

Atypical Revelation of Sheehan's Syndrome

Hind Asbar*, Fatima Ihibbane, Noura Tassi

Service des maladies infectieuses, Hôpital Arrazi, CHU Mohamed VI Faculté de médecine et de pharmacie de Marrakech Université Cadi Ayyad, Marrakech, Maroc

***Corresponding author**

Hind Asbar

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Abstract: Sheehan's syndrome is a disease of difficult and usually late diagnosis. It occurs as a result of ischemic pituitary necrosis due to severe postpartum hemorrhage. We report an atypical observation showing a disconcerting way of revelation of this syndrome. A patient treated as multifocal tuberculosis suspected in the presence of alveolar lung disease, pericardial effusion and altered general status. It was only when there was intolerance to antibacillary treatment that the patient was hospitalized with a rectification of the diagnosis. Our attention was drawn to the existence of clinical signs of hypopituitarism. Examination revealed a secondary amenorrhea for 20 years following a haemorrhagic delivery. The patient was placed under substitutive treatment based on hydrocortisone and L-thyroxine with a very good evolution. Sheehan's syndrome is a common cause of hypopituitarism in developing countries. Our observation illustrates the value of careful examination in the context of this difficult and late diagnostic pathology. Its inexpensive treatment is effective and avoids a dramatic evolution and sometimes serious complications.

Keywords: sheehan's syndrome; postpartum hemorrhage; secondary amenorrhea; hypopituitarism; pituitary MRI.

INTRODUCTION

Sheehan's syndrome is an ischemic necrosis of the pituitary gland following a severe hemorrhage of the delivery. It usually evolves slowly and hence is of delayed diagnosis.

Depending on the degree of pituitary dysfunction, symptoms may vary and occur early or late. The main symptoms are lack of lactation, secondary amenorrhea and loss of libido. Biologically, postpartum hypoglycemia and hyponatremia should suggest the diagnosis. However, these early signs are often overlooked and lead to delayed diagnosis.

In this article, we report the case of a patient treated as probable multifocal tuberculosis with no evidence of acid fast bacilli. In fact, the patient had a Sheehan's syndrome but symptoms and important anamnestic elements were overlooked leading to delay of diagnosis of 20 years.

CASE REPORT

A 47-year-old woman was admitted in the department of infectious diseases for intolerance to antibacillary treatment that she began taking one week before her admission.

The diagnosis of multifocal tuberculosis (pulmonary and pericardial) was highly suspected in the

face of multiple elements. The patient presented with profound alteration of the general state with fever, asthenia, anorexia and weight loss of 15 Kg in a 6 months' duration. She complains of a stage II dyspnea with atypical chest pain and productive cough.

The initial examination found a conscious, pale, tachypneic patient with a respiratory rate of 22 breaths/minute, a heart rate of 62 beats/minute, and a blood pressure of 110/50 mmHg. Lungs auscultation found left basithoracic crackles.

Laboratory findings revealed a normochromic normocytic anemia at 8.8 g/dl, leucocytes at 3980/mm³, neutrophils at 2480/mm³, lymphocytes at 1220/mm³, a platelet count at 112 000/mm³. Hyponatremia at 125 mmol/l, serum potassium at 3,8 mmol/l, CRP at 11 mg/l. Three negative acid-fast bacilli (AFB) in expectorated sputum were negative. Chest X-ray showed cardiomegaly with opacification of the lower left zone. Echocardiography revealed a pericardial effusion that wasn't punctured.

When admitted in our department, the upper gastrointestinal endoscopy revealed an erythematous and atrophic gastritis. The abdominal ultrasound was normal while the echocardiography showed a non-compressive circumferential pericardial effusion measuring 10 and 25 mm respectively in the anterior

and posterior portions that couldn't be punctured, with a preserved ejection fraction. ECG showed low voltage QRS complexes with a heart rate of 62 beats/minute.

Our attention was drawn to the existence of clinical signs of hypopituitarism with depressed mood, slowed speech, intense fatigability, axillary and pubic depilation and widespread depigmentation. The patient interrogation revealed a lactation loss and a secondary

amenorrhea for the past 20 years following a hemorrhagic delivery.

Exploration of the hypothalamic-pituitary-adrenal axis found a thyrotropic insufficiency with decreased T4 at 1,6 pmol/L while TSH was normal at 2 mU/L, cortisol level was low at 5,4 µg/dl, prolactin at 3 ng/ml, FSH at 3,9mUI/ml, LH at 1 mUI/ml and oestradiol at 10 pg/ml. Stimulation Testing couldn't be done at the time. An empty sella was found in the MRI (Figure 1).

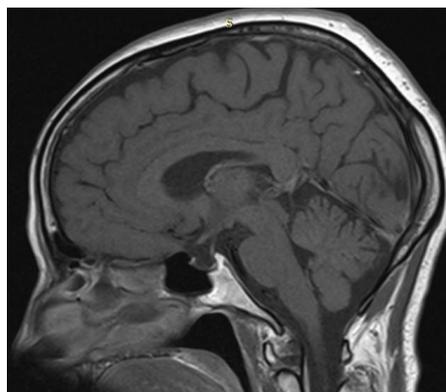


Fig-1: MRI T1-weighted sequence image showing an empty sella

These clinical, biological and radiological data as well as the absence of any aggravation following the discontinuation of antibacillary treatment allowed us to adjust the diagnosis to a Sheehan's syndrome.

The patient was started on replacement therapy by hydrocortisone and L-thyroxine with a good clinical, biological and echocardiographic evolution.

DISCUSSION

Sheehan's syndrome was described for the first time by Harold Leeming Sheehan in 1937 [1]. It is a result of ischemic pituitary necrosis due to severe postpartum hemorrhage, and is a common cause of hypopituitarism in underdeveloped and developing countries [2, 3].

The exact pathogenesis of Sheehan's syndrome is not yet well understood. It appears that the increase in size of the pituitary gland during pregnancy makes it more vulnerable to ischemia due to compression of superior hypophyseal artery. Autoimmunity and disseminated intravascular coagulation seem to be incriminated as well [4].

The latent period between the post-partum hemorrhage and symptoms is very variable, ranging from a few hours early in the postpartum period by acute, sometimes severe events (hypoglycemic coma, myxedema coma, pituitary coma) to several years by variable nonspecific manifestations [5]. This can make the time to diagnosis also variable. Previous studies reported delay in diagnosis up to 33 years [6]. In our case, the diagnosis was made after 20 years.

Clinical manifestations result of total or partial deficiency of anterior pituitary hormone secretion. This can lead to secondary hypothyroidism, hypocortism, hypogonadism and loss of growth hormone and prolactin production. Lactation loss after delivery and amenorrhea are common signs that shouldn't be overlooked. Other multiple complaints are reported in Sheehan's syndrome such as fatigue, weakness, anorexia, arthralgia, headache, dry skin, premature aging, hypotension, constipation, anemia, bradycardia, hypoglycemia, pubic and axillary depilation, infertility, and apathy [7]. Diabetes insipidus, as a sign of posterior pituitary dysfunction in Sheehan's syndrome, was reported in the literature but remains rare [8]. Diagnosis is confirmed by laboratory data, including hormone levels, hormone stimulation test results, and MRI evaluation of the pituitary. MRI is superior to CT in the diagnosis of Sheehan's syndrome. It may show a partial or completely empty sella like it was the case in our patient. Empty sella is a frequent imaging finding in Sheehan's syndrome but the latter is a rare cause of empty sella [9].

The treatment is based on hormone replacement therapy with essentially hydrocortisone and levothyroxin. Gonadal hormone replacement can also be helpful especially in premenopausal women to improve bone density and fertility. Growth hormone substitution remains controversial because of benefit / risk ratio, side effects and treatment cost [10].

CONCLUSION

Sheehan's syndrome is a common cause of hypopituitarism in developing countries. The clinical

signs are often nonspecific but are evocative by their association. That is to say the importance of putting all the elements together for the diagnosis of Sheehan's syndrome.

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