

Primary Empty Sella Syndrome: A Case Report

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ABSTRACT

Primary empty sella syndrome (PESS) is relatively rare and uncommon developmental disorder which involves the sella turcica and develops spontaneously. It presents as endocrine and visual disturbances, rhinitis, persistent intractable headache and rarely as idiopathic intracranial hypertension. It is characterised by herniation of subarachnoid space within the sella which is often associated with mild flattening of pituitary gland. We report a case of 35 year old obese female presenting with persistent intractable headache diagnosed to be a case of PESS.

KEYWORDS: Empty sella syndrome, Primary empty sella syndrome, Pituitary.

INTRODUCTION

Empty sella syndrome (ESS) is a rare disorder involving the sella turcica, which is a bony structure at the base of the brain and it protects and surrounds the pituitary gland. It is usually diagnosed as an incidental finding on radiological imaging done for evaluation of pituitary disorders¹. It is of 2 types – primary and secondary^{2,3}. Primary ESS is characterised by a small anatomical defect above the pituitary gland which increases pressure in sella turcica, ultimately making the pituitary gland to flatten out against sella turcica and displacement of the pituitary gland^{2,3}.

Secondary ESS occurs due to injury, surgery and radiation therapy so that the pituitary gland regresses in size within the sell aturcica cavity. Patients with secondary ESS usually present with symptoms pertaining to loss of pituitary functions like fatigue, intolerance to stress and infection as well as stoppage of menstrual periods.⁴ PESS occurs in obese females with multiple pregnancies.

CASE REPORT

A 35 year old obese multigravida woman presented with persistent intractable headache, easy fatigability, diminished vision and history of seeing small insects like images in the left visual field since 2 months. Dimness of vision was more marked in the left eye than right eye. There was no history of vomiting, photophobia and neck rigidity. She denied history of galactorrhoea before first pregnancy. There was no history of any other comorbid disease or addictions. She had four children, two sons and two daughters. Her menstrual history and breast development was normal. She had normal secondary sex characters. She was taking frequent pain killers for headache as prescribed by a general practitioner.

Ophthalmological examination revealed her visual acuity in the left eye as perception of light (PL) projection of rays (PR) present only.

The right eye had 6/6 vision with glass. Fundus examination showed myopia with optic atrophy in the left eye while the right eye was normal. Intraocular pressure was 17 mmHg and 14 mm of Hg in the left and right eye respectively. On physical examination, BMI was 28 kg/mm², BP 130/80 mmHg, Pulse 72 beats/min. Cardiovascular, abdominal, respiratory and central nervous system examination revealed no abnormality. Ultrasound examination, ECG and X-ray chest was normal. Laboratory investigations including hemogram, renal, liver and lipid profile, serum electrolytes were within normal limits.

Thyroid function tests revealed normal T₃ and T₄ with TSH 7.5 uIU/ml (N 0.4-5 uIU/ml) while evaluation of other hormones like growth hormone, prolactin, ACTH, LH and FSH were within normal limits. MRI brain focussing on the pituitary region revealed cerebrospinal fluid (CSF) density area in the sellar region which was compressing the pituitary gland, suggesting empty sella. (Fig 1) Based on history, clinical findings and MRI brain, a diagnosis of ESS was made. The patient was put on thyroxine 25 microgram and advised weight reduction and supportive measures. However she was lost on follow up.

DISCUSSION

The empty sella turcica is defined as an intresellar herniation of the suprasellar subarachnoid space with compression of the pituitary gland producing in many cases a remodelling of sella which results from increased CSF pressure as well as partial diaphragmsellae.⁵

PESS patients usually manifest as intractable persistent headache, endocrine and visual abnormalities and intracranial hypertension^{6,7}. Our patient presented with intractable headache and dimness of vision along with mild hypothyroidism. PESS is usually asymptomatic, incidentally detected and requires no specific treatment. If there are features of endocrine dysfunction, appropriate hormone replacement may be indicated. Presence of CSF rhinorrhoea may require surgical therapy. PESS may occur in many endocrine autoimmune diseases. PESS itself has been suggested to be due to lymphocytic hypophysitis suggesting that autoimmunity per se may not be significantly linked to the occurrence of PESS⁸. Pregnancy may induce PESS by enlarging the pituitary gland especially in case of multiple pregnancies. This may result in herniation of

subarachnoid space in case of hypoplastic diaphragm sellae and or cerebrospinal fluid hypertension^{3,7}. Our patient is also female in compatible with other studies³. PESS is more common in females with incidence of 5.5% to 23% in autopsy studies of sella region and 8-35% of general population². Various workers have reported pan hypopituitarism in PESS⁷. Obesity may be responsible for PESS by inducing hypercapnia which can lead to chronic CSF pressure elevation. This may further result in intrasellar herniation of the suprasellar subarachnoid space³. Our patient was also obese with BMI of 28 kg/m². MRI commonly confirms the diagnosis of empty sella. MRI can demonstrate the sella to be filled with CSF and the infundibulum can be seen to traverse the space, thus ruling out a cystic mass. This is known as the infundibulum sign.⁹



Fig 1: MRI Brain focussing on pituitary region showing findings of empty sella.

CONCLUSION

Any patient complaining of persistent intractable headache, dimness of vision, endocrine abnormalities and obesity must be screened for empty sella syndrome. Early recognition and management of such cases may improve the outcome.

REFERENCES

1. Mc Lachlan MSF, Williams ED, Doyle FH. Applied anatomy of the pituitary gland and fossa : radiological and histopathological study based on 50 necropsies. Br J Radiol 1968; 41: 782-88.
2. Bergland RM, Ray BS, Torack RN. Anatomical variations in the pituitary gland and adjacent structures in 225 human autopsy cases. J Neurosurg 1968; 28: 93-9
3. Jordan RM, Kendall JW, Kerber CW. The primary empty sellasynndrome : analysis of the clinical characteristics, radiographic features, pituitary function and cerebral fluid adeno-hypophysial concentrations. Am J Med 1977; 62: 569-80.
4. Hossain MS, Mumu MA, Moyenuddin PK. Primary empty sella syndrome:Acase report. Anwer Khan Modern Medical College Journal 2010;1(1):23-25.
5. Agrawal JK, Sahay RK,Bhutada SK,Reddy VS, Agrawal NK.Empty sella syndrome. Indian Academy of Clinical Medicine. 2001;2(3)198-02.
6. Becejac B, Vizner B, Berkovic M, Gnjudic Z, Vrkljan M. Neuroendocrinological aspects of primary empty sella. CollAntropol 2002; 26 (suppl) : 159-64.

7. Bianconcini G, Bragagni G, Bianconcini M. Primary empty sella syndrome : observations on 71 cases. *Recent Prog Med* 1999; 90: 73-80.
8. Thomas R, Cupps, Paul D. Primary empty sella syndrome with panhypopituitarism. *Diabetes insipidus and visual field defects. ActaEndocrinologica* 1978 ; 89(3) : 445-60.
9. Houghton VM, Rosenbaum AE, Williams AL. Recognizing the empty sella by CT: The infundibulum sign. *Am J Roentgenol* 1981; 136(2):293-5.

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