A RARE CAUSE OF ANEMIA-SHEEHAN'S SYNDROME-A CASE REPORT
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Abstract:
Sheehan's syndrome caused by necrosis of the pituitary gland is a rare complication of post partum haemorrhage occurring in 1-2 percent of cases. The diagnosis can be difficult when presenting many years after child birth and when presenting with partial deficiencies rather than panhypopituitarism. This disorder primarily presenting as a haematological abnormality is quite rare. Here we present a case of 24 yr old female who presented 4 yrs after her child birth with severe refractory anemia.

Keyword : sheehan's syndrome- post partum haemorrhage- hypopituitarism-anemia

Introduction
Sheehan's syndrome occurs as a result of ischemic necrosis of the pituitary gland caused by severe postpartum haemorrhage. Although decreasing in frequency in recent years, it is still one of the commonest causes of hypopituitarism in developing countries owing to the lack of effective management of postpartum bleeding. The clinical presentation of this syndrome is variable; the patient can present abruptly with acute hypopituitarism or insidiously with non-specific features. Its diagnosis is based on the clinical features of associated hormone deficiency, a suggestive obstetric history, laboratory finding of decreased hormone levels and related radiological features. Its treatment requires lifelong replacement of the deficient hormones.

ARTICLE:
24 yr old female patient presented with easy fatiguability, asthenia and grade 2 breathlessness over the past 4 yrs. she had no history chest pain,palpitations or syncope; no history of swelling of legs,abdominal distension, facial puffiness or decreased urine output;no history of jaundice; no history of hematemesis/melena/bleeding diathesis; No history of passing worms in stools; No cough with expectoration or fever. She had no history suggestive of connective tissue disorder either. She had no exposure to potential drugs or toxins . She had her only child birth 4 yrs ago. Antenatal period was uneventful. She had an emergency Caesarean for foetal distress and had significant post partum bleeding. Her menstrual cycles were irregular initially and now is amenorrheoic for the past 4 yrs. On examination she was pale, poorly built and nourished, had absent axillary and pubic hair. Her B.P. was 96/60 mm Hg.

Her cardiovascular,respiratory and abdomen examination were normal. We started evaluating her for anemia.

INVESTIGATIONS:
RBC Count – 3.0 million/cmm(3.8-5.8),Hb – 7.7 g%(11-15), PCV – 31.2%(34-44),ESR 10/22.Total and differential WBC counts, platelet counts were within normal limits. Peripheral smear study showed reduced count of RBC, predominantly normochromic, normocytic. No nucleated RBC seen. No sickle cells, targetcells, inclusion bodies,WBC and plateletes – Count normal. Distribution and morphology normal. No immature cells.No blood parasites. Blood sugar, urea, creatinine, S.electrolytes,liver function tests, serum proteins were normal. Urine analysis was normal, negative for haemoglobin, no occult blood in stool analysis either. Chest x-ray, ECG and ECHO were normal. USG abdomen & pelvis showed reduced size of Ovaries. Upper GI scopy was normal. Iron studies were normal ( S.Iron 75ng/ml, S.TIBC 326 ng/ml, S.Ferritin 68 ng/ml);serum Vit B12(245pg/ml) and Folic acid (9nmol/L) were within normal limits,RDW – 16.3% (11-18),Reticulocyte count – 1.3% (0.2-2%),S. LDH 205 U/L (135 -220 ),Coombs test: negative. Hemoglobin electrophoresis was normal with no specific hemoglobin variants. Bone marrow study revealed Hypocellular marrow with Normoblastic erythroid progenitors,granulocytic progenitors with preserved sequence of maturation. Thyroid function tests showed Free T3 0.26 pg/ml( 2-4.4),Free T4 0.07ng/dl(0.8- 2.o), TSH 1.750mU/l(0.35-5.50),all were remarkably low. On probing the history, the patient revealed that she was transfused with 4 units of whole blood for severe post partum haemorrhage during child birth and that she had never breast fed the child and had not resumed her menstrual cycles since then. We proceeded with other hormone profile studies. Se.FSH 1.39mU/ml and Se.LH 0.51 mIU/ml. Se Prolactin -0.92mg/ml( 4.79-23.3 ng/ml). Se Cortisol - 0.639 mcg/ dl( 6.2 –19.4 mcg/dl).Se ACTH – 6.88 pg/ml (7.2 –63.3 pg/ ml) .Serum GH-1.007ng/ml( 0.010-3.607). All anterior pituitary hormones were low. Posterior pituitary hormones oxytocin and vasopressin were normal. MRI Brain showed Empty Sella sign of pituitary.

MRI BRAIN SHOWING EMPTY SELLA
Thus based on history of post partum haemorrhage, lactation failure, amenorrhoea, asthenia, low hormone profiles and empty sella, a diagnosis of Sheehan’s syndrome and associated anemia was established. Our patient was started on hormone replacement, initially steroids were started followed later by thyroxine and estrogen, progesterone supplementation. Her anemia improved remarkably after hormone supplementation (Hb 12.2 gms) and is on regular follow up.

AFTER TREATMENT

DISCUSSION

Sheehan’s syndrome also known as Simmond’s syndrome, was first described in 1937 by the British pathologist Cindy Sheehan(5,9). It refers to postpartum hypopituitarism as a result of pituitary necrosis due to severe hypotension or shock secondary to postpartum haemorrhage. Its prevalence is decreasing now due to improving obstetric care(10). Hypertrophy and hyperplasia of lactotrophs during pregnancy results in enlargement of the anterior pituitary, without a corresponding increase in blood supply. The anterior pituitary is supplied by a low pressure portal venous system, thereby rendering it more susceptible to ischemia. These factors, when affected by haemorrhage or hypotension during the peripartum period, can result in ischemia of the affected pituitary region leading to necrosis. The mean duration between postpartum bleeding and the subsequent development of symptoms varies from 1 to 33 years. The presenting signs and symptoms are related to the underlying hormone deficiencies and include weakness, fatigue, hypoglycemia, or dizziness( corticotrophin deficiency), amenorrhoea, oligomenorrhea, hot flashes, or decreased libido( gonadotrophin deficiency), fatigue, decreased muscle mass(growth hormone deficiency), agalactia ( prolactin deficiency), slowed mentation, cold intolerance(hypothyroidism), asthenia ,anemia(4), pancytopenia(1,2), headache, and hyponatremia (8). Some women unknowingly live for years with pituitary insufficiency, then go into adrenal crisis triggered by extreme stressors, such as infection or surgery. The diagnosis of Sheehan’s syndrome is based on the features of hormone deficiency, a suggestive obstetric history and neuroimaging. A characteristic MRI finding in pituitary apoplexy is an enlarged pituitary gland bulging under the optic chiasm with peripheral enhancement surrounding an hypointense gland, "pituitary ring sign" later becoming the empty sella sign.

The treatment of Sheehan’s syndrome is hormone replacement. Hydrocortisone or prednisolone is replaced first because thyrxine therapy can exacerbate glucocorticoid deficiency and can induce an adrenal crisis. Growth hormone replacement in adults is controversial. The anemia that develops in Sheehan’s syndrome is due to cortisol deficiency, hypothyroidism and hypogonadism. While glucocorticoids stimulate erythropoiesis, thyroid hormones stimulate not only erythropoietin production, but also the proliferation of erythroid progenitor cells. Sheehan patients presenting with anemia have normochromic-normocytic anaemia (55%) or hypochromic-microcytic anaemia (45%). Anaemia is frequently associated with Sheehan’s syndrome and responds to appropriate replacement therapy(6,7). Hypopituitarism should be considered as a possible cause of anaemia, and a hormone examination should be undertaken promptly, particularly in patients with anaemia resistant to therapy and/or with a history suggestive of Sheehan’s syndrome.(4)

REFERENCES:
