Sheehan’s syndrome with reversible dilated cardiomyopathy: A case report and brief overview

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Sheehan’s syndrome is a rare condition characterized by post-partal panhypopituitarism due to necrosis of adenohypophysis resulting from severe post-partum hemorrhage. Lethargy, amenorrhea and failure of lactation are the usual presenting features. Cardiac involvement in Sheehan’s syndrome is rare. The case presented here describes dilated cardiomyopathy in a 36-year-old lady who failed to respond adequately to the standard anti-failure treatment. Further investigation revealed the diagnosis of Sheehan’s syndrome. Besides other manifestations, cardiac function reverted to normal after giving replacement therapy with glucocorticoid, levothyroxine and sex hormone. Physicians, specially those in developing countries, should have high index of suspicion for the diagnosis of Sheehan’s syndrome while dealing with a case of ‘peripartal dilated cardiomyopathy’. Persistent amenorrhea and failure of lactation may be important clues in this context. Timely diagnosis and appropriate treatment can lessen the sufferings of the patients.

Keywords: Sheehan, Hypopituitarism, Reversible, Cardiomyopathy

Introduction

Sheehan’s syndrome is a rare condition characterized by post-partum panhypopituitarism due to necrosis of adenohypophysis resulting from severe post-partum hemorrhage. Lethargy, amenorrhea and failure of lactation are the usual presenting features. Cardiac involvement in Sheehan’s syndrome is rare. The case presented here describes dilated cardiomyopathy in a 36-year-old female who failed to respond adequately to standard anti-failure treatment. Further investigation revealed Sheehan’s syndrome. Besides other manifestations, cardiac function reverted to normal after replacement therapy with glucocorticoid, levothyroxine and sex hormone.

Case presentation

A 36-year-old non-diabetic female with history of childbirth 2 years prior, presented with progressive breathlessness, effort intolerance and...
dependent edema over a period of 6 months. She was provisionally diagnosed as a case of peripartum dilated cardiomyopathy (DCM). Standard treatment was offered, and she was finally referred for further evaluation and management due to poor response to standard treatment. Physical examination revealed lethargy, pallor, sparse body hair, atrophied breasts, husky monotonous voice, dyspnea and leg edema. Her pulse was 68/min, blood pressure 90/60 mmHg, shifted, diffuse apex beat and third heart sound (S₃). Further enquiry revealed complicated, vaginal home delivery with severe, post-partum hemorrhage requiring 6 units of blood transfusion, followed by progressive effort intolerance, loss of libido, failure of lactation and amenorrhea. Her blood counts were normal with hemoglobin (Hb) 12.7 g/dl, normal iron profile, renal and hepatic functions, and serum electrolytes. There was cardiomegaly in chest X-ray (CXR), ascites in abdominal ultrasound (USG) and inverted T waves in precordial leads of electrocardiogram (ECG). Echocardiography showed dilated cardiac chambers with severe global left ventricular (LV) hypokinesia, severely impaired LV ejection fraction (LVEF 29%) and grade III mitral regurgitation (MR) (Fig. 1). Considering her history, inquiry into endocrine status revealed low thyroid stimulating hormone (TSH) (0.68 micro IU/ml; normal 0.47–5.01 micro IU/ml), low tetraiodothyronine (T₄) (0.86 µg/dl; normal 4.50–12.0 µg/dl), low follicle stimulating hormone (FSH) 3.22 m IU/ml, normal 4.0–13.0 m IU/ml, and low luteinizing hormone (LH). However, serum prolactin and ACTH levels were within normal limits. MRI showed empty sella. (Fig. 2) She was diagnosed as a case of Sheehan’s syndrome with dilated cardiomyopathy. Along with diuretics, she was given ACE inhibitor, digoxin, and hydrocortisone, 100 mg six-hourly IV for 2 days followed by prednisolone 5 mg in the morning and 2.5 mg orally in the evening. This was followed by administration of increasing dose of oral levothyroxine, up to 100 µg/day. With this treatment, the patient’s general wellbeing improved dramatically, breathlessness decreased, edema disappeared, and psychological status improved. She re-commenced menstruating, and, in her own words, she found her ‘second life’. In
follow up, serial echocardiography over 6 months showed reduction of chamber dimensions, improved myocardial contractility, disappearance of MR and finally, complete recovery of systolic dysfunction. The dose of glucocorticoid, and levothyroxine were adjusted according to clinical parameters (sense of well-being and body weight) and serum free T4 levels.

Discussion

Sheehan’s syndrome occurs as a result of ischemic pituitary necrosis, usually due to severe postpartum hemorrhage [1]. Vasospasm, thrombosis, vascular compression of the hypophyseal arteries, enlargement of pituitary gland, small sellar size, disseminated intravascular coagulation and autoimmunity are the suggested role-players in the pathogenesis of Sheehan’s syndrome [2,3]. Though rare in the developed world due to improved obstetric care, Sheehan’s syndrome remains the most common cause of hypopituitarism in developing countries [2]. In women, its prevalence was found to be 3% in the Kashmir valley of the Indian subcontinent, [4] and 5.1 per 100,000 in Iceland [5].

Cardiac disorders including DCM in association with hypopituitarism have been described previously. Laway et al. [6] reported a case of Sheehan’s syndrome with DCM and pulmonary tuberculosis in a 25-year-old Indian woman, in whom the DCM completely reversed after replacement therapy in the form of glucocorticoids and levothyroxine, in addition to antitubercular treatment. Wang et al. [7] described a young woman with Sheehan’s syndrome who presented with ventricular arrhythmia and congestive heart failure, and responded to appropriate replacement therapy.

Some of the cardiac abnormalities associated with Sheehan’s syndrome have been attributed to growth hormone deficiency, and improvement of cardiac function was described with growth hormone replacement [8,9]. On the other hand, replacement of levothyroxine resulted in reversal of dilated and hypertrophic cardiomyopathies, found in patients with hypothyroidism [10]. In association with hypopituitarism, DCM may be multifactorial: deficiency of thyroid, corticosteroid and growth hormone may contribute to varying degrees. In the index case, DCM was diagnosed readily, but the underlying etiology was not initially diagnosed, and was most probably Sheehan’s syndrome, not idiopathic, peripartum DCM. This is evidenced by the concomitant presence of hypopituitarism, and further, rapid improvement by hormone replacement. The diagnosis was initially missed probably because of inadequate attention to the typical history of complicated childbirth, failure of lactation and amenorrhea. In comparison to Sheehan’s syndrome, peripartum etiology is presumably a more common cause of DCM, so bias to the latter diagnosis is more logical. Sophisticated stimulation tests were not done, because of typical features of Sheehan’s syndrome, including MRI findings, and very limited availability of the tests.

Figure 2. Magnetic resonance imaging of patient’s brain: enlargement of pituitary fossa which is filled with CSF along with thinned, compressed pituitary gland (white arrow) in non-contrast T1 weighted sagittal image (left panel); also the pituitary infundibulum (yellow arrow) traverses the enlarged sella to meet the thinned, compressed pituitary gland at the bottom, i.e., the ‘infundibular sign’ in T2 weighted coronal image (right panel).
For the treatment of Sheehan’s syndrome, deficient hormones are replaced and glucocorticoid is replaced prior to starting levothyroxine in order to avoid precipitation of adrenal crisis. Hydrocortisone is preferable to prednisolone, but the oral formulation of hydrocortisone is not readily available locally. Sex hormone was given because the patient was of reproductive age, and to prevent osteoporosis in the future. Growth hormone replacement in hypopituitarism in adults is a controversial issue, though GH replacement therapy in patients with Sheehan’s syndrome has been shown to be beneficial [11,12]. In the present case, growth hormone was not given due to satisfactory improvement without it, and due to high cost. Replacement of the deficient hormones with simple, inexpensive medications in this case led to marked subjective and objective improvement of the patient, and at the same time, reversal of functional impairment of the heart.

Sheehan’s syndrome is an uncommon diagnosis, and is a very rare cause of DCM in females of reproductive age. Physicians, especially those in developing countries, should have high index of clinical suspicion and low threshold while dealing with a suspected case of ‘peripartum DCM’ for the very rare association of Sheehan’s syndrome. Persistent amenorrhea and failure of lactation may be important clues in this context. Timely diagnosis and appropriate treatment can lessen the sufferings of this younger population.

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References