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**Case Report**

# A case of partial empty sella turcica syndrome: Sheehan syndrome

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## ABSTRACT

Sheehan syndrome is the clinical manifestation that results due to necrosis of pituitary gland resulting from severe postpartum haemorrhage during pregnancy and is one of the causes of empty sella, complete absence of the pituitary gland. Partial empty sella is the term given when part of the gland is visible on imaging. The hormonal deficiencies and resulting clinical syndromes are thus respectively named as complete/empty sella syndrome and partial empty sella syndrome. Incidence of empty sella especially partial empty sella are extremely rare. Also, Sheehan syndrome as a cause of the same has not been frequently observed. Here, we present a case of a young female who presented with secondary amenorrhoea postpartum and was diagnosed ultimately to have partial empty sella syndrome and partial Sheehan syndrome.

**Keywords:** Partial empty sella turcica syndrome, Sheehan syndrome, Necrosis of pituitary gland

## INTRODUCTION

Empty sella syndrome is a condition in which the pituitary gland which occupies the sella turcica, shrinks or gets flattened. As a result, it is replaced with fluid (cerebrospinal fluid) completely and when seen on the magnetic resonance imaging (MRI) scan, it looks like an empty sella.[1] When some of the pituitary gland is visible on the MRI scan it is called partial empty sella. Empty sella resulting from an underlying condition such as infarction, infection, and trauma is called secondary whereas when cause is unknown or congenital it is called primary empty sella syndrome.

Ischemic necrosis of the pituitary gland due to post-partum haemorrhage leading to hypopituitarism is known as Sheehan syndrome. It is characterized by varying degrees of anterior pituitary dysfuntion.[2] It was first described in 1937 by Harold Sheehan when he reported a series of patients with hypopituitarism following massive postpartum haemorrhage.[3] It is rare complication, is the mostly seen in developing and under-developed countries due to lack of medical resources. It is a rare complication with incidence of 10 out of every 100,000 births globally and accounts for 0.5% of all known cases of hypopituitarism in females.[4] The pituitary gland is physiologically enlarged in pregnancy as a result of hyperplasia of prolactin-secreting cells due to elevated estrogen levels. Infarction of this enlarged gland occurs due to the compression of the blood vessels supplying the gland by the enlarged gland itself or due to grossly decreased blood supply during intrapartum or postpartum events. Other probable causes causing infarction are vasospasm, autoimmunity, small sella size, and disseminated intravascular coagulation. Necrosis of pituitary gland can cause its degeneration leading to either partial or complete empty sella, seen on imaging which depends on the extent of pituitary gland degeneration. Gland degeneration leads to accumulation of cerebrospinal fluid (CSF) in the sella turcica resulting in a very small associated pituitary gland lying in the floor of the sella

(partially empty sella) or completely CSF filled with no visualized pituitary gland (completely empty sella).

The cause is unknown (idiopathic) or it occurs as a secondary disorder, due to an underlying condition. Conditions like a pituitary tumor, head trauma, or a condition known as idiopathic intracranial hypertension (also called pseudotumor cerebri), total absence of diaphragmatic sella, endocrine autoimmune diseases, obesity, type 2 diabetes, hypertension, use of certain drugs, pregnancy, post-partum pituitary necrosis (Sheehan syndrome) could promote the onset of partial or complete empty sella.

Sheehan syndrome can present either immediately after birth or several months or years following delivery. A recent epidemiological study from the Kashmir valley of the Indian subcontinent estimated prevalence to be about 3% for women above 20 years of which two-third delivered at home.[5] Also, Sheehan syndrome can be partial or complete with variable levels of hormonal deficiencies ranging from panhypopitutirism to only selective or no pituitary deficiencies. Sheehan syndrome can have varied presentations with most common symptom being agalactia and/or amenorrhea or it can result in severe crisis with circulatory collapse, severe hyponatremia, diabetes insipidus, hypoglycaemia, congestive cardiac failure, hypothyroidism lethargy or psychosis.[6,7]

Sheehan syndrome is an uncommon entity today, given the progressing global medical advancements. Post-partum haemorrhage causing partial empty sella leading to atypical presentation of Sheehan syndrome or partial Sheehan is even rare and difficult to diagnose considering the need to integrate the clinical and investigative profiles. Initial partial presentation of the syndrome could later manifest as full-blown Sheehan.

It is imperative that early diagnosis of Sheehan syndrome (with either complete/partial manifestation) is made, as undiagnosed or a delayed diagnosis is associated with long-term morbidity and mortality. Our case is one such, where an early diagnosis was made which resulted in appropriate, timely treatment of our patient.

## RESULTS

A 23-year-old female, parity 1/intrauterine fetal death 1 (P1IUFD1) who had undergone previous lower segment cesarian section (LSCS) presented to our outpatient department (OPD) with complaints of secondary amenorrhea for 4 years. Patient was affected by polio in childhood. On probing, the patient gave history of preeclampsia in 1st pregnancy resulting in intrauterine fetal death and underwent caesarean section in view of cephalopelvic disproportion. She also gave a history of blood loss during the surgical procedure and subsequent transfusion of 4 blood units in the intrapartum and postpartum period. Patient was started on breast milk suppressants because of fetal death hence she did not give history of failure of lactation. However, she presented with secondary amenorrhoea for which she had been taking oral contraceptive pills prescribed by various practitioners and had withdrawal bleeding. A proper evaluation of her symptoms was not done. She had no complains of generalized fatigue abdominal pain, nausea, vomiting, no history of weight loss, anorexia, dizziness, cold intolerance, myalgia, arthralgia, no headache, and no blurring of vision or diplopia. She had no history of head trauma or irradiation, and no history of polyuria or polydipsia. She was conscious well oriented. Her blood pressure was 110/60 mmHg, orthostatic hypotension was not observed, pulse rate 86 beats per minute, respiratory rate 16 breaths per minute, and temperature of 37.2°C. Physical examination revealed mild pallor, average built, short stature due to polio affection, no facial hair, age appropriate normal appearing pubic and axillary hair. Mild breast atrophy was seen. Systemic examination was unremarkable. On per speculum examination vaginal mucosa was seen to be dry and cervix was pulled up and on per vaginum, uterus seemed atrophied and fornices were free.

Her investigations were as follows haemoglobin (Hb) 9.4, total leucocyte count (TLC) 8k, platelets 410k, and random blood sugar was 86 mg/dl. Urine examination, liver and renal function tests, and serum electrolytes were with normal limits. Lipid profile was normal. She had mild vitamin D3 (14 ng/ml) and calcium deficiency (ionic calcium: 1 mmol/l).

Hormonal profile is given in Table 1.

**Table 1: Hormonal profile.**

| Hormone | Value | Interpret-ation |
|---|---|---|
| **FSH (mIU/ml)** | 7.73 | Normal |
| **LH (mIU/ml)** | 4.68 | Normal |
| **Prolactin (ng/ml)** | 6.49 | Normal |
| **TSH (uIU/ml)** | 2.43 | Normal |
| **AMH (ng/nl)** | 1.96 | Low normal |
| **Serum estradiol (pg/ml)** | 11 | Low |
| **Serum cortisol (mcg/ml )** | 7.39 | Low |

Serum levels of growth hormone and adrenocorticotropic hormone (ACTH) could not be done due to financial constraints and so, surrogative (both clinical and biological) markers of these hormonal deficiencies were used for interpreting. Ultrasonography of the abdomen revealed mild atrophy of uterus and ovaries. A dedicated MRI of the hypothalamus suspecting Sheehan syndrome was planned considering the history of blood loss during labour. MRI brain revealed most of the sella turcica to be filled filled with CSF. Pituitary gland appeared to be thinned out with concave upper borders, confirming partially empty sella. There was no other significant parenchymal abnormality.

The case was finally diagnosed as partial empty sella resulting in partial Sheehan's syndrome causing variable hypopituitarism. The patient was started on supplements of estrogen, progesterone and hydrocortisone along with calcium and vitamin D3 supplements and discharged. Patient was advised to follow up regularly, for monitoring endocrinological profile and to treat infertility.

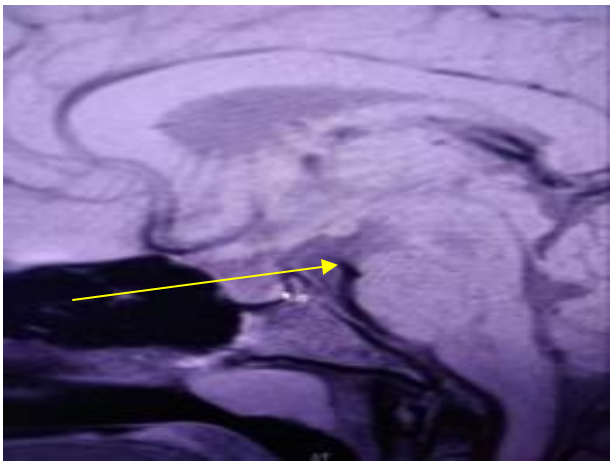

**Figure 1: Arrow points to bright posterior pituitary gland.**

## DISCUSSION

The diagnosis of Sheehan syndrome and partial empty sella in this case were mainly determined by the patient's history of blood loss during caesarean section and physical examination and confirmed by MRI revealing empty sella and low values of pituitary hormones. Failure to lactate is often a common complaint in patients with Sheehan syndrome who present immediately after deliver, but in our case, because of fetal death patient was given lactation suppressants. Many of them also report amenorrhea after delivery like our patient who had chief complaint of that of amenorrhea. Most of the women present with failure to resume menstruation and failure to lactate.[8-10] Adeno-hypophyseal ischaemic necrosis following hypoperfusion because of excessive blood loss is the most common cause of adeno-hypophyseal insufficiency resulting in Sheehan syndrome.[6] At times degeneration of pituitary is only partial and the syndrome can present in atypical and incomplete forms causing difficulty in diagnosis. The patient who came to us had suffered from partial necrosis of pituitary and hence had amenorrhea as the only symptom. The diagnosis of Sheehan syndrome can be missed in such cases and patient might be apparently asymptomatic until her body is exposed to stressful situations like surgery or infection many years after her delivery, and then she presents with adrenal crisis. Sheehan syndrome presents with varied symptoms as a result of hormonal deficiencies like lactation failure (due to prolactin deficiency), amenorrhea or genital hair loss due to gonadotropins deficiency. Corticotrophin deficiency can result in generalized fatigue, weakness, hypoglycaemia, or dizziness. Growth hormone deficiency causes fatigue, decreased quality of life, and weight loss. Patients also present with symptoms of central hypothyroidism Sheehan's syndrome can be acute or chronic based on time of presentation of symptoms after the inciting event. Acute cases have features like failure of lactation or amenorrhea. Our patient could not breastfeed following her pregnancy due to lactation failure, and became amenorrhoeic, indicating an acute presentation. But our patient did not have signs and symptoms of chronic Sheehan syndrome, such as asthenia, anorexia, weight loss, dizziness, nausea, vomiting, and abdominal pain or severe adrenal insufficiency. Our patient was diagnosed early because of amenorrhea, failure of lactation and history of blood loss during labour. Thus, with a thorough history, physical and endocrinological examination with help of imaging modalities, a rare disease was diagnosed.

All organic factors such as premature ovarian insufficiency resulting in estradiol deficiency were ruled out. Also, the patient had no menstrual complaints prior to the last pregnancy. Our case was diagnosed as a case of secondary empty sella (partial) as this patient had no hormonal deficiencies prior and inciting cause was haemorrhagic shock causing Sheehan syndrome. Treatment of our patient with variable hypopituitarism included replacement of hydrocortisone first and then replacement of estrogen with progesterone, the standard dose of hydrocortisone of 20 mg/day for an adult (15 mg every morning and 5 mg every evening) was started.[11] Replacement of growth hormone was not done as our patient was an adult and didn't have any significant manifestation of its deficiency. For our patient, we replaced her endocrinological deficiencies with available hormonal supplements considering her age, fertility desire, and financial ability. Patient was advised regular follow-up and discharged.

## CONCLUSION

The clinical manifestations of hypopituitarism secondary to partial empty sella due to pituitary necrosis are mostly subtle and may take years before the diagnosis is made following the inciting delivery. Careful reading of the history of postpartum hemorrhage, lactation failure and amenorrhea lead to the suspicion of Sheehan syndrome though, surprisingly the endocrinological profile of the patient did not reveal any profound hormonal deficiencies. The case that we studied was a rare case of empty sella with varied involvement of pituitary gland. As per clinical and radiological diagnosis, this can be considered as partial empty sella. Early diagnosis and prompt treatment are necessary to stop progression of disease and reduce the morbidity and mortality of patients. It is imperative that every woman who has history of acute blood loss during and pregnancy and labour be evaluated for Sheehan syndrome as it is associated with long-term morbidity and young age mortality. It is crucial that clinicians have a strong suspicion so that an early diagnosis can be made in routine clinical visits in order to avoid complications

arising with delayed diagnosis and failure of adequate treatment in later stages. Awareness among clinicians is essential so that such cases are not overlooked, especially in developing nations and in unfortunate circumstances even developed nations.

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