Sheehan’s Syndrome: A Rare but Potentially Life-Threatening Complication of Postpartum Haemorrhage

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Description

Sheehan’s syndrome (SS), also known as postpartum pituitary gland necrosis, is a rare but potentially life-threatening complication of postpartum haemorrhage. SS is characterized by varying degrees of anterior pituitary dysfunction [1]. With marked improvement in obstetrical care, it has become uncommon in developed countries. However, it still remains a common cause of hypopituitarism in less developed countries.

The underlying process leading to Sheehan’s syndrome is the infarction of the pituitary gland, particularly the anterior lobe, secondary to the grossly decreased blood supply as a result of severe haemorrhage during and/or after parturition. The pituitary gland is physiologically enlarged in pregnancy and prone to infarction from hypovolemic shock. Vasospasm, thrombosis and vascular compression of the hypophyseal arteries have also been described as possible contributors to the pathogenesis.

The damage to pituitary can be mild or severe, and can affect the secretion of one, several or all of its hormones [1]. At least 75% of pituitary must be destroyed before clinical manifestations of SS become evident. The order of frequency of hormone loss has generally been found to be growth hormone (GH) and prolactin, followed by gonadotropins, adrenocorticotropic hormone and thyrotropin. The most common hormone to get affected is GH 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 [2].

Breast involution, failure of postpartum lactation and failure to resume menses after delivery are the most common presenting symptoms of SS. However, absence of these symptoms does not rule out the diagnosis. Loss of genital and axillary hair is also common. Patients progressively develop clinical manifestations of central hypothyroidism and secondary adrenal insufficiency. These may include malaise, non-specific fatigue, anorexia, joint pains and rarely hypotension. Some patients also develop psychiatric manifestations. It must be remembered that patients of SS can have varied presentations and the clinical manifestation may change from one patient to another [1,3].

Although in a small percentage of patients with SS, abrupt onset severe hypopituitarism may occur immediately after delivery, most patients have a mild disease and remain undiagnosed and untreated for a long time. In a study of 60 patients, the average time between the previous obstetric event and diagnosis of SS was 13 years [3].

The diagnosis of Sheehan’s syndrome is based on the features of hormone deficiency, a suggestive obstetric history and decreased levels of basal hormones. Diagnosis of pan-hypopituitarism is straightforward, but partial deficiencies are often difficult to elicit.

The great majority of the patients have empty sella on CT or MRI.

The general principle of treatment of hypopituitarism holds good for the treatment of SS also. 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. In patients who have secondary hypothyroidism and hypocortisolism together, glucocorticoids should be replaced before the replacement of thyroid hormone. Gonadotropin deficiency and hypogonadism should be treated with hormone replacement therapy [4].

We had a similar experience of a 32-year-old woman, para2 live2, with a past history of full term vaginal delivery at home 9 years back which was complicated by severe postpartum haemorrhage secondary to retained placenta. She was then taken to hospital where manual removal of placenta was done, and PPH was managed conservatively. Patient received 2 units of blood transfusion. Patient was subsequently discharged on the fourth post delivery day. She subsequently developed failure of lactation, secondary amenorrhoea, and loss of pubic and axillary hair. Over the years, she also developed symptoms of generalised weakness and lethargy. She had been ignoring these symptoms for past 9 years. But now when her symptoms worsened, she presented to us. Physical examination was unremarkable. From history a working diagnosis of Sheehan’s Syndrome was made, and subsequently diagnosis was confirmed by laboratory investigations and imaging.

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MRI brain which showed empty sella turcica (Figure 1). The patient recovered completely after initiation of hormonal replacement therapy with corticosteroids and thyroid hormone.

In conclusion, SS is a frequent cause of hypopituitarism in underdeveloped countries. The clinical features of hypopituitarism are often subtle, and years may pass before the diagnosis is made following the inciting delivery. History of postpartum haemorrhage, failure to lactate and cessation of menses are important clues to the diagnosis. Even if PPH is well managed, this complication cannot be excluded. It is necessary to consider this diagnosis in all the patients presenting with classical signs of pituitary insufficiency and having a past history of postpartum haemorrhage with or without cardiovascular collapse.

Early diagnosis and appropriate treatment are necessary to reduce the morbidity and mortality in such patients.

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