

Empty sella syndrome: A case report

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Abstract:

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Empty sella syndrome (ESS) is a symptom complex characterized by dull headache, vomiting, occasional giddiness and associated visual field defects along with an enlarged sella. We present here a 51-year-old obese female patient having persistent headache and endocrine abnormalities who was diagnosed to have empty sella syndrome.

Keywords: Empty sella syndrome, endocrine abnormalities, visual field defects.

Introduction:

Empty sella syndrome (ESS) is a disorder that involves the sella turcica, a bony structure at the base of the brain that surrounds and protects the pituitary gland (1). Empty sella syndrome (ESS) is a condition in which the sella turcica is partially or completely filled with CSF resulting in a displacement of the normal pituitary gland (2).

Case History:

A 51-year-old, obese, female patient known case of hypothyroidism since 3 years on regular treatment, attained menopause 2 years before, came to our hospital with complaints of headache, easy fatigability, feeling of uneasiness, milk discharge from both breasts and mental depression since last 2- 3

years. She has been taking treatment for headache from other general practitioners since last 3 years. She was on Tab. Thyrox 100 mcg OD and on analgesics as required. She was diagnosed as a case of migraine. She had three children - two sons and one daughter. There was no history of any other co morbid disease. There was no history of any addictions.

On examination, she was obese (BMI = 26.4), vitals were stable, breasts were normal and there was no neurodeficit. Visual fields were normal. Rest systemic examination was within normal limits.

On investigations, her MRI brain was suggestive of marked thinning of pituitary gland with widening of sella filled with CSF. Findings were suggestive of empty sella syndrome.

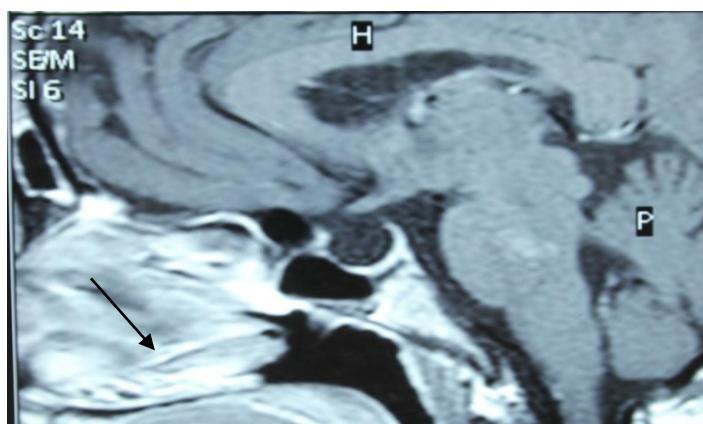


Figure 1: MRI Brain- Arrow showing the empty sella filled with CSF

Hormonal studies were done which showed serum TSH was 1.46 (normal 0.27-5.1), serum Prolactin levels were 84.95 ng/ml (normal 4.79- 23.3), serum growth hormone levels were below 0.05 (normal 0-8), serum testosterone levels were below 0.02 (normal 0.029- 0.408), serum LH levels were 14.58 (normal 7.7- 58.5) serum cortisol levels were 12.23(normal 6.2-19.4) and serum ACTH levels were 6.01 (normal 0- 46).

Her serum cholesterol levels were 250 mgm% (normal 135- 250mgm %), triglycerides were 416 mgm% (normal 60- 150), serum HDL was 62.5 mgm%(normal 30-95 mgm%). Rest of the investigations was within normal limits.

Thus the diagnosis of Empty sella syndrome was made. She was started on Tab. Cabergoline 0.5 mg once a week, Tab Thyrox 100 mcg OD, Calcium

and vitamin D supplements, Tab. Atorvastatin 20 mg HS. Dietary advice was given and patient was discharged to be on regular follow up.

Diagnosis:

Imaging: The lateral radiograph of the skull may reveal a normal sized sella or it may be enlarged. The typical empty sella shows “symmetrical ballooning”, that is, a smoothly curving regular cavity of the sella. Computerised tomography scans will show the pituitary fossa to be occupied largely by substance of CSF or water density rather than a normal gland.

Magnetic resonance imaging (MRI) can readily confirm the diagnosis of an empty sella. On T1 sagittal MR images, extension of CSF into the sella is easily identified and remaining gland is compressed along the floor. Typical central position of the infundibulum is a useful sign in an empty sella which helps to rule out a cystic lesion in the suprasellar region (3). MRI will demonstrate the sella to be filled with CSF and the infundibulum can be seen to traverse the space, thereby excluding a cystic mass. This is known as the infundibulum sign (5).

Hormonal profile:

To establish the diagnosis of hypopituitarism (partial or complete) and to determine the hypophyseal cause of dysfunction, measurement of following hormones are required – serum thyrotropin and T4, corticotropin and cortisol, LH and testosterone, basal and stimulated growth hormone and prolactin level. Usually, the hormonal profile is normal in patients with empty sella. However, mild hyper-prolactinaemia (usually <100 µg/l) with or without galactorrhoea occurs in approximately 15% of patients.

Complications:

- a) In view of the increased intracranial pressure and traction over the optic chiasma caused by the post-surgical adhesions, visual field defects are known to occur and often require treatment.
- b) Cerebrospinal fluid rhinorrhoea can occur presenting as a non-traumatic and persistent nasal discharge.
- c) Long standing increase in the intrasellar pressure can also lead to pituitary dysfunction, which needs to be corrected with appropriate hormone replacement therapy.

Treatment of empty sella syndrome:

The primary empty sella is generally asymptomatic and incidentally detected, it requires no specific treatment. However, when it is accompanied

by endocrine dysfunction, replacement therapy of the appropriate target gland hormones would be required. Presence of CSF rhinorrhoea may require surgical correction.

In secondary empty sella, adhesions are formed between the diaphragma sellae and the chiasma. Retraction of these adhesions due to reduction in tumour size may pull the chiasma into the empty sella resulting in visual field defects. This needs to be corrected by chiasmopexy. Packing the sellar cavity after transphenoidal removal of the pituitary tumour can also be done to avoid herniation of the chiasma into the surgical empty sella (3).

Discussion:

The empty sella turcica is defined as an intrasellar herniation of the suprasellar subarachnoid space with compression of the pituitary gland producing in many cases a remodelling of sella which results from a combination of incomplete diaphragma sellae and an increased CSF fluid pressure(3). There are two types of ESS: primary and secondary. 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. The primary empty sella syndrome is generally found in middle-aged women who are obese and hypertensive (4). 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 (1).

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