A Case Of Idiopathic Intracranial Hypertension In Background Of Empty Sella Syndrome

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Abstract: Background: Empty sella syndrome is a rare disorder characterized by enlargement or malformation of a structure in the skull known as the sella turcica. Most individuals with empty sella syndrome do not have any associated symptoms, but the finding raises concerns about hormone deficiencies. Empty sella syndrome may occur as a primary disorder (idiopathic) or as a secondary disorder in which it occurs due to an underlying condition or disorder such as a treated pituitary tumor, head trauma, or a condition known as idiopathic intracranial hypertension (also called pseudotumor cerebri) during which elevated intracranial pressure causes empty sella syndrome. Here we present a case of 25 year married female having symptoms and signs of raised ICP and decreased secondary sexual characters with blood investigations showing hypothyroidism. Her MRI Brain showed features of raised intracranial pressure (ICP) suggestive of Idiopathic Intracranial Hypertension with partially empty sella. Patient was diagnosed as Idiopathic Intracranial Hypertension with partial empty Sella with hypothyroidism and hypogonadotrophic normogonadism secondary to raised ICP. Patient was treated with IV Mannitol, thyroxine was started according to weight and other supportive treatment. Patient consciousness improved and got better. Patient was discharged with follow up on oral acetazolamide, thyroxine and hormone replacement therapy. [Vaghela N Natl J Integr Res Med, 2024; 15(1): 54-56, Published on Dated: 26/01/2024]

Key Words: Sella Syndrome, Hypothyroidism, Idiopathic Intracranial Hypertension

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Introduction: Empty sella syndrome is a rare disorder characterized by enlargement or malformation of a structure in the skull known as the sella turcica. Most individuals with empty sella syndrome do not have any associated symptoms, but the finding raises concerns about hormone deficiencies. Empty sella syndrome may occur as a primary disorder, for which the cause is unknown (idiopathic), or as a secondary disorder, in which it occurs due to an underlying condition or disorder such as a treated pituitary tumor, head trauma, or a condition known as idiopathic intracranial hypertension (also called pseudotumor cerebri) during which elevated intracranial pressure causes empty sella syndrome[1].

Case Study: A 25 year married female presented with headache, blurring of vision, anasarca, for 10 days and decreased level of consciousness since 1 day. No complain of focal neurological deficit, decreased urine output, hematuria, vomiting, convulsion, fever, perspiration, syncope, jaundice and trauma. No history of previous any medical illness.

Obstetric History: P0A0L0, Menses irregular (2-3 month), scanty (2-3 days)

Family History: No significant family history. On general examination patient was drowsy, short stature with sparse axillay and pubic hair and cold clammy dry skin.

Vitals: Temp: cold, Pulse: 55 bpm, BP: 140/90 mmHg

RS: BAE clear, CVS: S1S2 +, CNS: Plantar B/L extensor, Reflexes Superficial and deep reflexes diminished.

Investigations: Hb: 10.4 g/dl, Hematocrit: 42%,

TLC: 10680/cumm, Platelet Count: 2.1 lac/cumm.

Creatinine: 0.7 mg/dl, Urea: 14 mg/dl, Na+: 136 mEq/L, K+: 3.8 mEq/L, RBS: 92 mg/dL

Total Protien: 6.7 g/dl and Albumin: 3.2 g/dl,

CRP: 0.6 mg/dl, ESR:17 mm/hr.

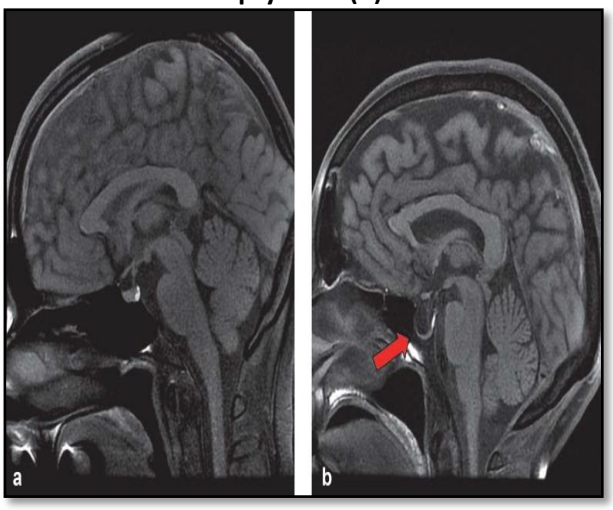
Sr. TSH: 70 mIU/L, F. T3: 0.08 nmol/L, F. T4: 38 nmol/L Sr. Cortisol: Normal

Urine Analysis: NAD, ANA titre: Negative

Fundus Examination: Early changes of papilloedema.

MRI Brain: Features of raised ICP suggestive of Idiopathic Intracranial Hypertension. Partially empty sella with residual height of pituitary gland is 3.8 mm. No definite focal enhancing leison.

Image 1: Cerebral MRI Scan Of Normal Sella Turcica With Regular Pitutary Gland (a) And Empty Sella (b)



Patient was diagnosed as Idiopathic Intracranial Hypertension with partial empty Sella with hypothyroidism and hypogonadotrophic normogonadism secondary to raised ICP due to altered pitutary function resulting in hormonal imbalance.

Patient was treated with IV Mannitol, thyroxine was started according to weight and other supportive treatment. Patient conciousness improved and got better. Patient was discharged with follow up on oral acetazolamide, thyroxine and hormone replacement therapy.

Discussion: We present a case of Idiopathic Intracranial Hypertension with partial empty Sella with hypothyroidism and hypogonadotrophic normogonadism.

The exact role that defects in the diaphragma sella play is the development of primary empty sella syndrome is unknown.

In some affected individuals a tear in the diaphragma sellae allows the underlying membranes to push through (herniate), which allows cerebrospinal fluid to leak out and accumulate in the sella turcica. The pressure exerted by the fluid can flatten or enlarge the sella turcica. Consequently, the pituitary becomes compressed and flattened as well. These results indicate a presence of hypothalamic or pituitary stalk dysfunction[2].

Some studies suggest autoantibodies that recognize antigens in mouse pituitary and cerebrum were detected in the patient's serum by immunobloting analysis that suggested autoimmune mechanisms were involved in the impairment both of hypothalamus and pituitary[3].

Generally, hyperprolactinemia is frequently observed in empty sella syndrome with stalk dysfunction. Low concentration of fT4 and slightly elevated levels of TSH, and delayed response of TSH to TRH administration suggest hypothalamic dysfunction or TRH deficiency.

In addition, polyuria and limited elevation of urinary osmolality during hypersaline test suggest a presence of partial central diabetes insipidus.

Taken together, it is strongly suggested that the panhypopituitarism is caused by hypothalamic dysfunction[4]. Although, several studies have reported hypothalamic insufficiency with empty sella in children, it is extremely rare in adulthood.

In children, congenital deficiency or structural anomaly in the hypothalamus has been suggested as a cause[3]. However, the cause of hypothalamic dysfunction with empty sella syndrome in adulthood has been totally unknown.

Conclusion: In summary, we present a case of Idiopathic Intracranial Hypertension with partial empty Sella in adulthood.

Endocrinological findings strongly suggest hypothalamic dysfunction in form of hypothyroidism and hypogonadotrophic normogonadism secondary to raised ICP.

Patient responded well to ICP lowering drugs and was discharged with follow up with oral acetazolamide, thyroxine and hormone replacement therapy.

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