query of her obstetric history, it was known that she had postpartum hemorrhage after her second delivery followed by lactation failure after 2 weeks postpartum. She developed amenorrhea and slurring of speech since then.

Investigations

Her complete blood count showed anemia (Hb- 7g%, PCV- 21%) with ESR- 55mm and other blood counts being normal. Random blood sugar was 85 mg/dL. Urine routine test, Renal function tests, serum electrolytes, Ultrasonogram of abdomen were normal. Viral markers were negative. Her thyroid profile was suggestive of primary hypothyroidism (fT3 - 0.46 pg/mL, fT4- 2.58 pmol/L, TSH- 8.64 uIU/mL). Thyroid antibodies: Anti-thyroglobulin (33.72 IU/mL) and Anti-thyroid peroxidase (178.13 IU/mL) were elevated confirming primary hypothyroidism. Growth hormone

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Biochemical marker	Value	Interpretation
Serum cortisol	2.7 ug/dL	Low
FSH	6.14 mIU/mL	Low Normal
LH	1.80 mIU/mL	Low Normal
Growth Hormone	Not detected	Absent
ACTH	53.53 pg/mL	Normal
Prolactin	1.73 ng/mL	Low
TSH	8.64 uIU/mL	High
Free T3	0.46 pg/mL	Low
Free T4	2.58 pmol/L	Low
Anti Thyroperoxidase (Anti TPO) Antibody	178.13 IU/mL	High
Anti-thyroglobulin Antibody	33.72 IU/mL	High
Serum Estradiol	17.4 pg/mL	Low normal

Table-1: Biochemical profile

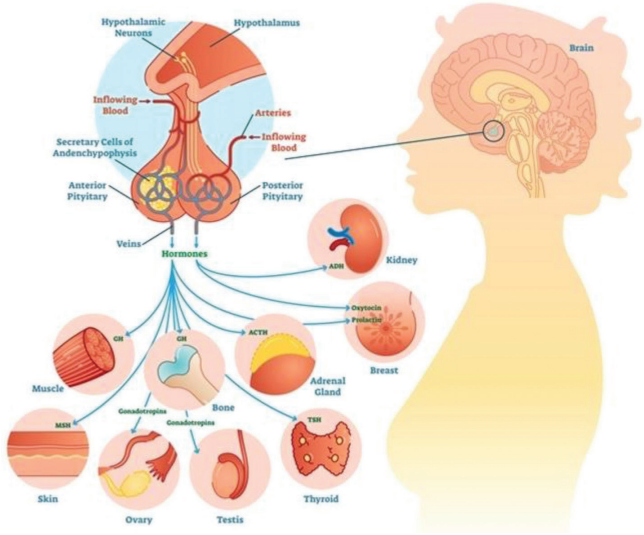

Figure-1: Pathophysiology of Sheehan's syndrome

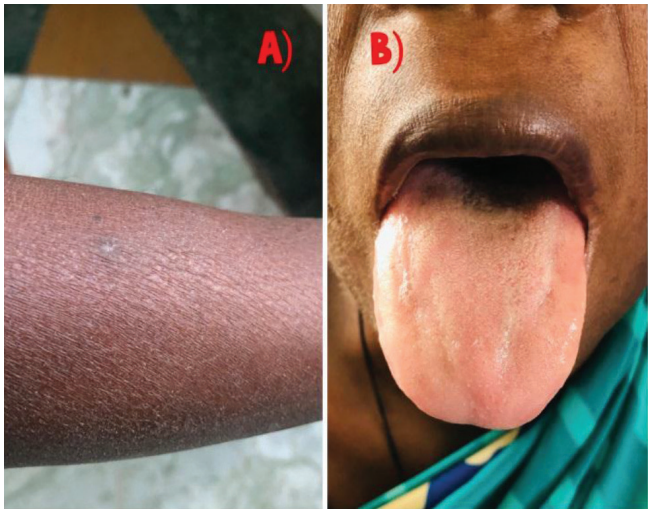

Figure-3: A) Dry, coarse skin; B) Atrophic glossitis of Iron deficiency anemia

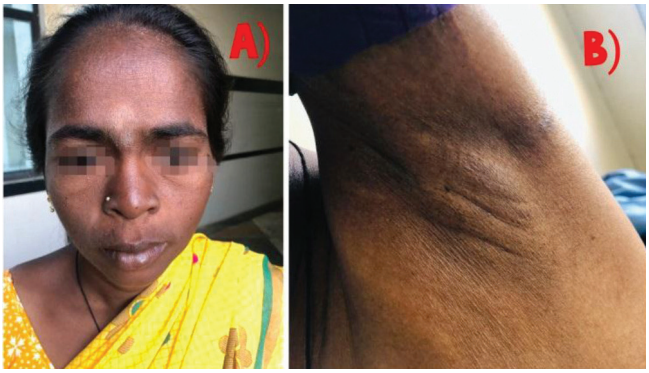

Figure-2: A) Receding hair line; B) Loss of axilla hair

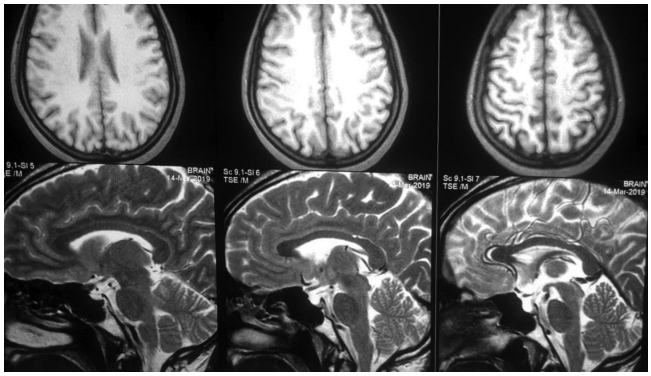

Figure-4: MRI brain showing Empty sella.

(not detected) and Prolactin (1.73 ng/mL) levels were low suggestive of hypopituitarism. LH (1.80 mIU/mL), FSH (6.14 mIU/mL) levels were in the lower limit of normal range. ACTH (53.53 pg/mL) was normal. Serum cortisol (2.7 ug/dL) was low. Serum estradiol (17.4 pg/mL) was low normal.

MRI brain showed empty sella (figure 4).

The case was diagnosed as Partial Sheehan's syndrome with primary hypothyroidism. T. Levo-thyroxine 100 mcg and Hydrocortisone supplementation was started and patient discharged. Patient is on follow up for monitoring the biochemical profile.

DISCUSSION

A recent epidemiological study from the Kashmir valley of the Indian subcontinent estimated the prevalence to be about 3% for women above 20 years of age, almost two-thirds of whom had delivered babies at home.[4]

Features of Sheehan's syndrome (SS) are lactation failure, postpartum amenorrhea, asthenia, low libido, loss of axillary and pubic hair, dry skin, thin eyebrows. The characteristic facies is described as waxy white and sallow because of low

melatonin. Thyroid may not be palpable. Untreated patients may have a low pulse rate with an often low and labile blood pressure. Atypical patients may have normal menstrual cycle and with subsequent pregnancies, symptoms may improve because of the adrenocortical hormones secreted by the placenta.[5]

The diagnosis of SS was based on the history of postpartum hemorrhage, empty sella in MRI brain, with the deficiency of one or more pituitary hormones. It has been reported that pituitary function is relatively preserved for LH, FSH, and TSH but not for prolactin, growth hormone and cortisol.[6] Jialal et al conducted a study in Africa with 10 SS patients and reported the pituitary hormone reponse to a combined intravenous insulin (0.1 unit/kg), TRH (200 mg), and GnRH (100 mg) challenge test. 100% of these women had both prolactin and GH deficiency, 90% had cortisol deficiency, 80% had TSH deficiency, 70% had LH deficiency and 40% had FSH deficiency.[7]

The basis of SS pathology has been identified as infarction and ischemic necrosis that develops due to the interruption of blood supply to the anterior pituitary. However, the cause of interruption of the blood supply is not crystal clear. The possible mechanisms considered are arterial occlusion by thrombosis similar to stroke, arterial spasm due to severe hypotension from massive haemorrhage during delivery or due to pituitary gland enlargement in pregnancy leading to compression of blood vessels of the gland.[8] Additionally, in many patients autoantibodies have also been detected against the pituitary gland. It is proposed that sequestered antigens due to tissue necrosis could trigger autoimmunity and may cause delayed hypopituitarism in these patients.[9]

In a study of 1034 hypopituitary adults, SS was the sixth most frequent cause of growth hormone deficiency, being responsible for 3.1% of cases.[10] It is imperative to consider SS in all patients presenting with cardiovascular collapse during delivery and also to look for signs of hypopituitarism. In the present case, patient survived acute blood loss and related pituitary failure for several years without any hormone replacement. Despite lactational failure and amenorrhea, indicating pituitary insufficiency dating from the postpartum period, she did not seek medical attention until slurring of speech occurred which she thought was due to CVA. This sequence of loss of pituitary trophic hormone function suggests that destruction of the pituitary gland was partial from the inception with progressive loss over time involving somatotroph, thyrotroph and corticotroph function. Such delayed presentation in patients with Sheehan's syndrome could be due to the subtlety of symptomatology and thus the associated failure to recognise hypopituitarism.

Our patient was hypothyroid previously. During all these 10 years, she gradually developed lactation failure, weakness, fatigue, anorexia, amenorrhoea. All anterior pituitary hormone panel was either low or lower limit of normal range except TSH which is raised and ACTH which was normal. Both Anti TPO antibody and Anti thyroglobulin antibody were high which indicates "Partial Sheehan's Syndrome with Primary Hypothyroidism". Pituitary autoantibody is not possible in our setting due to lack of facility. Therapy should be initiated by hormone replacement with hydrocortisone and thyroxine, and ovulation induced with follicle-stimulating hormone and luteinizing hormone.

CONCLUSION

The clinical manifestations of hypopituitarism are mostly subtle and may take years before the diagnosis is made following the inciting delivery. History of postpartum hemorrhage, lactation failure and amenorrhea are indication to the diagnosis. Early diagnosis and prompt treatment are necessary to reduce the morbidity and mortality of patients.

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