Case report

# Empty Sella Syndrome: A case report

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**Abstract**

A 55 year old female patient presented with history of giddiness and fatigue of one year duration. She had pallor, diffuse hyperpigmentation of skin and mucosal surfaces, low blood pressure (90/60 mm Hg), hypoglycemia, hyponatremia (118 mEq/L), hypokalemia (2.7 mEq/L) and elevated serum ACTH levels (21 pmol/L) suggestive of hypocortisolism. On imaging, MRI Brain showed features suggestive of an empty sella. In the absence of any other inciting factors, a diagnosis of Primary Empty Sella (PES) syndrome was made. The patient was treated with levothyroxine, steroids, hypertonic saline and potassium supplements following which she recovered rapidly. An empty sella occurs due to herniation of the arachnoid membrane through an incompetent diaphragma sellae. Primary empty sella syndrome is considered as a less common entity and is usually asymptomatic and can be an incidental finding.

**Keywords:** empty sella, Empty Sella Syndrome, hypopituitarism, pituitary dysfunction

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## Introduction

Empty Sella Syndrome (ESS) is a symptom complex which is characterized by the presence of an enlarged sella turcica that is partially or completely filled with cerebrospinal fluid on imaging studies with symptoms like headache, giddiness, vomiting, visual field defects and endocrine abnormalities.[1]It can be either primary or secondary. Two factors are known to play a role in the development of primary empty sella syndrome, namely the inherent weakness associated with the diaphragm sellae, and the concomitant increase in the intracranial pressure which increases the chance of herniation of the arachnoid membrane into the pituitary fossa.[2] Primary Empty Sella (PES) syndrome is an incidental radiological diagnosis and usually remains asymptomatic. It is more commonly seen in middle-aged, obese females who usually present with headache and is occasionally associated with endocrine or visual abnormalities.[3] Secondary empty sella occurs in the background of previous pituitary surgery or cranial irradiation. It can also be due to hemorrhage or infarction of the pituitary gland in the postpartum period (Sheehan's syndrome).[3]

## Case presentation

A 55 year old lady presented with history of giddiness and fatigue since 1 year. She had a few episodes of vomiting for two days prior to admission. She had attained menopause 2 years back. Her obstetric score was P2 L2 and the postpartum period was unremarkable. On examination, she was found to be pale with no icterus. Her vitals were stable, except for a blood pressure of 90 / 60mm Hg. There was diffuse hyperpigmentation of the lips and tongue (Figure 1) and ichthyosis of the lower limbs (Figure 2). Fundus examination was normal.

**Figure 1: Diffuse hyperpigmentation noted over the tongue of the patient**

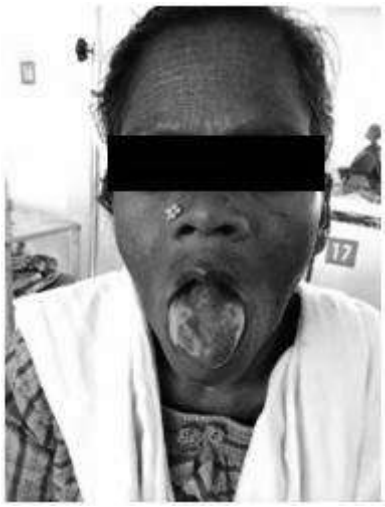
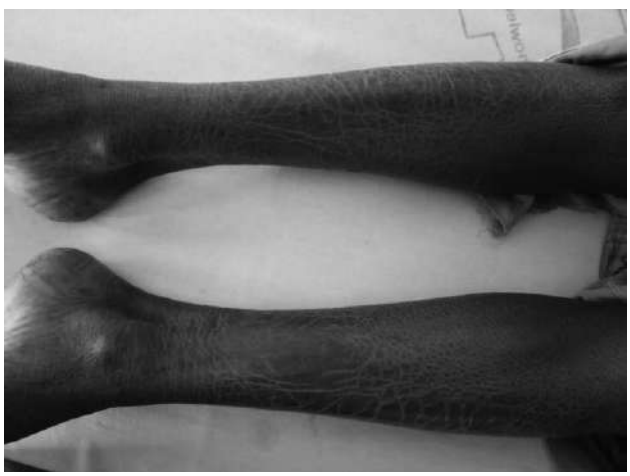


**Figure 2: Ichthyosis of the lower limbs**



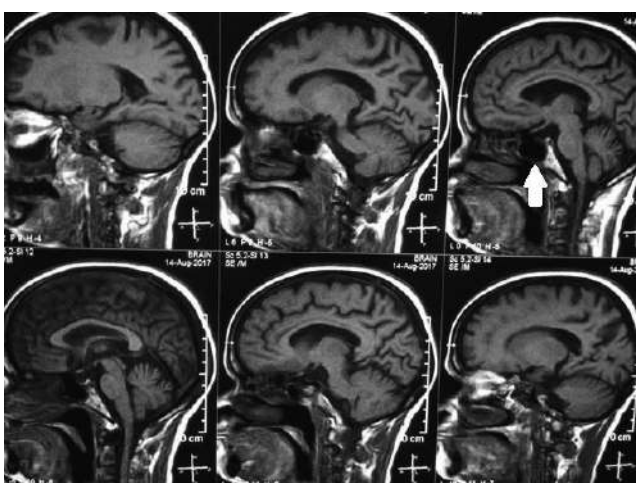
Systemic examination was found to be normal and there were no visual field defects. Complete blood counts and other biochemical blood investigations were carried out. Her hemoglobin concentration was 7.8 g/dL, serum sodium was 118 mEq / L and serum potassium was 2.7 mEq / L. Direct Coombs test was positive. She had repeated episodes of hypoglycemia which required correction with 25% dextrose infusion. All other blood investigations were within the normal range. Despite adequate electrolyte correction, her symptoms persisted and hence, she was subjected to further tests. The reports are as seen in Table 1.

**Table 1: Hormonal assays of the patient**

| Hormone | Values | Reference Range |
|---|---|---|
| Thyroid Stimulating Hormone (TSH) | 3.13 | 0.27 – 5.5 mIU/L |
| Free $T_4$ | 0.03 | 0.93 – 1.7 ng/dL |
| Free $T_3$ | 0.26 | 2.0 – 4.43 pg/mL |
| Cortisol (8 a.m.) | 10.2 | 3.7 – 19.4 µg/dL |
| Adrenocorticotrophic Hormone (ACTH) | 21 | < 20pmol/L |
| Prolactin | 1.41 | 4.79 – 23.3 ng/mL |
| Follicle Stimulating Hormone | 2.29 | 25.8 – 134.8 mIU/mL |
| Luteinizing Hormone | 0.68 | 7.7 – 58.5 mIU/mL |

MRI Brain showed marked thinning of the pituitary gland along the sellar floor, with the posterior pituitary appearing normal which was suggestive of partial empty sella (Figure 3). In the absence of the other precipitating events, the diagnosis of primary empty sella syndrome was made. The patient was treated with levothyroxine, intravenous hydrocortisone followed by oral prednisolone, hypertonic saline and potassium supplements. There was a rapid improvement in her condition and subsequently her blood pressure and sodium levels returned to normal with no further hypoglycemic episodes.

**Figure 3: MRI Brain of the patient showing Empty sella (white arrow)**



**Discussion**

Primary Empty Sella (PES) syndrome accounted for most cases of empty sella and it was more commonly noted in females with higher parity, as was the case in our patient.[4] Enlargement of the pituitary during pregnancy may lead to weakening of the sellar diaphragm, thus predisposing to herniation of cerebrospinal fluid into the sella.[5] This could also be the contributory cause in our patient. Obesity has also been found to be more common among patients with PES.[5] Obesity causes obstructive sleep apnea leading to hypercapnia and increased CSF pressure predisposing to empty sella.[6]

There is a wide variation in the reported prevalence of endocrine abnormalities in PES. Ghatnatti *et al.* noted endocrine dysfunction in 50% of their PES patients in Guwahati, India in 2012, while De Marinis *et al.* found endocrine abnormalities in 19% of their PES patients in northern and central Italy in 2005.[5, 6] Hyperprolactinemia was the most common endocrine abnormality observed in both the studies.[5, 6] In PES particularly, mild hyperprolactinemia has been frequently reported; this is has been attributed to altered CSF dynamics and the remodeling of the hypothalamo-pituitary region leading to a compression of the pituitary stalk.[7] Patients also have abnormalities of the thyroid and adrenal axis. This leads to features of hypocortisolism (low blood pressure, hyponatremia, hypokalemia) as seen in our patient. To the best of our knowledge, diffuse ichthyosis has not been reported previously in patients with primary empty sella syndrome.

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.[7] Treatment depends on the type of empty sella. There is no specific treatment for primary empty sella, as the patient is usually asymptomatic and pituitary hormones are in the normal range. For secondary empty sella syndrome, treatment involves replacing the hormones that are deficient.[8]

**Conclusion**

Empty sella syndrome may be an incidental finding in a patient who presents with headache or other symptoms. Empty sella syndrome should be kept as a differential diagnosis for patients presenting with non-specific persistent headache, fatigue and giddiness, especially in multiparous females. In our patient, headache was conspicuously absent and she had diffuse ichthyosis which was unique and not previously reported in literature. She also had no prior history of migraine or hypertension which are usual co-morbidities

**Acknowledgment:** The authors would like to acknowledge the contribution of Dr. Prasanna Karthik and Dr. Kannan for their help in the preparation of this manuscript.

**Conflicts of interest:** Nil

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