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CASE REPORT

A RARE CASE OF SHEEHAN'S SYNDROME WITH ACUTE PRESENTATION

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ABSTRACT: Sheehan's Syndrome (SS) also known as postpartum Hypopituitarism or postpartum pituitary necrosis is a condition in which hypopituitarism develops after severe bleeding during or immediate after childbirth. The incidence of Sheehan's syndrome is 0.5% of all cases of Hypopituitarism. Reporting a rare case of SS who presented post Obstetric hysterectomy within 5 months with signs of agalactia, severe hypoglycemia, hyponatremia.

KEYWORDS: Sheehan's Syndrome, Hypopituitarism, obstetric hysterectomy, agalactia.

INTRODUCTION: Sheehan's syndrome (SS) is a rare but potentially serious postpartum complication. The diagnosis can be erratic and often delayed.

SS first described by Sheehan in 1937.[1] Estimated prevalence is about 3%,[2] SS was the sixth most frequent cause of growth hormone deficiency (GHD), being responsible for 3.1% of cases[3] in a retrospective nationwide analysis in Iceland, the prevalence of SS in 2009 was estimated to be 5.1 per 100, 000 women.[4]

Sheehan's syndrome is a well-known cause of panhypopituitarism, secondary to pituitary apoplexy. Sheehan's syndrome occurs after an intra or postpartum bleeding episode characterized by severe hypotension or hemorrhagic shock results from postpartum necrosis of anterior lobe of the pituitary gland. It is one of the most common causes of hypopituitarism in developing countries.[4]

In the literature only few reports are available on the early diagnosis of acute-onset cases with severe signs and symptoms; here we report the case of a woman with an early diagnosis of Sheehan's syndrome.

CASE REPORT: A 26 years female P3L3 with history of obstetric hysterectomy done for postpartum hemorrhage 5 months back reported to OPD. She came with chief complains of weight loss, generalized weakness, continuous fatigability, loss of interest associated with mood changes predominantly of depressive type since 3 months. There was significant history of lactation failure. There was no history of evening rise fever / cough or no history suggestive of malabsorption syndrome.

She had under gone 3 lower segment cesarean section (LSCS) for contracted pelvis. At the time of last LSCS she had undergone obstetric hysterectomy for atonic postpartum hemorrhage. She had been transfused 10 bottles of whole blood and 4 fresh frozen plasma. Rest postoperative period was uneventful.

She was not having any complain except failure of lactation for 3 months post obstetric hysterectomy. After 3 months She was admitted in ICU for one episode of unconsciousness with acute hypotension, hypoglycemia. Patient received supportive management.

After this episode she was having complains of insomnia, loss of interest, depression, failure of lactation continued along with no resumption of menstruation.

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On general examination, she was lethargic, cachexic, poorly built, poorly nourished. Her body mass index was 15kg/m2. She was conscious, well oriented to time, place and person. Her vitals were stable, her skin was dry, Mild pallor was present. Her speech was slow & slurred. Thyroid swelling was not present.

On her CVS & RS examination no abnormality was detected. Her abdomen was soft, no distention, rigidity or tenderness detected, pfannesteil incision scar was healthy. On per speculum examination vaginal vault was atrophied and narrow. On per vaginum examination no abnormality detected.

In routine investigations her hemoglobin was 9gm% with peripheral blood smear has mild hypochromia, Her HIV, HbsAg were nonreactive. USG abdomen & pelvis revealed absence of uterus with normal ovaries.

Blood sugar level random was 64 mg% Her electrolytes were deranged with Sr.sodium-123 meq/L, Sr potassium- 5.3meq/L. Her liver function test, renal function test, ECG, X-ray chest were within normal limits.

On endocrinological investigations her luteinizing hormone (LH) was 0.58mIU/ml, follicle-stimulating hormone (FSH) was 1.87 mIU/ml, thyroid-stimulating hormone (TSH) was 1.25µg/dL. Free T3 was 1.14 pg/ml (1.7-4.2) and Free T4 was 0.38ng/dl (0.7-1.8). Sr estradiol – 7.74pg/ml, s. prolactin was 0.1 ng/mL Sr. cortisol- 1.05 ugm/dl. All were remarkably decreased. Adrenocorticotropic hormone (ACTH) stimulation test was done. Her 8am Pre-ACTH was 0.22 µg/dL (5-25) & 9am Post - ACTH was 1.41 µg/dL (3-5 times basal level). Normally it should increase by 25µg/dL. It was suggestive of adrenocortical insufficiency.

Brain MRI showed (gadolinium-enhanced T1-weighted MR images) a large low intensity signal in sellar lesion with enhancement of the rim suggestive of ischemic necrosis of anterior pituitary.

Clinical examination, laboratory findings and MRI imaging were suggestive of Hypogonadism, hypothyroidism, Hypoprolactinism and hypocortisolism confirming Sheehan's Syndrome.

Endocrinologist started intravenous Hydrocortisone 100mg for 2 days after that Tb. Prednisolone 10 mg OD, Tb Eltroxine 100 µg OD was given for one month. Hormonal replacement therapy (HRT) planned after 1 month. There was significant improvement in her sign and symptoms. Her thyroid function test, estrogen, LH, FSH, serum electrolytes were within normal limit.

HRT was started, Tb Premarin 0.625mg OD, Tb Eltroxin 100 µg continued. Tb Prednisolone was continued.

DISCUSSION: Sheehan's Syndrome presents with clinical signs and symptoms of deficiencies of gonadotropins, cortisol, thyroid and prolactin. Selective defects of certain pituitary functions are possible.

The primary causative factor is a widespread ischemic lesion of the pituitary gland resulting in the impairment of anterior pituitary function. The pituitary gland is particularly vulnerable to necrosis due to vascular risk that may arise during pregnancy, especially in the peripartum period. It is also believed that the acute loss of blood causes spasm in the arteries supplying the anterior pituitary leading to necrosis.[5]

Vasospasm, thrombosis and vascular compression of the hypophyseal arteries have also been described as possible causes of the syndrome. Enlargement of pituitary gland, small sellar size,

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disseminated intravascular coagulation and autoimmunity have been suggested to play a role in the pathogenesis of SS. SS is characterized by varying degrees of anterior pituitary dysfunction.[6]

Lactation failure is a very common clinical feature. Uncommonly, it can present acutely with circulatory collapse, severe hyponatremia, diabetes insipidus, hypoglycemia, congestive cardiac failure or psychosis.[7] Usually diagnosis is delayed. In a study of 60 patients, the average time between the previous obstetric event and diagnosis of SS was 13 years[8] unlike in our case.

The extent of anterior pituitary dysfunction varies in different series. The main involvement was the secretion of growth hormone (GH) and prolactin (90-100%), while deficiencies in cortisol secretion, gonadotropin and thyroid stimulating hormone (TSH) ranged from 50 to 100%[9, 10] as it is seen in our case. GH deficiency is very common in SS because somatotrophs are located in the lower and lateral regions of the pituitary gland and are most likely to be damaged by ischemic necrosis of the pituitary.[10] Gonadotrophic function may be preserved in an occasional patient and there are several reports of patients with SS who maintained regular menstrual cycles and even became pregnant spontaneously.[11] Syndrome of inappropriate antidiuretic hormone secretion (SIADH) and volume depletion are the other factors leading to hyponatremia.[13]

Whereas hypopituitarism associated with pituitary tumors often presents in an incomplete form, in Sheehan's syndrome the opposite is true. The diagnosis of Sheehan's syndrome is based on the patient's history and physical examination, laboratory investigations. Tests include hormone levels and hormone stimulation tests, CT scans, preferably MRI scans. Lab tests will reveal hypopituitarism with low thyroxin, estradiol, and cortisol levels, and with inadequately low levels of TSH, FSH, LH and ACTH. Frequently, lab tests will also reveal hyponatremia, which occurs from 33% to 69% of cases and represents the most common electrolyte disorder in Sheehan's syndrome.[12]

Hyponatremia has a late onset and can be induced by different causes, such as volume depletion, cortisol deficiency, hypothyroidism, or a syndrome characterized by inappropriate secretion of ADH.

The presence of anti-pituitary antibodies (APAs) has been demonstrated in some patients with SS, suggesting that an autoimmune pituitary process could be involved in this syndrome.[13] The main radiological finding of SS is the image of an empty sella (around 70% of patients) or partially empty sella (30%). The time-dependent evolution of the findings on magnetic resonance (MR) imaging in SS has been described and begins acutely with nonhemorrhagic changes in signal intensity consistent with central infarction, along with peripheral and heterogeneous central enhancement in an enlarged pituitary gland. The findings are consistent with patchy central ischemic necrosis in an enlarged gland and are followed by pituitary gland atrophy and an empty sella. These findings on MR imaging characterize SS and provide early confirmation of the clinical diagnosis.[14]

The goal of therapy is to replace deficient hormones. Treatment is important not only to correct endocrine abnormalities, but also to reduce mortality due to hypopituitarism.[6] In patients who have secondary hypothyroidism, hypocortisolism glucocorticoids should be replaced before the replacement of thyroid hormone. Gonadotropin deficiency and hypogonadism should be treated with a hormone replacement therapy.[6] Hormone replacement therapy, with careful follow-up of laboratory and clinical results, is the treatment of choice.

CONCLUSION: Early diagnosis and appropriate treatment of SS are necessary to reduce the morbidity and mortality of patients. Identification of all cases with postpartum hemorrhage or the deliveries

complicated with bleeding in order to identify the potential cases that may develop SS is very essential. Proper follow up of such patients is must to diagnose and treat the rare condition. The idea of reporting this case is not just rarity of this case but to give a message that Good antenatal and delivery care is a must to be able to prevent Sheehan's syndrome.

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