A young lady with hypotension and engorged neck veins

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A 25-year-old woman presented to us with history of breathlessness. The lady had noticed progressively increasing fatigue in the past six months and had a sense of generalized weakness. Over the past week or so she had been unable to do even routine chores at home and had been breathless since the past two days. The dyspnoea increased with lying down. She denied any history of fever.

She appeared pale, had dry skin and sparse pubic and axillary hair. Her neck veins were engorged. She had a pulse rate of 86/min, blood pressure of 76/40 mm Hg. Pulsus paradoxus was absent. Her chest was clear and heart sounds were distant.

What is the Possible Diagnosis?

The presentation of the case suggests a subacute to chronic process. The history of orthopnea, engorged neck veins and hypotension indicate a cardiac pathology. The features of engorged neck veins, hypotension and muffled heart sounds constitute the classical Becks' triad of cardiac tamponade.[1]

A chest roentgenogram revealed massive cardiomegaly. Electrocardiogram revealed low voltage complexes. An echocardiography revealed pericardial effusion with features of cardiac tamponade with collapse of the right atrium and ventricle in diastole. An urgent pericardiocentesis was performed and around 350 ml fluid removed. The condition of the patient improved.

What are the Possible Causes of Cardiac Tamponade in this Patient?

The classical Beck’s triad is not usually seen in slow-developing effusion. However, the presence of pallor, dry skin and prolonged progressive fatigue suggest a long-drawn process. The possibilities to be considered in chronic pericarditis include chronic infections (tuberculosis, fungal), neoplastic (lymphoma, metastasis from breast, lung), uremia, myxedema, collagen vascular diseases (systemic lupus erythematosus (SLE), rheumatoid arthritis, scleroderma), severe chronic anemia, chylopericardium etc.[1]

In this patient in view of the chronic nature of complaints, tuberculosis is a distinct possibility. The presence of dry hairless skin suggests a possibility of hypothyroidism. However, pericardial effusions secondary to hypothyroidism only rarely result in tamponade.[1][2] Although tamponade is rare in SLE, cases of patients of SLE actually presenting with tamponade are known.[1] Even drug-induced SLE has occasionally presented with tamponade as first manifestation.[4] In view of her young age collagen vascular disease can be a possibility. Also tamponade can be the presentation of a yet occult malignancy.[1]

Investigations revealed Hb- 9.8 gm/dl, total leucocytes of 9800 with 75% polymorphs and 23% lymphocytes. ESR was 32. Red blood cells were normocytic and normochromic. Liver and renal function tests were normal. The serum Na+ was 132, serum K+ was 3.7 and serum Ca++ was 9 mg/dL. The pericardial fluid of the patient was slightly yellow. Pericardial fluid examination revealed proteins of 4.1 g%, sugar 42 mg% and total cells 50/ml predominantly lymphocytes. No atypical cells or cholesterol crystals were observed. Gram and AFB staining, culture and PCR for mycobacterium were negative. Adenosine deaminase levels were 6 IU/ml.

The patient said that she was amenorrhoeic since her last childbirth four years back. She had delivered a child at her home four years back and had needed hospitalization for severe postpartum bleeding. Following this she had a failure of lactation and had not had a pregnancy ever since. She also complained of loss of libido, hair loss, increasing fatigue.

What is the Diagnosis in this Patient?

The history of postpartum bleeding suggests Sheehan’s syndrome. The classical history of lactational failure followed by failure of menstruation virtually clinches the diagnosis of Sheehan’s syndrome.[5]
The manifestations in this are believed to result from the postpartum necrosis of pituitary due to postpartum haemorrhage. This is because of higher blood supply needed by pituitary as a result of physiological hypertrophy during pregnancy. However, evidence suggests a role of autoimmune response to sequestered antigens released as a result of pituitary necrosis in the causation of Sheehan’s syndrome. This ongoing autoimmune phenomenon explains the late presentation of Sheehan’s in the form of circulatory collapse (hypocortisolism and hypothyroidism) occurring long after the initial manifestations of acute loss of pituitary function (agalactia and amenorrheic). In fact it has been seen that a higher percentage of patients with Sheehan’s syndrome had pituitary antibodies as compared to controls.\[6\]

Sheehan’s syndrome can present with variable manifestations depending on the involvement of the pituitary. The features may include postpartum lactational failure and amenorrheic, fatigue, weakness, hair loss, fine wrinkles around face, loss of libido, polyuria, hypoglycemia and hypotension etc.\[5\]

Her hormonal studies revealed a serum TSH-0.6 (0.35-5.5 mU/L), serum free T3- 0.8 (2.3-4.2 pg/mL), serum free T4-0.2 (0.9-2.80 ng/dL), serum FSH-4.75 (23-116 IU/L), serum LH-1.83 (15.9-54.0 U/L), serum cortisol (8 AM morning)-1(5-23 ug/dL).

These are consistent with the possibility of panhypopituitarism. The serum TSH can be normal, low or sometimes raised in central hypothyroidism. Adrenocorticotropic hormone is usually low (not available in our patient).

What are the Causes of Panhypopituitarism?

The possible causes to be considered in this patient apart from Sheehan’s include lymphocytic hypophysitis or other infiltrative diseases, pituitary adenomas or a parasellar mass, tuberculosis or rarely, empty sella syndrome. Magnetic resonance imaging of the brain using gadolinium contrast revealed a normal pituitary, sella and parasellar region. Imaging in Sheehan’s usually reveals an empty sella of normal size although enlarged or partially atrophied pituitary may be seen depending on the stage of the disease. Rarely, as in the present case, sella can be normal (Figure 1).\[5\] Although some authors have suggested a correlation between the amount of pituitary remnants and hormonal secretory capacity this does not appear to be true in this case.\[7\]

What are the Cardiac Manifestations of Hypothyroidism and are they Different in Primary and Central Hypothyroidism?

Cardiac manifestations of hypothyroidism include sinus bradycardia, diastolic hypertension, narrow pulse pressure, pericardial effusions, coronary artery disease etc. Pericardial effusions although common with primary hypothyroidism, are rare in central hypothyroidism.\[12\] Hypothyroidism-related pericardial effusions are usually clear and straw colored. Occasionally they may be yellow (gold paint) due to cholesterol pericarditis.\[16\] They only rarely cause tamponade even in primary hypothyroidism. Usually, the hypothyroidism in Sheehan’s syndrome is less severe than in primary hypothyroidism.\[19\] This patient presented with features of cardiac tamponade which was confirmed on echocardiography.

Is Cardiac Tamponade Described with Sheehan’s Syndrome?

Cases of Sheehan’s syndrome presenting as tamponade have been reported earlier.\[8-10\] Cardiac tamponade is rare with hypothyroidism due to earlier detection of hypothyroidism. But in cases presenting late this can be seen. Cardiac tamponade occurs when the amount of fluid in the pericardium is sufficient to restrict the inflow of blood into the ventricles. The amount of fluid necessary to cause tamponade varies with the rapidity of development of effusion. In hypothyroidism, since the process is slow, tamponade is unusual. In such cases tamponade can develop slowly and may mimic heart failure with features of dyspnea, orthopnea, tender hepatomegaly.\[11\] In Sheehan’s syndrome the presence of low cortisol may result in low-pressure tamponade. In low-pressure tamponade, usually occurring in hypovolemia, the features can be subtle and easily missed. Here the jugular venous pressure is usually normal or only slightly elevated, the arterial pressure is normal and pulsus paradoxus usually absent.\[2,8\]

What are the Lessons from this Case?

With two-thirds of deliveries occurring at home and the poor state of obstetric and general healthcare, Indian women are at a significant risk of developing Sheehan’s syndrome. Although not much literature is available about the epidemiology of Sheehan’s syndrome, a study in Kashmir valley projected that more than 3% of all parous women ≥20 years might be having Sheehan’s syndrome. Very often these women present late with life-threatening complications as in our case.\[11\]

This case indicates the need to consider hypothyroidism, even central, as a possible etiology in patients with unexplained pericardial effusion. Since Sheehan’s can present at a variable duration after the last pregnancy it has to be considered even if history of childbirth is remote. Lactational failure is seen in most severe cases of Sheehan’s syndrome. Therefore follow-up
and the cardiomegaly disappeared [Figure 2]. The patient is still on regular follow-up.

References


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