

Sheehan's Syndrome: New Perspective into Old, Preventable but Life-threatening Disease

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Abstract

Background: Sheehan's syndrome (SS) which is the result of necrotic damage to the anterior pituitary gland after severe postpartum bleeding is preventable but life-threatening condition for a woman. Due to postpartum hemorrhage, the hypothalamic-pituitary-gonadal axis gets disturbed and results in severe hypotension, shock, and coma. The important causes highlighted for SS are tumors of pituitary gland and vascular conditions, autoimmune diseases such as hypophysitis. The major characteristics of SS are hormonal disturbances such as prolactin, growth hormone, follicle-stimulating hormone, luteinizing hormone, and adrenocorticotropic hormone which lead to lactation failure, amenorrhea, loss of pubic and axillary hair, poor concentration, and many more to mention. It is often remained unrecognized so the diagnosis of SS is quite troublesome, but magnetic resonance imaging, computed tomography scan, and level of hormones in the blood are helpful enough. Although it is a rare condition in developed countries due to proper availability of medical treatment, yet in developing countries, it is usually undiagnosed many years.

Aim: The purpose of this article is to bring attention about this rare but crucial disease condition after postpartum hemorrhage that must be diagnosed early and the review focuses on risk factors, causes, pathophysiology, symptoms, diagnosis, and treatment of one of the critical disease conditions after noticeable postpartum bleeding.

Keywords: Sheehan's syndrome, hypopituitarism, postpartum hemorrhage, parturient, agalactorrhea, amenorrhea

INTRODUCTION

Sheehan's syndrome (SS) is also known by other names such as postpartum pituitary gland necrosis,^[1] hypopituitarism syndrome, postpartum panhypopituitarism, postpartum pituitary insufficiency, and pituitary apoplexy.^[2] SS is a serious life-threatening condition in which pituitary gland is unable to produce sufficient amount of most of its important hormones necessary to maintain essential body functions.^[2] Amenorrhea, failure to lactate, loss of pubic, and axillary hair are common symptoms faced by the woman suffering from SS. Sudden drop in blood pressure (B.P) may lead to serious consequences and sometimes woman may present with psychotic disorders.^[3]

Every woman with complicated delivery must be taken care for symptoms of SS. It was first described by the British pathologist Harold Leeming Sheehan, in 1937, who established a strong association of SS with postpartum hemorrhage (PPH) which is considered as an obstetrical emergency^[4] that means it is specifically related to the woman who experienced severe blood loss (PPH) after delivery.^[5] As stated by the WHO that every year 14 million women are affected with PPH.^[6] Postpartum bleeding or PPH is often defined as the loss of more than 500 ml or 1000 ml of blood within the first 24 h following childbirth.^[7]

Incidence and prevalence

Globally (1 of every 100,000 births) women get experience SS.^[8] Of course, actual data of incidence and prevalence are troublesome to get due to delaying in diagnosis as the disease is chronic in nature.^[9] Delay in diagnosing this condition is just due to lack of awareness and focus on puerperium period.^[9]

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RISK FACTORS

Nowadays, it is a very rare complication due to advancement of medical sciences. However, there are some predisposing factors which confronted the parturient with this serious life-threatening issue. Pregnancy can be considered as a major risk for developing SS. During pregnancy, pathophysiologic changes occurred in pituitary gland and make it more prone to develop ischemic event due to severe blood loss after delivery.^[10] According to one case analysis that those women who are diagnosed with SS have significantly higher disseminated intravascular coagulation score.^[11] Some of the high-risk conditions associated with pregnancy such as miscarriage, stillbirth, and antepartum hemorrhage can lead to severe blood loss and subsequently SS.^[12,13] A woman with multiple pregnancies,^[14] sickle cell anemia, giant cell arteritis, and any placental abnormality is also associated with it.^[14]

CAUSES

As SS is characterized by severely low B.P (hypotension) and followed by hemorrhagic shock after PPH, the important causes of SS may be considered as vasospasm, thrombosis, and vascular compression of superior hypophysial arteries, which supply the anterior pituitary with low-pressure flow.^[15] All these conditions make the pituitary gland more vulnerable to damage when woman suffers from PPH and outcome can be ischemic state of ant. pituitary gland and finally the tissue death (necrosis).^[16]

PATHOPHYSIOLOGY

Pregnancy is the state of high hormones level, especially estrogen and progesterone. Hence, hyperplasia is a normal phenomenon during pregnancy. As like uterus, anterior pituitary gland is also underwent in size enlargement under the influence of estrogen, as it is essential for the growth of prolactin-secreting lactotrophs for breast milk production.^[17,8] However, the blood supply to the gland is not changed in response of increased nutritional demand. This endangered the pituitary gland to damage easily if blood loss is more. Due to excessive blood loss just after delivery, there may be decreased blood supply to the pituitary gland and decreased oxygen supply, which can create necrosed area and loss of its functions and due to significant blood loss, it can cause SS in the parturient.^[17] Although it is a very rare complication in postpartum, it is assumed that there must be 75–90% damage to the anterior pituitary gland for developing SS as stated by medical community.^[4] Due to anterior part is affected, the hormones secreted from anterior pituitary now may not be enough of a specified quantity. Hence, the patient may develop deficiency of understated hormones affected with SS; these are growth hormone (responsible for body growth and metabolism), adrenocorticotrophic hormone (ACTH) (which acts on adrenal glands to produce mineralocorticoids [aldosterone], glucocorticoids [cortisol], and androgens), thyroid-stimulating hormone (stimulate secretion of thyroid hormone T3, T4), prolactin (responsible for milk production after baby birth), and follicle-stimulating hormone and luteinizing hormone

(responsible for production of female sex hormones [estrogen and progesterone] from ovary) as shown in Figure 1.^[18]

SYMPTOMS

The major symptoms are depend on the degree of damage occurred to the pituitary gland and level of hormones secreted by it.^[19] The symptoms can range from panhypopituitarism to pituitary hormone deficiencies. Most common symptoms exhibited by woman suffering from SS are secondary adrenal insufficiency, agalactorrhea (absence of lactation), amenorrhea (secondary), feeling of tiredness, hair loss of pubic and axillary area, hypotension,^[3] disturbed metabolism, hard to pass stool (constipation), distinct attention, hyponatremia may also be appeared acutely after baby birth, and osteoporosis is related to hypogonadism in woman affected with SS.^[20]

DIAGNOSIS

Medical history has a major role in diagnosing SS. Inquiry about postpartum hemorrhage should be made. The patient must be asked about difficulty in breastfeeding (lack of milk production),

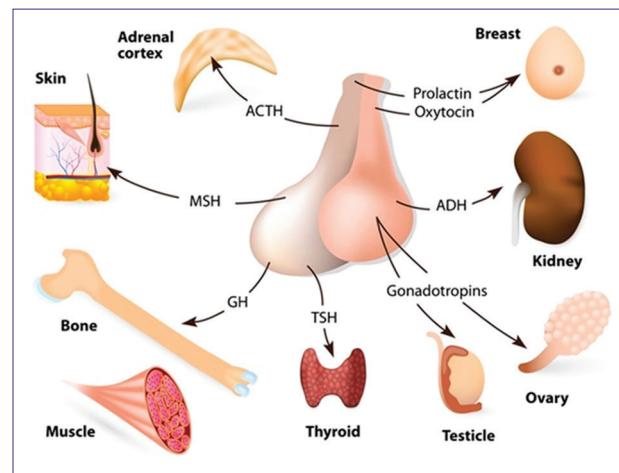


Figure 1: Hormones secreted from pituitary gland^[18]

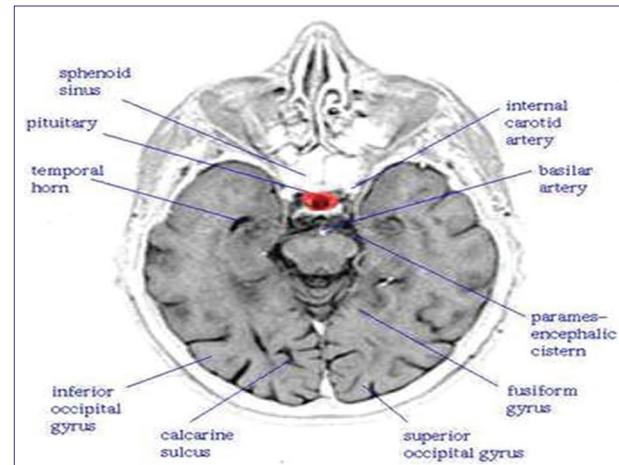


Figure 2: Location of pituitary gland in sphenoid bone^[21]

amenorrhea after childbirth, and unexplained weight gain or loss after delivery.^[17] Imaging: Pituitary gland lies in the sella turcica, as shown in Figure 2 (a groove in the sphenoid bone) which will be emptied after necrosis of pituitary gland. Most of cases showed empty sella turcica.^[21] Detection of hormonal level of pituitary gland and other gonads must be carried out to rule out the lack of functioning of the glands;^[22] magnetic resonance imaging of pituitary gland must be done to eliminate any tumor or abnormal conditioning. Computed tomography scan is also beneficial to know about the size and structure of pituitary gland.^[23]

TREATMENT

Hormone replacement therapy will be needed for rest of the life which includes estrogen, progesterone, thyroid, and adrenal hormones. Hydrocortisone or prednisone is prescribed for the treatment of ACTH. Periodically monitoring of hormonal levels is essential.^[24]

COMPLICATIONS

SS can be a disastrous condition if not diagnosed and treated expeditiously. Due to adrenal insufficiency, sudden drop in B.P occurred which leads to dizziness or fainting, shock, and loss of consciousness and finally leads to death.^[25]

ADDITIONAL AND RELEVANT USEFUL INFORMATION FOR SS

The US National Organization for Rare Disorders categorizes SS as a “rare disorder.”^[4]

PREVENTION

Although with the support of excellent medical services, we can handle the situation, yet the only prevention of SS is prevention of postpartum hemorrhage or severe blood loss after childbirth; otherwise, it is inescapable.^[26]

CONCLUSION

SS is included under one of the rarest disorders after severe blood loss during postpartum period. Severe hemorrhagic loss makes the anterior pituitary gland more prone to get damaged and leads to deficiency of lots of hormone secreted by it. The only treatment which can subside the symptoms is the correction of deficient hormones.

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