

Delayed presentation of Sheehan's syndrome: a case report

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Abstract

Sheehan's syndrome, though rare, is still one of the commonest causes of hypopituitarism in developing countries like ours. The clinical presentation is variable with abrupt or insidiously developing pituitary insufficiency after a heavy intrapartum or postpartum haemorrhage. We present an elderly lady with this syndrome who had slowly progressive panhypopituitarism 24 years after a severe haemorrhage associated with the delivery of triplets.

Sheehan's syndrome occurs as a result of ischemic necrosis of the pituitary gland caused by severe postpartum haemorrhage. Although decreasing in frequency in recent years, it is still one of the commonest causes of hypopituitarism in developing countries owing to the lack of effective management of postpartum bleeding. The clinical presentation of this syndrome is variable; the patient can present abruptly with acute hypopituitarism or insidiously with non-specific features. Its diagnosis is based on the clinical features of associated hormone deficiency, a suggestive obstetric history, laboratory finding of decreased hormone levels, and related radiological features. Its treatment requires lifelong replacement of the deficient hormones.

Case-report

A 65 year-old lady, from the remote district of Khotang, presented to our hospital with lethargy, loss of weight and appetite, and alternating diarrhoea and constipation for a duration of eight months. The patient had noticed progressively increasing weakness, skin pallor and gradual loss of weight for the past several years before she came to our hospital.

During this period she had been admitted on six separate occasions to different hospitals in Kathmandu with various diagnoses like acid peptic disease, ischemic heart disease and dyselectrolytaemia. Usually the lab investigations showed a low to low-normal blood sugar level, a low serum sodium level, and nonspecific ST-T changes in the ECG. There was no history of meningitis, head injury, tuberculosis or diabetes mellitus in the past.

On further inquiry, the patient volunteered the history of heavy vaginal bleeding with retention of placenta following home delivery of triplets 24 years back. Immediately after the event the patient had become very sick; she was unconscious for hours and bedridden for many days. All of the three triplets died

a few days after the delivery. The patient recalled that she had had no menstruation after that incidence but became pregnant again after a period of two years and gave birth to her last son.



Figs 1 & 2: The patient at presentation and after 1 month of treatment On

examination, the patient was sick looking, thin, with sunken eyes. The pulse was 88/min and blood pressure was 90/70mmHg (supine) and 70/60mmHg (with dangled legs). The skin was pale with absence of hair from skin, axillae, and groin. She was dehydrated. Examination of the lungs, heart and abdomen were normal. Examination of the central nervous system revealed generalized muscle and fat wasting and delayed relaxation of the ankle reflexes, the remaining examination was normal including fundoscopy.

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Investigations revealed a haematocrit of 31% (with normocytic normochromic red cells), total WBC was 6900/ μ L with a normal differential count, urea was 10 mg/dL, creatinine 0.8 mg/dL, Na⁺ 120 mEq/L (previously as low as 107), K⁺ 3.3 mEq/L, calculated serum osmolality 252, and urinary specific gravity 1.004.

Other investigations showed free T3 1.3 pg/mL, free T4 0.4 ng/ mL, thyroid stimulating hormone (TSH) 0.69 μ u/ mL, serum cortisol (at 9 am) 75 ng/mL (when blood sugar was 52 mg/dL), oestrogen 13.66 pg/mL, follicle stimulating hormone (FSH) 3.4 μ u/mL, leutinizing hormone (LH) 2.8 μ u/mL, prolactin 0.8 ng/mL and normal lipid profile.

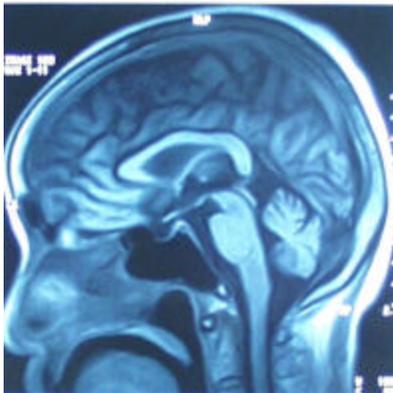


Fig. 3 MRI of the brain showing empty sella

Chest x-ray and abdominal ultrasound were normal. MRI of the brain showed a normal-sized sella that was empty with no pituitary parenchyma inside. There was no abnormality in hypothalamic, supra-sellar, or para-sellar regions in the MRI.

Discussion

Sheehan's syndrome refers to postpartum hypopituitarism as a result of pituitary necrosis occurring during severe hypotension or shock secondary to massive bleeding during or just after delivery. Though first described by HL Sheehan in 1837, it was known as Simmond's disease until 1939 when Sheehan described the disease was due to postpartum necrosis of the anterior pituitary following postpartum haemorrhage.¹ Owing to improved obstetric care and effective management of postpartum haemorrhage in more developed countries, the prevalence of Sheehan's syndrome is decreasing. However, in a developing country like ours, it is still encountered at times as postpartum bleeding is common and timely intervention is not possible in many remote and rural areas.

The underlying process leading to Sheehan's syndrome is the infarction of the physiologically enlarged pituitary gland, particularly anterior lobe, secondary to the grossly decreased blood supply during intrapartum or postpartum events. Though vasospasm, autoimmunity, small sella size, and disseminated intravascular coagulation may also have role in the development of Sheehan's syndrome, none has been conclusively proven.²

The clinical presentation of Sheehan's syndrome ranges from long-standing non-specific features such as weakness, fatigue, and anaemia to profound abrupt hypopituitarism resulting in coma and death. The mean duration between postpartum bleeding and the subsequent development of symptoms varies from 1 to 33 years;³ this period lasted more than two decades in our patient. Clinical features are the result of the deficient hormones that may be single or many, but symptoms due to GH deficiency usually appear earliest. Our patient had clinical features consistent with GH, ACTH and TSH deficiency. Although failure of postpartum menstruation due to deficiency of FSH and LH is quite common, spontaneous pregnancies have been reported.⁴ Our patient also conceived after the event; this may be possible because of continual production of gonadotrophins, a little in amount but sufficient for ovulation.

The diagnosis of Sheehan's syndrome is based on the features of hormone deficiency, a suggestive obstetric history, and decreased basal hormone levels (free T3, free T4, TSH, cortisol, ACTH, FSH, LH, oestrogen, prolactin and insulin like growth factor-1). The presenting symptoms, though vague, were suggestive of hypopituitarism in our patient. The finding of a normal pulse rate in the presence of significant postural drop was noteworthy as she had coexisting hypothyroidism and hypocortisolism. The delay in diagnosis was probably due to her vague symptoms and inadequate obstetric history. A dynamic pituitary function test like insulin tolerance test (ITT) is helpful to assess the pituitary reserve of GH and ACTH. We did not carry this test out in our patient as she already had a low blood sugar. MRI or CT of pituitary often shows an empty sella, like the one our patient had, in the late stages of the disease. Criteria have been suggested for the diagnosis of Sheehan's syndrome.²

The treatment of Sheehan's syndrome is replacement of the deficient hormones. ACTH and TSH deficiencies should be replaced with glucocorticoids and thyroxin respectively; mineralocorticoid replacement is usually not required. Our patient

showed a remarkable improvement with steroids (hydrocortisone was switched over to prednisolone) and thyroxin within a few days. Sex hormone replacement is important in premenopausal patients and GH replacement has shown improved lipid profile and quality of life in these patients.⁵

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