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Reproductive Outcome in a Rare Case of Empty Sella Syndrome

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Abstract: *Empty sella refers to radiological appearance of enlarged or deformed sella turcica that is partially or completely filled with cerebrospinal fluid. It is of 2 types – primary and secondary. Present case report refers to a middle aged female presenting with persistent headache and having obesity, hypertension, irregular menstrual cycles and infertility.CT scan revealed Empty sella turcica.*
Keywords: *ESS, Sella turcica.*

I. Introduction

The term "empty sella" was first coined by Busch in 1951. Empty sella syndrome refers to the radiological appearance of an enlarged or deformed Sella Turcica that is completely or partially filled with Cerebro spinal fluid. The Empty sella syndrome is defined as an intra-sellar herniation of the suprasellar subarachnoid space with compression of the pituitary gland, which becomes flattened or may appear shrunken.

Classification:
1) Primary ESS – Those without an antecedent cause.
2) Secondary ESS – With an identifiable cause.

Clinical features:

The symptoms of ESS may vary from one person to another. Many patients are asymptomatic and endocrinologically normal. Some may be associated with hypopituitarism and hyperprolactinemia. Most common presenting symptom is chronic headache. Sometimes there may be associated visual disturbances, CSF rhinorrhoea, and benign intracranial hypertension. In females, there may be irregular or absent menstruation, galactorrhoea and fatigue.
Primary ESS has strong female predilection with obesity.

Investigations: X-ray, CT scan, MRI
MRI is the modality of choice for confirming the diagnosis. Sella is filled with CSF and infundibulum can be seen traversing the space (Infundibulum sign).

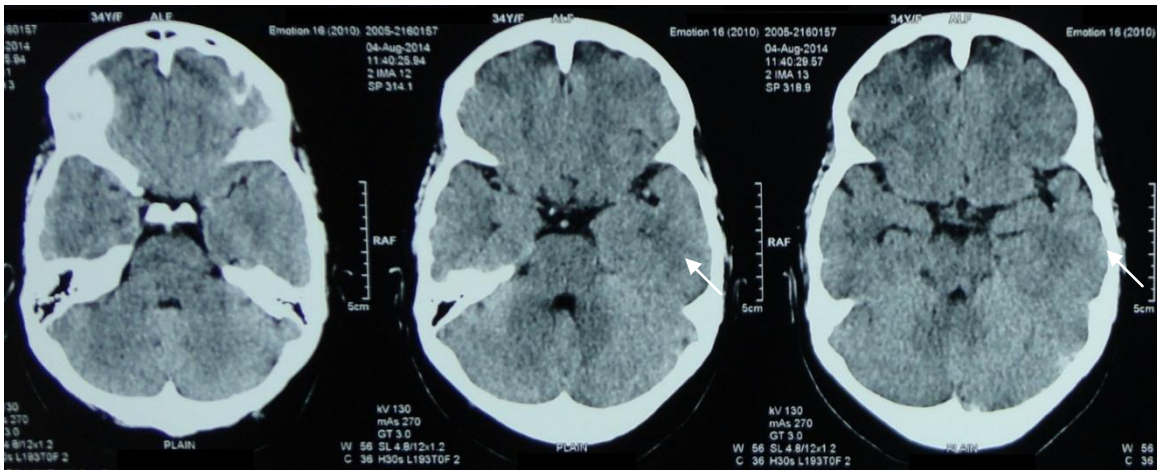
II. Case Report

A 34 year old female, P1 L1, having irregular menstrual cycles, presented with history of heavy bleeding with passage of clots, and pain abdomen since 1 week preceded by 5 months of amenorrhoea. Patient is obese (Weight = 105 kg, Height = 160 cm, BMI = 41). Pulse rate – 96/min, Blood Pressure = 140/100 mm Hg, Heart and lungs are normal. Per- abdomen – Fatty abdominal wall, Per-speculum – Cervix normal with bleeding from inside the os, Per-vaginal examination – Exact size of the uterus could not be made out due to obesity, Cervical os closed.

Investigations:

Complete blood picture, bleeding time, clotting time – normal, Random blood sugar, Serum creatinine, Serum electrolytes – within normal limits, T3, T4, TSH – normal, FSH, LH – normal , Prolactin – mildly elevated (34.7 ng/ml, Normal – 3.4-24.1 ng/ml)
Ultrasonogram revealed enlarged uterus with single intrauterine gestational sac and fetal pole. Crown Rump Length – 25 mm, Gestational age 9 weeks 1 day with no cardiac activity. Both ovaries normal. No ascites, other abdominal organs normal. Impression – missed abortion.

CT scan of brain:



Menstrual history:

Age of menarche – 14 years. Irregular cycles, 7-10 days/5-6 months. Heavy flow, no dysmenorrhoea. History of withdrawal bleeding after using hormones without any normal menstrual cycles. Oral Contraceptive pills were used for several years, on and off.

Marital life – 9 years. Conceived 5 years after marriage, she has taken treatment for infertility with ovulation induction drugs (Gonadotrophins). Pregnancy maintained with supportive progesterone therapy till term and delivered by caesarian section under spinal anesthesia. Male child, birth weight 3 kgs with good APGAR. Post-operative period uneventful. Lactated for 2 years.

Past history: Patient had reeling sensation, chronic headache, with heaviness of body since a long time for which she was evaluated by the physician. All routine investigations were within normal limits. CT scan was performed for persistent headache which revealed an empty sella turcica. Fundoscopy normal.

Treatment: Suction-evacuation of the products of the conception followed by check curettage done under local anesthesia. Procedure uneventful. Patient was advised follow-up.

III. Discussion

Primary ESS is of unknown etiology. Presence of a congenital defect in the diaphragm sellae may play a role in the development. Secondary ESS is caused by a variety of different conditions like injury or trauma to head, pituitary tumors, infection, radiotherapy, surgery on pituitary and rare conditions like Sheehan's syndrome. Primary ESS affects women more often than men. Most often detected in middle aged women who are obese and may have hypertension. In females, inspite of anovulation with prolonged menstrual cycles, they may conceive with ovulation induction drugs (Preferably gonadotrophins) followed by supportive therapy for maintenance of pregnancy. In the present case, she has conceived with gonadotrophin treatment in the 1st pregnancy, continued to full term with progesterone support. She was unaware of second pregnancy due to irregular menstrual cycles (spontaneous conception) and did not receive any hormonal therapy during the present pregnancy.

IV. Conclusion

This is a rare case of primary ESS diagnosed in a middle aged female presenting with menstrual abnormalities, infertility, obesity, hypertension and mild hyperprolactinemia. Conceived with gonadotrophin treatment. Primary ESS may be asymptomatic and usually does not affect life expectancy. It is discovered incidentally during an MRI/CT scan of head and brain. In case of secondary ESS, symptoms are related to the cause of pituitary gland disease and hormone deficiency.

Since the advent of non-invasive radiological techniques, primary ESS is being diagnosed with increasing frequency. As the incidence of pituitary dysfunction is high, patients with ESS should be evaluated periodically.

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