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Case Report

Partial Empty Sella Syndrome in a Middle Aged Female-A Case Report

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Abstract

Empty sella syndrome is a condition where there is shrinkage or flattening of the pituitary gland that leads to filling of the sella turcica with CSF either partially or completely. It is usually asymptomatic and is an incidental finding. When symptomatic, it can present with endocrine abnormalities, headache and visual field defects. Here we report the case of a 40 year old multiparous, obese, hypertensive female who presented with persistent headaches, menstrual irregularities and was diagnosed to have empty sella syndrome on the basis of radiological features. Hormonal assays revealed high prolactin levels. She was managed conservatively and regular follow up was advised.

Keywords: *Empty Sella; Obese Female; Headache*

Introduction

Empty sella syndrome is a condition where the sella turcica is deformed or enlarged and is partially or completely filled with csf. The term empty sella is a misnomer as the sella is not empty and a pituitary gland is always present though may be displaced downwards due to excessive csf pressure [1]. It is more common in middle aged obese women and increases with age [2].

The presence of incomplete sellar diaphragm is a prerequisite for its development. It can be classified into two types. Primary empty sella which occurs either due to the congenital anatomical defect or factors leading to the increased CSF pressure causing herniation of arachnoid membrane into pituitary fossa [3]. Most patients with primary empty sella are generally asymptomatic and this is found as an incidental radiological finding. Common symptoms are of headaches due to increased CSF pressures, visual symptoms due to arachnoidal adhesions and traction on optic apparatus and endocrine symptoms [4]. Secondary empty sella can be due to iatrogenic causes like radiation or pituitary surgery and non-iatrogenic causes like reduction in the size of pituitary gland due to physiological involution, sheehan's syndrome or primary organ failure (thyroid, adrenal, gonads).

Case Report

A 40 years obese multiparous lady presented with history of intermittent episodes of headache accompanied with nausea since 1 year which has aggravated since last 2 days. She gives history of menstrual irregularities and sleep disturbances since last 1 year. There was no associated history of vomiting, fever and visual disturbances. No history suggestive of galactorrhea. On examination, her BMI was 34.7 kg/m^2, BP 150/90 mmhg, heartrate was 80/minute. She was pale with no lymphadenopathy. Thyroid gland was normal. Neurological, cardiological examination were normal. Ophthalmological examination revealed normal vision and fundus. ECG was within normal limits. MRI

brain was done which revealed presence of partial empty sella with pituitary gland normal in morphology (Figure 1 and 2). Her pituitary hormonal assays were done which revealed FSH 6.09 mIU/ml(normal), high prolactin levels of 42.37 ng/ml (normal range 2.80 - 29.20 ng/ml), normal cortisol levels 74.32 and HbA1c of 6.4 gm%. She was treated symptomatically for her headache to which she responded well and was advised regular follow up.

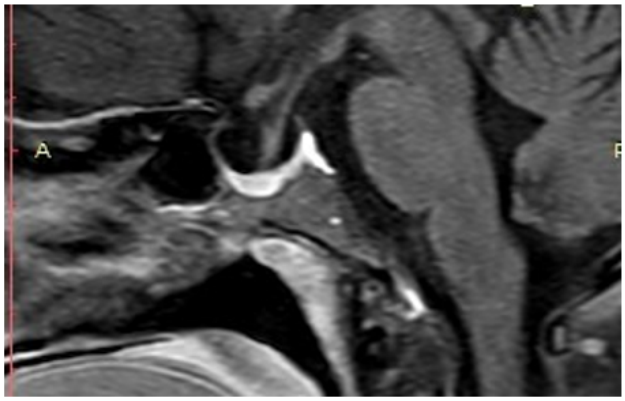

Figure 1: *MRI brain showing presence of partial empty sella.*

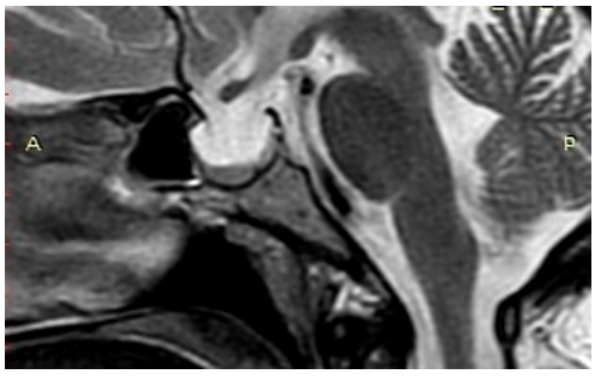

Figure 2: *MRI brain showing presence of partial empty sella.*

Discussion

Empty sella syndrome is a radiological entity which requires an anatomical defect of an incompetent diaphragm sella. Total absence of diaphragm sella is present in 20.5% of normal subjects [1]. It is said to be partial if less than 50% of the sellar space is filled with CSF or complete if more than 50% space is filled with CSF and pituitary gland thickness is less than 2 mm [2]. In primary empty sella, incompetent diaphragm sella being the prerequisite, there is a constant or intermittent rise in the CSF pressures which allow the subarachnoid space to be forced into the pituitary fossa. Whenever there is a rise in intracranial pressure, it leads to accentuated suprasellar cistern pressure and the CSF pulsations fall directly onto the pituitary gland. Rise in CSF pressure can be due to obesity, sleep apnea, hyperten-

sion, pregnancy and labor. Intracranial hypertension is associated with tumours, venous thrombosis, infections and malformations [2]. The elevated CSF pressure along with the incompetent diaphragm forces the subarachnoid space into the sella due to hydrostatic pressure and CSF pulsatile movement [1]. Primary empty sella is very common in females above the age of 40 yrs and is associated with obesity, hypertension and migraine [3]. Women of child bearing age can present with menstrual irregularities, galactorrhea and sterility while men may have complaints of gynaecomastia and sexual disturbances [1]. During pregnancies, there is increase in the size of the pituitary gland leading to weakened diaphragm and herniation of CSF into the sella [4]. After menopause, there occurs reduction in pituitary volume and so explains the occurrence after the age of 40 years. Obesity leads to obstructive sleep apnea causing hypercapnia and increased CSF pressure predisposing to empty sella. Pseudotumour cerebri is one such cause occurring due to increased CSF production and capillary permeability and impaired CSF absorption [5]. The prevalence of ESS in idiopathic intracranial hypertension ranges from 70 - 94% [6]. Headache is the most common symptom present in 80% of patients. In secondary empty sella, there are iatrogenic causes like pituitary surgery, irradiation and non-iatrogenic causes like infection, infarction. The predominant complaint here is visual abnormality like diplopia, blurred vision, optic neuritis due to arachnoidal adhesions and traction on the optic apparatus. Endocrine abnormalities like panhypopituitarism, secondary hypogonadism, hyperprolactinemia, isolated ACTH deficiency and diabetes insipidus (DI) have been reported. Ghatnati., *et al.* described endocrine dysfunction in 50% of PES patients [7]. Hyperprolactinemia is the most common one [8]. The most probable reason is remodeling of the hypothalamo-pituitary region and altered CSF dynamics leading to pituitary stalk compression. Degree of raised prolactin levels is usually less than 100 ng/ml in ESS and more than 200 ng/ml in prolactinomas. If pituitary is normally functioning, there is no medical treatment required. If high prolactin interferes with functioning of ovary or testes, drugs lowering prolactin levels are given. In a study by Atmaca, 29% patients had diabetes insipidus and thirst was increased in all [9]. For secondary empty sella, replacement of the lacking hormones is done. Dutta., *et al.* described a male diagnosed with empty sella along with symptoms of hypothyroidism which got reverted with levothyroxine therapy [10]. In obese patients, weight reduction plays an important role in alleviating symptoms. Visual disturbances and CSF rhinorrhea are the only indications of surgery. Sinus bradycardia has also been found to be an unusual and rare presentation of empty sella which signifies the importance of keeping this as one of the differentials while evaluating causes of bradycardia [11]. Lennon., *et al.* reported a case of empty sella syndrome in a woman with cerebral venous sinus thrombosis as a rare presentation of polycythemia vera [12]. On MRI brain, CSF filling is seen in continuity with subarachnoid space and pituitary gland is seen flattened against the sellar floor. Indirect signs of intracranial hypertension may be seen like prominent subarachnoid spaces along the optic nerve, flattened posterior sclera, vertical tortuosity of optic nerve sheath and increased width of optic nerve sheath [13]. Our patient had an incidentally detected PES most probably due to her multiparity, age and obesity. Hormonal assays revealed hyperprolactinemia which was treated conservatively.

Conclusion

ESS is a rare entity which occurs due to elevated CSF pressures causing herniation of CSF into the sella. It should be kept as a differential diagnosis in women who are multiparous and obese presenting with headaches, visual disturbances and endocrine abnormalities. Hormonal replacement if indicated along with weight reduction is the mainstay of management. Surgical correction is required when visual disturbances and CSF rhinorrhea are present.

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