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Case Report

A severe adrenal crisis occurred in late diagnosed Sheehan's syndrome developed in a 17-year-old woman

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Abstract

Sheehan's syndrome occurs as a result of severe postpartum hemorrhage and is characterized by varying degrees of anterior pituitary dysfunction. It is an important health problem especially for women who deliver at home in developing countries. In this report, we presented a 17-year-old female patient who gave her second birth at home. During her delivery severe vaginal hemorrhage occurred and she applied to a healthcare facility too late. Since then, she hasn't had menstruation and hasn't been able to get pregnant. She was diagnosed with Sheehan's syndrome 16 years after giving birth. The patient didn't use her medicine regularly after the diagnosis because she wasn't properly informed about the disease, and she has stopped taking it for the last six months. In the current conditions; the 69-year-old patient is presented with hypocortisolemia, hypoglycemia, hyponatremia, and advanced osteoporosis. The patient with severe adrenal crisis was taken in treatment by us.

Keywords: Sheehan's syndrome; adrenal crisis; hypocortisolemia; hypoglycemia; hyponatremia; osteoporosis

INTRODUCTION

Sheehan's syndrome (SS) is rare but crucial complication that occurs as a result of postpartum hemorrhage (PPH) in women. As a consequence of the cardiovascular collapse developed after a life-threatening obstetric hemorrhage, ischemic necrosis may evolve in the pituitary gland (1-4). It was first published by Sheehan in 1937 and is characterized by varying degrees of anterior pituitary dysfunction (5). It may commence after birth with a mild headache, weakness, nausea, lactation loss, prolonged amenorrhea and hypoglycemia crises. In certain occurrences, it may be found partial pituitary necrosis. As a result, when a peculiar and inadequate clinic occurs, it may cause a delay in the diagnosis of the patient (2,6).

CASE REPORT

A 69-year-old woman was presented with symptoms such as weakness, nausea, and weight loss. The patient had a facial appearance looking older than her age and had profound facial lines on her face (Figure 1). She was hypotensive and apathetic, and the muscle potency was debilitated. The patient was unable to walk and confined to bed. We asked about her medical history and came to know that she was 17 years old when she gave her second birth. After the delivery at home, she had severe the postpartum hemorrhage which continued for about a few hours before she was taken to the hospital and treated there. The lactation stopped 4 weeks after the birth and she hasn't had menstruation since then. Because of her socio-economic conditions, the patient wasn't able to go to a medical center. She was treated in the hospital for hypoglycemic crisis 16 years after giving birth. The patient was diagnosed with SS, however, she didn't receive medical treatment regularly. Neither the patient nor her relatives were informed enough about the disease. They didn't know that current treatment had to continue for life. Moreover, since a relative who is a health worker thought steroid might cause osteoporosis, she has stopped replacement treatment for the last six months. We have conducted the laboratory tests, and the results are mentioned below. Glucose: 44 mg/dL, calcium: 8.39 mg/dL, sodium: 125 mEq/L, potassium: 3.2 mEq/L, chlorine: 90 mEq/L, sT3: 1.02 (2.3-4.2) pg/mL, sT4: 0.15 (0.89-1.76) ng/mL, TSH: 1.60 (0.35-5.5) µIU/mL, prolactin: 1.30 ng/mL, FSH: 4.23 µIU/mL, LH: 0.55 µIU/mL, estradiol: <11.80 pg/mL, cortisol: 1.27 (4.3-22.4) µg/dL, somatomedin-C: <15.0 (37-219) ng/mL, 25-OH vit

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D: 6.8 (30-100) ng/mL. Comparative laboratory results of the patient's admission vs discharge values are shown in a table (Table 1).

Table 1. The laboratory results comparing patient's admission vs discharge values

Laboratory tests	Reference range	Admission	Discharge
Glucose	74-106 mg/dL	44	73
Calcium	8.7-10.4 mg/dL	8.39	8.54
Sodium	132-146 mEq/L	125	133
Potassium	3.5-5.5 mEq/L	3.2	4.5
Chlorine	99-109 mEq/L	90	94
Urea	10-49 mg/dL	9	20
Creatinine	0.5-1.3 mg/dL	0.46	0.62
Leukocytes	4-10 109/L	2.69	4.15
Hemoglobin	11-16 gr/dL	10.3	11
Platelets	13-400 109/L	175	176

Because the thickness of the pituitary gland was decreased, it couldn't be clearly identified in magnetic resonance imaging. Cerebrospinal fluid was seen in the cella cavity and this finding was compatible with totally empty cella. Following the intravenous paramagnetic injection, pituitary became homogeneous, diffused substance involvement and no adenoma was detected (Figure 2, 3). We observed severe osteoporosis in the bone mineral densitometer (L1-L4 vertebrae T and Z scores: -4.2, -2.2 respectively; L2-L4 vertebrae T and Z scores: -4.3, -2.3 respectively) and multiple vertebral fractures at X-ray graphy. We have started essential hormone replacement therapy: hydrocortisone 40 mg/day, levothyroxin 50 mg/day and D vitamin 50.000 unit/week for 8 weeks. We planned bisphosphonate therapy for osteoporosis. In a short time the patient's clinic ameliorated and we discharged her from the hospital. Prednisolone 5 mg/day, levothyroxine 75 mg/day, calcium carbonate/colecalciferole 1000 mg/880 IU and ibandronic acid 150 mg/month was prescribed when she was discharged.

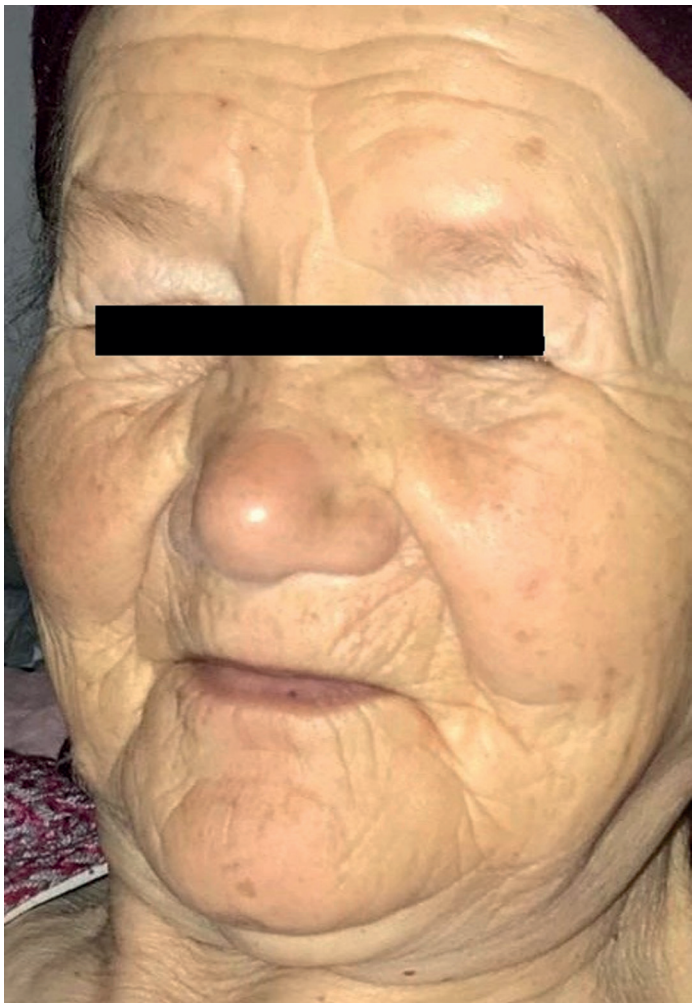

Figure 1. The appearance of striking wrinkles on the patient's face compared to her age

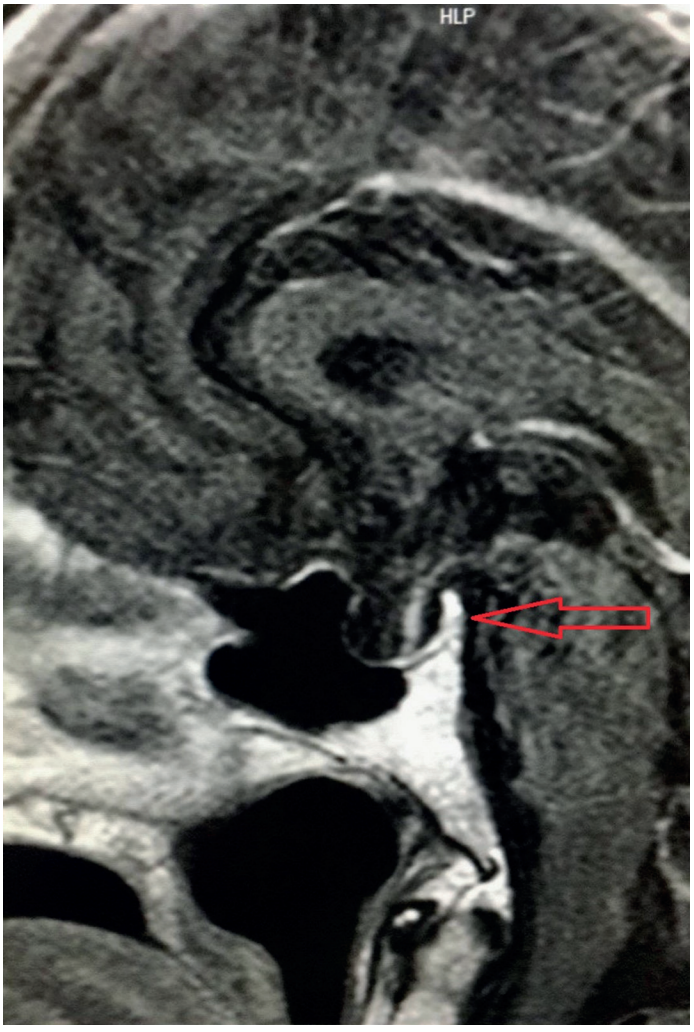

Figure 2. The pituitary thickness was 0.6 mm in the T1 sequence sagittal section (totally empty cella)

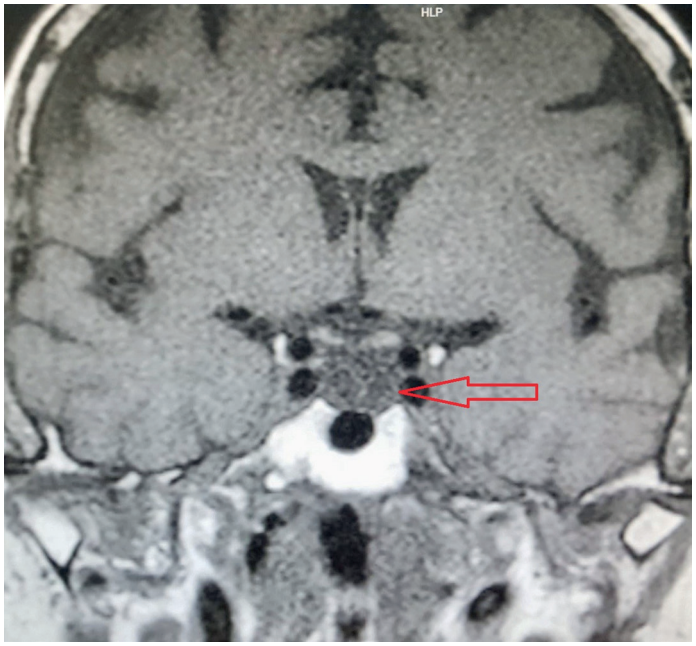

Figure 3. The pituitary wasn't seen in the T1 sequence transverse section

DISCUSSION

In this report, our case delivered her second birth at home when she was 17 years old. She was exposed to severe PPH and treated in a hospital after vaginal bleeding pursued for several hours. She was diagnosed with SS approximately 16 years after this occurrence and didn't have a regular medical follow-up until the age of 69. What we want to emphasize in this case is all hypophysis axis should be evaluated during the diagnosis phase and together with the proper and sufficient replacements therapy; the patient should be informed that this is a lifelong treatment. When the patients are not fully informed, they can hinder or totally stop the treatment. Especially sudden stopping of replacements of the hormones (cortisol, thyroid hormones) having vital importance, can be the reason of mortality or morbidity together with adrenal crisis as in our case.

Obstetric hemorrhage is leading common cause of maternal mortality in the world. In developed countries, maternal deaths are 100 times lower, but PPH remains the cause of maternal death for about ten women per 100 000 births (7). The incidence of Sheehan's syndrome, which consists of ischemic necrosis and hormone deficiency in pituitary due to hemorrhage, has decreased in developed countries. Thanks to better obstetric conditions and advances in anesthesia and reanimation, ranges from 0.2 to 2.8 per 100 000 women in developed countries (8). However, it is a life threatening health problem in societies with low socioeconomic conditions, especially in women who are delivering at home (9,10).

In a study conducted between 2004 and 2005, it was reported that the incidence of PPH in France was 3.1/1000. Moreover, the bleeding-related mortality rate was 2.5 times higher than the European average (7,11). According to a retrospective study conducted in Iceland in 2009, the prevalence of SS was found to be higher than expected (5.1 per 100 000 women) (12). In England, which was another developed European country, the SS prevalence was 100-200 per million people in the 1960s (13). According to another study in England, it was found that less than 1% of 1.014 hypopituitarism patients had SS in 2001 (14).

In Turkey, 20 patients who had SS were analyzed in a study conducted in 2005. The mean age of the patients was 51.12 ± 9.44 years, and the average duration of diagnosis was 16.35 ± 4.74 years after birth (15). In 2014, in a study realized in Turkey 114 patients with SS were evaluated. In accordance with the results of this study, during the diagnosis, panhipopituitarism was found in 55.3% of the patients and partial hipopituitarism was found in 44.7% of the patients. Average time of diagnostic delays was 19.7 years. 52.6% of the patients had non-specific complaints, 30.7% of them had adrenal insufficiency and 9.6% of them had hypogonadism (16). In accordance with another study conducted in Turkey, average time of diagnostic delays was 20.3 years. 62% of the patients with SS gave birth at home and only 5.7% of these patients were literate (17).

The National Family Health Survey in India (1998–1999) revealed that 66% of the deliveries occurred at home, and the incidence of PPH was 11%. In a study conducted in the Kashmir region of India, it was found that 3% of women over the age of 20 had hypopituitarism and 2/3 of them gave birth at home (9).

These results show that women with low socio-economic level, received not enough medical support during their pregnancy, and gave birth at home and living especially in less developing countries are at risk for SS. When the diagnosis is late and the patients aren't informed sufficiently, there can be disruptions in the treatment. In the patients receiving irregular treatment, life threatening complications such as severe adrenal crisis, severe osteoporosis can cause mortality or morbidity.

CONCLUSION

Although SS appears to occur more frequently in women in the developing countries, it is more than expected in the developed countries. It is an important health problem especially for women having low socioeconomic status and living in countryside. Considering the duration of SS, crucial delays occur in diagnosis and treatment of the disease. Informing the patients about their disease properly is as important as the replacement treatment.

Conflict of interest: The authors declare that they have no competing interest.

Financial Disclosure: There are no financial supports.

Patient informed consent : The patient approval was received for this case report and she signed informed consent form.

Ethical approval: Authors followed the ethical principles according to the Declaration of Helsinki. Due to the covid-19 pandemic, local

ethics committee meetings were stopped. So we didn't get the ethics committee approval.

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