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Primary Empty Sella Syndrome Presenting with CSF Rhinorrhea: Successful Endoscopic Repair

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ABSTRACT

Primary empty sella syndrome (ESS) is characterized by herniation of the subarachnoid space into the sella turcica with pituitary flattening. While usually asymptomatic, primary ESS can rarely present with cerebrospinal fluid (CSF) rhinorrhea.

We report the case of a 67-year-old man with a 3-month history of headache and left-sided clear nasal discharge. Neurological and ophthalmological examinations were normal. Brain magnetic resonance imaging (MRI) revealed an empty sella. Endocrinological tests were within normal limits. An endoscopic transnasal transsphenoidal repair was performed using autologous fat grafting, resulting in complete resolution of symptoms. CSF leaks in ESS are often attributed to elevated intracranial pressure and diaphragmatic incompetence, leading to sellar floor erosion. Endoscopic repair offers a minimally invasive approach with high success rates and low morbidity.

In cases of ESS complicated by CSF rhinorrhea, surgical repair is mandatory. The endoscopic transnasal transsphenoidal approach remains the preferred treatment option.

Keywords: Empty sella syndrome, cerebrospinal fluid rhinorrhea, endoscopic repair, transnasal transsphenoidal surgery, case-report

INTRODUCTION

Empty sella syndrome (ESS) is defined by the herniation of the subarachnoid space into the sella turcica with flattening of the pituitary gland, as first described by Busch in 1951. It is classified as primary or secondary. While the secondary form is known to be due to a tumor, surgery, radiation therapy or any harmful event leading the pituitary gland to shrink and disappear on imaging causing frequent pituitary hormonal dysfunction, the primary form typically arises in the context of congenital diaphragmatic weakness and elevated intracranial pressure and is often asymptomatic. It may rarely present with CSF rhinorrhea, representing a significant risk of meningitis.
We report the case of a 67-year-old male patient with primary ESS complicated by CSF rhinorrhea, successfully treated by endoscopic transnasal transsphenoidal repair.

Case Report

A 67-year-old man, without relevant medical history, presented with a 3-month history of persistent headache and intermittent left-sided watery inodorous nasal discharge with a metallic taste. Neurological and ophthalmologic examinations were unremarkable. Brain MRI revealed an empty sella with an intrasellar arachnoidocele. Hormonal assessment was within normal ranges.

Surgery was performed via an endoscopic transnasal transsphenoidal approach. The sellar defect was exposed, and a CSF leak into the sphenoid sinus was identified. The defect was repaired with autologous abdominal fat grafting and reinforced with fibrin glue. Postoperative recovery was uneventful, with complete resolution of symptoms at follow-up.

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DISCUSSION

Spontaneous CSF leak is common. In case of empty sella, the elevated intracranial fluid pressure results in bone thinning in sphenoid sinus and cribriform plate [3]. It may eventually cause brain herniation or bone erosion with CSF leakage. Number of hypotheses may explain the cause of primary ESS [4]. A reasonable explanation is that the condition arises in a patient who has an elevation in intracranial pressure associated with an incompetent diaphragma sella which allows the subarachnoid space to be forced into the sella by the hydrostatic pressure and pulsatile movement of CSF. The bony erosion, especially if augmented by increased intracranial pressure, can cause communication of the intrasellar subarachnoid space with the sphenoid sinus. CSF rhinorrhea may be attributed to benign intracranial hypertension, which can be associated with ESS [5]. The site of the leak is usually into the sphenoid sinus but may be through the cribriform plate and can be distinguished after the injection of the intrathecal contrast. Visual changes are noted. They result from herniation of the suprasellar cisterns into the sellar space [4]. This causes downward displacement of the optic nerves, optic chiasm and exposes the optic structures to a more intense CSF pulsation [4].

The MRI represents the gold standard for the diagnosis of the empty sella. It shows a large sella filled with CSF. Because of the high risk of CSF rhinorrhea and infection, the intradural technique was replaced by the extradural technique which represents the current treatment modality [4,6]. Several materials have been suggested for filling the sellar space and reconstruction of the sellar floor. As recorded by many authors, the fat was prefered over muscle because it results in less necrosis or scar retraction over time [4]. The technique consists of inserting an amount of fat inside the sella which will push the optic structure into their normal position. Extradural transsphenoidal chiasmapexy can be indicated if the optic chiasm herniated inside the sella causing progressive visual abnormalities [7]. They include muscle, fat, dural substitutes, cartilage, bone fragments, ceramic substances and titanium plates.

The endoscopic approach is considered the preferred procedure for treatment of sinus CSF leaks [6,8]. The success rate is estimated between 90 and 95%. It is associated with less complication than the open skull approaches. The surgery consists of separation of the communication from the nose and sinuses from the brain compartment. The endoscope helps to identify the site of CSF leak and to place the grafts precisely [9].

CONCLUSION

Primary ESS is usually asymptomatic but can occasionally present with CSF rhinorrhea, requiring surgical repair. The endoscopic transnasal transsphenoidal approach offers a safe, effective, and minimally invasive solution with excellent outcomes.

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Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict of Interest:

A conflict of interest exists when an author's financial interests or other opportunities for tangible personal benefit may compromise, or reasonably appear to compromise, the independence of judgment in the research or scholarship presented in the manuscript submission. In our case, there is no conflict of interest that can potentially affect the material contained in the manuscript submitted to this Journal.

Ethics committee approval is deemed not necessary in our institution for case reports.

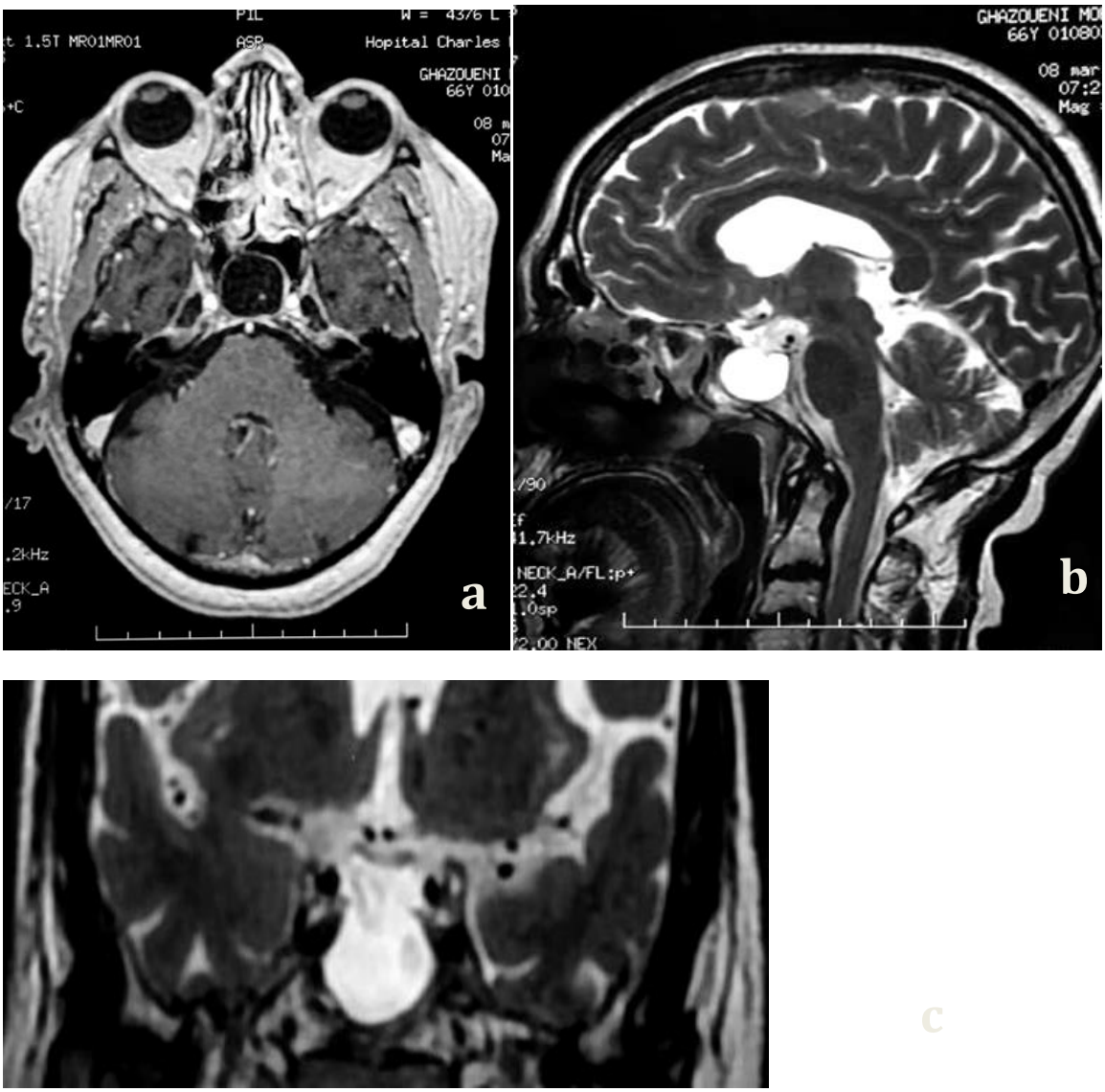

Fig 1: Brain MRI sequences in axial (a), sagittal (b) and coronal (c) sections show the intrasellar arachnoidocele