A CASE REPORT ON EMPTY SELLA SYNDROME WITH SHEEHAN SYNDROME

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Abstract

Empty Sella Syndrome (ESS) is an Endo-neurological disease characterized by shrinkage of the pituitary gland due to primary or secondary causes in which an imbalance of pituitary secreting hormones and headache vomiting are commonly seen. In this case, we saw two more conditions with ESS, namely Sheehan Syndrome and Pituitary apoplexy, we observed hormonal imbalance, X-Ray findings, MRI findings, and psychiatric/Gynecological evaluation with a significant history of amenorrhea but no history of surgery, after relevant examination and differential diagnosis we concluded that the patient was suffering from ESS due to unknown or secondary reasons and in this particular case we couldn't find any anti pituitary antibodies so we concluded that it is not caused autoimmunity.

INTRODUCTION

The term "Empty Sella Syndrome" was first coined in 1951 for neurological or pathological anatomical exposure of an empty sella turcica. (1)

Its pathophysiological characteristic by either anatomic abnormalities in the diaphragm sellae (primary ESS), or it can also occur by damage to the pituitary by irradiation surgery or autoimmunity in resulting the availability of "void" space in the sella (secondary ESS).(2)

Due to pituitary shrinkage or structural changes and intracranial pressure, the most common hormone secretion deficiency is growth hormone, affecting 35% to 61% of adult patients. and other hormonal secretion dysfunctions are LH, TSH, FSH, ADH, and ACTH

Its most common symptom is headache, which is either deep, dull, centrally situated or severe, along with vomiting.(2)

Classification

1. **Primary or idiopathic**

   This is due to inheritance and idiopathic with autoimmunity condition leading to deficiency of diaphragma sellae, idiopathic intracranial hypertension, congenital hypopituitarism, and antipituitary antibody either causing shrinkage of the pituitary gland or herniation CSF into the pituitary cavity. (3)

2. **Secondary**

   This type of EES is due to possible conditions like medications and medical surgery irradiation, traumatic head injury, Sheehan syndrome, pituitary tumor, impaired CSF dynamic, CSF rhinorrhoea, menopause, multiple pregnancies, obesity, sleep apnea, an autoimmune disease.(4)

Sheehan syndrome is a condition at which postpartum hypopituitarism or a decrease or absence of one or more of the secretions of the hormone due to necrosis of the pituitary gland. Due to the above condition, severe hypotension or shock owing to massive uterine bleeding during or soon after childbirth. Sheehan's syndrome is always associated with a sella turcica that is partially or completely empty. Pituitary remnants were found in 15 out of 54 patients. (5)

In 57 patients with Sheehan's syndrome and 17 female controls, high-resolution computerized tomography was used to assess the size and contents of the pituitary fossa. There were large variations in the volume of the sella. In contrast, a mean sella volume of 922 mm3 (SD 155) was found in controls, whereas the average for all patients was 565 mm3. By using the Mann-Whitney test (P less than 0.001), the difference was statistically significant. 81 percent of patients (46 of them) had the stalk visible (20 of these also had other pituitary tissue in their samples). Only pure CSF density was found in the remaining 11 patients, and there was no pituitary tissue. (6)

Pituitary apoplexy condition in which oxygen supply decreases due to acute ischemic infarction or hemorrhage of the pituitary gland. Apoplexy of the pituitary (PA) has been implicated in the development of empty sella (ES). Most cases described in the literature were

diagnosed through anamnesis. The following study looked at three patients diagnosed during a PA episode. It was necessary to periodically evaluate hormone levels and computed tomography images of the brain. Within 6 and 18 months, all three patients developed ES in the anterior pituitary.(7)

Case Representation

This patient was a 29-year-old, North Indian female with second-degree amenorrhoea from two years, central hypothyroidism and cerebrovascular accident (chronic microbleeds) past medical history and no significant surgical history. She lived in a rural area. She presented with fever general weakness, lethargy, dizziness and loss of appetite for 7 days in. She suddenly experienced fever up to 40°C. she sometimes felt ocular pain without visual disturbance. Vomiting and abdominal pain were experienced. Physical examination was notable for a blood pressure of 110/ 80mm,pulse 80bpm, respiratory rate 18 breaths per min and peripheral capillary oxygen saturation (spo2) was 98%, Her body temperature recovered to the normal level of 37°C.she experiencing significant hair loss Laboratory test results were as follows. Routine blood examination showed WBC of 6680 /cumm, percentage of neutrophils of 77%, percentage of lymphocytes of 20%, red blood cells of 3 and platelets of 93,000 cells/mL. The urine examination had nothing found significant. Liver enzymes were increased (Glutamic-oxalacetic transaminase (SGOT) 49.3 U/L, glutamic-pyruvic transaminase (SGPT)38.0 U/L, gamma-glutamyl transferase 48.0 U/L). Then, the examination of pituitary function was performed. All of the results were fluctuation than the reference range thyroid-stimulating hormone: 0.028mIU/mL (0.27–4.2), free T3 3.46 pmol/L (3.1– 6.8), free T4 5.0 pmol/L (12–22); prolactin: 451.00mIU/L (55.97–278.36); follicle-stimulating hormone: 0.970mIU/mL (1.27–19.26); GH: 0.026 ng/mL (0.003– 0.971); luteinizing hormone: 0.216mIU/mL (1.24–8.62); Renal function was found normal. The regular blood cell test result was normal.Iron profile was taken to rule out type of anemia and showed a low level of TIBC 237 ug/dl (265-497ug/dl). She felt fatigued and lost her appetite. A urine culture showed no growth of microorganisms. The brain MRI result showed that signals of cerebrospinal fluid could be observed in sella. X-ray examination and anti-mullerian hormone found normal as well as nothing particularly significant. The history interview questions and gynecological reference revealed the history of Sheehan syndrome with significant data and the possibility of past pituitary apoplexy but a lack of significant data. other references were also done to understand possible comorbid conditions such as psychiatry, neuro physician for the betterment of therapeutic outcomes.

During 10 days of hospitalization, she experienced constipation, generalized weakness, mood swings, fever but recovered with treatment and counseling.

Laboratory Data

THYROID PROFILE AND PITUITARY HORMONE			
S no.	Tests	Value	Normal range
1.	TSH	0.028	0.470-4.680MIU/L
2.	T3	3.46	4.26-8.10 pmol/L
3.	T4	5.0	10.0-28.2 pmol/L
4.	PROLACTIN	451.00mIU/L	55.97–278.36 mIU/L
5.	FSH	0.970mIU/mL (1.27–19.26)	1.27–19.26 mIU/mL
6.	LH	0.216mIU/mL	1.24–8.62 mIU/mL
COMPLETE HEMOGRAM (CBC)			
1.	HB	10.7	12-15g/dl
2.	TLC	6680	4000-11000/cumm
3.	NEUTROPHILES	77	44-68 %
4.	LYMPHOCYTES	20	25-48 %
5.	RBC	3.73	3.8-4.8
6.	PCV	33.9	36-46%
LIVER FUNCTION TEST			
1.	SGOT	49	14-36U/L
2.	SGPT	38	4-35U/L
3.	GGT	48	12-43U/L
4.	GLOBULIN	3.6	2.3-3.5g/dl

IRON PROFILE			
1.	TIBC	237	265-497ug/dl

Discussion

The empty sella turcica is defined as an intrasellar herniation of the suprasellar subarachnoid space with compression of the pituitary gland producing a remodeling of sella, resulting in many cases a combination of incomplete diaphragma sellae and an increased CSF fluid pressure. There are two types of ESS: primary and secondary.(8) Primary ESS happens when a small anatomical defect above the pituitary gland increases pressure in the sella turcica and causes the gland to flatten out along the interior walls of the sella turcica cavity. (8) The primary Empty Sella syndrome is generally found in middle aged women who are obese and hypertensive.The disorder can be a sign of idiopathic intracranial hypertension. Secondary ESS is the result of the pituitary gland regressing within the cavity after an injury, surgery, or radiation therapy. Individuals with secondary ESS due to destruction of the pituitary gland have symptoms that reflect the loss of pituitary functions, such as the ceasing of menstrual periods, infertility, fatigue,and intolerance to stress and infection.

As we know empty sella syndrome is caused by various etiological reasons and certain populations or risk factors are associated which makes it a life-threatening condition. Different etiologies are 1. inherited or idiopathic (idiopathic intracranial hypertension, deficiency of diaphragma sellae or inherent weakness of EST, congenital hypopituitarism), 2. Disease condition or traumatic injury (Sheehan syndrome, Traumatic head injuries, pituitary tumor, menopause, Hypophysitis, viral infections, meningitis), 3. Autoimmunity (AITD, Antipituitary Antibody), 4. Impaired CSF dynamic (CSF rhinorrhoea), 5. Medical therapy and surgeries (pituitary surgery, irradiation, medications), 6. Risk factors or population (multiple pregnancies, female sex(5:1), obesity, sleep apnea, head injury, hypertension.

As above mentioned, etiologies lead to progressive CSF herniation into the pituitary cavity whichfurther leads to shrinkage of the pituitary gland.

When the pituitary gland shrinks cause dysfunction in the secretion of pituitary hormones it leadsto three major conditions that are hypopituitarism, pan pituitarism, and hyperpituitarism. These three conditions differently show signs & symptoms which are visible physically and on laboratory data.

Sign and symptoms are anorexia, abdomen pain, nausea, vomiting, weight loss, hypoglycemia, loss of pubic hairs (women), reduce bone mineral density, impaired thermogenesis, decrease sweating, increase central adiposity, decrease muscle mass & strength, cold intolerance, constipation, psychomotor retardation, polyuria, polydipsia, nocturia, lactation failure, erectile dysfunction, oligomenorrhea, breast atrophy, loss of libido, flushes, infertility.(8)

In this case, we found physical symptoms like dizziness, lethargy, visual disturbance, loss of appetite, weight, fever, and generalized weakness.

Laboratory examination revealed central hypothyroidism, cerebrovascular accident, and dysfunction in the secretion of pituitary hormone-like TSH, prolactin, LH,and Liver function test and thyroid function test shows elevated liver enzyme, and thyroid hormones respectively.(9)

The renal function test was found to be normal. Normal blood count (CBC) shows low levels of hemoglobin, from which anemia was suspected for ruling out a specific type of anemia; an iron profile test was done and TIBC was found to be low. X-ray of lungs was found to be normal History Interview revealed history of Sheehan’s syndrome possibilities of the past

history of pituitary apoplexy. MRI revealed structural changes in the pituitary which led to micro bleeding to above-mentioned signs and symptoms in this case representation. (10)

Conclusion

Empty Sella syndrome is either idiopathic/inherited or due to secondary reasons as clinicalfinding and literature studies shows. Most of the patients who has progressive elevation in Intracranial pressure, incompetent diaphragm sella, obesity, multiple pregnancies, head surgeries, and idiopathic intracranial hypertension. It is usually found in obese patients, predominantly in females. the differential diagnosis for migraine, pituitary tumor, hormonal imbalance, meningitis, some viral infections, and other disease conditions is always kept in mind while diagnosing empty sella syndrome.

Recent studies show antipituitary antibodies are also responsible for it, which causes the shrinkage of the pituitary gland, but the particular mechanism and reason for the development of antibodies against the pituitary gland is still a concern of the research.

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