

Empty Sella Syndrome

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ABSTRACT

An Empty Sella occurs due to herniation of the arachnoid through an incompetent diaphragma sellae. Over time, cerebrospinal fluid (CSF) pulsations may enlarge the sella and compress the gland against the floor of the sella. Empty Sella Syndrome is a considered a less common entity and is usually asymptomatic and an incidental finding. However, it can be a manifestation of increased intracranial pressure and can be occasionally severe. Compression of the pituitary gland may affect function, or traction on the optic chiasm and nerves may cause visual symptoms. An empty sella may be classified as primary when this occurs in persons who have not received pituitary radiation or pituitary surgery, while an empty sella discovered following such procedures is classified as secondary empty sella. We had a 41 year old patient who came to us with symptoms of headache and left sided hemi-sensory disturbance. An evaluation revealed multiple comorbid illnesses with MRI showing features of Empty Sella.

Key words: Empty Sella Syndrome, Empty Sella, Pituitary Dysfunction

INTRODUCTION

Empty Sella Syndrome (ESS) is a condition in which the sella turcica is partially or completely filled with CSF. This results in a displacement of the normal pituitary gland¹. Usually the sella is enlarged and the pituitary gland is compressed and reshaped². ESS is divided into primary and secondary types depending on the presence of previous surgery or irradiation to the pituitary gland³. Primary empty sella is usually due to an inherent weakness of the diaphragma sella and or to an increase in the intracranial pressure which promotes the herniation of arachnoid membrane into the pituitary fossa⁴. Although most individuals who have primary ESS are asymptomatic, a few present with clinical symptoms and signs related to the condition. It is more common in middle-aged obese females, usually presents with headache, and only occasionally associated with endocrine or visual abnormalities⁵. Surgical therapy is rarely required except for cases presented with CSF rhinorrhea or progressive visual loss⁶. Secondary Empty Sella (SES) may be caused by pituitary adenomas undergoing spontaneous necrosis (ischemia or hemorrhage). Other causes known to cause SES are infective, autoimmune, traumatic, radiotherapy, drugs, and surgery. Numerous studies have shown that empty sella syndrome may be associated with pituitary dysfunction contrary to the notion of it being an incidental finding⁷. In this report we present a patient who came to us with left sided hemisensory disturbance and headache and had an associated incidental finding of Empty Sella Syndrome.

CASE REPORT

A 41 year old obese lady came to us with numbness of left side of the body (Face, Left Upper Limb and Left Lower Limb) since 6 years. Symptoms are intermittent and are associated with intermittent headache and neck pain. She also gives history of left facial weakness six years ago which recovered gradually. There is no weakness of extremities or imbalance. She also complained of visual blurring of recent onset. Her menstrual history was normal. On examination her Blood Pressure was 140/80 mm of Hg and heart rate was 80 beats per minute. Cardiovascular and respiratory system examination was normal. Central Nervous system examination was unremarkable too.

An MRI Brain was done here which revealed presence of Few Lacunar Infarcts in the high parietal area and also Empty Sella (Figures 1a and 1b).

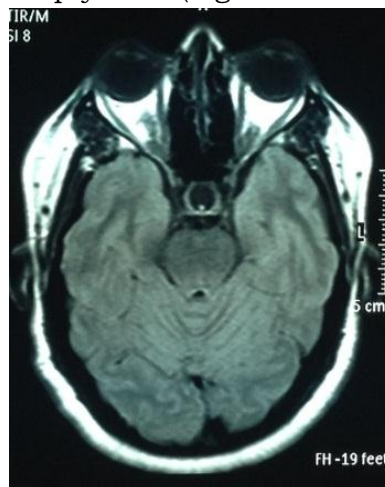


Figure 1a: MRI Brain (T2 FLAIR) showing Empty Sella

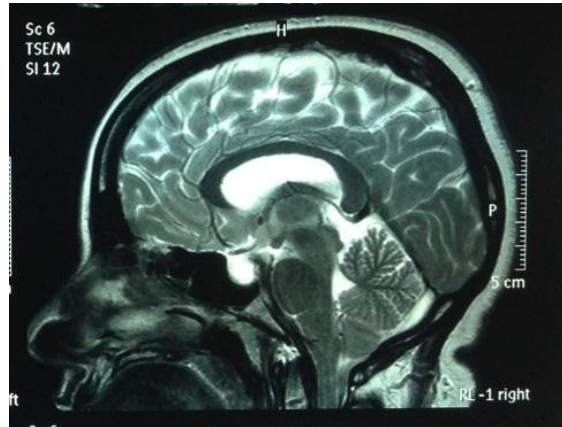


Figure 1b: MRI Brain (T2 Saggital View) showing hyper intense signal intensity in the region of Sella Turcica suggestive of CSF filled Sella.

An Ophthalmologic evaluation revealed normal vision with mild bilateral optic disc edema (Figure 2a and 2b). Conservative management was advised and patient has been asked to follow up after 1 month for repeat fundoscopy.



Figure 2a – Left Eye



Figure 2b – Right Eye

Figure 2a, 2b: Fundoscopy Image Showing Optic Disc Edema in Both the eyes

In view of the finding of Empty Sella, pituitary hormonal studies were recommended. The TSH was 1.18 micro IU/mL (Normal Range: 0.27 – 4.2), Prolactin level was 128.9 micro IU/mL (Normal Range: 102 – 496), Follicular Stimulating Hormone level was 10.06 micro IU/mL and Lutenising Hormone (LH) level was 3.59 micro IU/mL. All were within normal limits. Serum 25-OH Vitamin D Level was 13.8-ng/mL (Insufficient: 10 – 29). She was also detected to have Impaired Post-prandial Blood Glucose and mild Dyslipidaemia.

She is currently under conservative treatment with Vitamin D Supplements and Statins/Anti-platelets for Infarcts. A close follow up was recommended to evaluate the optic disc edema.

DISCUSSION

The term ESS describes a distinct radiological and anatomical entity in which the subarachnoid space extends significantly through an incompetent diaphragma sella into the sella turcica⁷. In normal individuals, the diaphragm sella almost completely covers the

pituitary body. There is only a small central opening for the passage of the infundibulum. The term empty sella is an incorrect representation for this condition, because in these cases the sella is not empty. It is rather completely filled by the pituitary gland with its stalk, the arachnoid, the CSF and occasionally, the optic system and the third ventricle. That is why some authors prefer to use the term intrasellar arachnoidocele because it expresses in a simple and clear way the findings in this entity⁴. The condition is more common and benign in adults, with a female predominance, but its occurrence in children has been reported^{8,9}. It has been found in association with several conditions, such as obesity, hypertension, and migraine¹⁰. A number of hypotheses have been offered to explain the cause of primary ESS such as pituitary infarction, pituitary apoplexy, and rupture of an intrasellar cyst¹¹. Although one or more of these conditions may play role in the development of ESS, a reasonable explanation is that the condition arises in a patient who has either a transient or constant elevation in intracranial pressure and who has incompetent diaphragm sella that allows the subarachnoid space to be forced into the sella by the hydrostatic pressure and pulsatile movement of CSF¹². Secondary ESS is generally associated with a previous surgery, radiotherapy, or medical treatment for tumors of the sellar region^{1,3}. The reported prevalence of primary empty sella (PES) in general population is 8-35%¹³.

In a recent study, Ghatnatti et al.¹⁵ reported that PES accounted for most cases of empty sella and it was more commonly noted in females with higher parity like in our patient. Enlargement of the pituitary during pregnancy may lead to weakening of the sellar diaphragm, thus predisposing to herniation of cerebrospinal fluid into the sella.¹³ They also noted that obesity was more common among patients with PES. Obesity causes obstructive sleep apnea leading to hypercapnia and increased CSF pressure predisposing to empty sella. This could also be the contributory cause in our patient.

There is a wide variation in the reported prevalence of endocrine abnormalities in PES. Ghatnatti et al.¹⁵ noted endocrine dysfunction in 50% of PES patients while De Marinis *et al.*¹³ found endocrine abnormalities in 19%. Hyperprolactinemia was the most common endocrine abnormality observed. In PES, mild hyperprolactinemia has been frequently reported which can be explained by pituitary stalk compression as a consequence of the remodeling of the hypothalamo-pituitary region and altered CSF dynamics.

Surgical treatment for symptomatic ESS is controversial. Visual disturbances and CSF rhinorrhea are the main indications for surgery. When surgery is indicated, the type of surgery depends on clinical presentation and radiological findings. The surgical outcome of cases with ESS is favorable, as most patients report improvement or stabilization of their symptoms¹⁴.

CONCLUSION

Empty Sella Syndrome is an incidental finding which arises in a patient who has either a transient or constant elevation in intracranial pressure and who has incompetent diaphragm sella that allows the subarachnoid space to be forced into the sella by the hydrostatic pressure and pulsatile movement of CSF. It is usually seen in obese patients with slight female preponderance. It should be kept as a differential diagnosis for patients with non-specific headache especially in obese females.

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