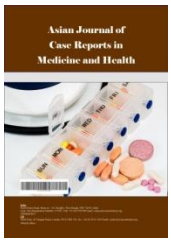

Asian Journal of Case Reports in Medicine and Health

7(2): 1-5, 2022; Article no.AJCRMH.86465

Repair of Cerebrospinal Fluid Rhinorrhea in a Patient Presenting Empty Sella Syndrome: Case Report and Review of Literature

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/86465

Case Study

Received 12 February 2022
Accepted 20 April 2022
Published 30 April 2022

ABSTRACT

Introduction: The cause of Empty Sella syndrome is unknown, however it occurs when intracranial contents herniate through the sellar diaphragm, filling the sella turcica with cerebrospinal fluid (CSF) and giving the appearance of an absent pituitary gland on radiography.
Case Presentation: Author presents a rare case of a 67 years-old woman who was admitted into our department with a complaint of dripping clear fluid from the left side of her nose. Clinical examination and biological, radiological investigations found a CSF leakage through a minute bony defect in the floor of the sella associated with empty Stella syndrome. Treatment consisted of the reparation of the leak using a fascia lata graft, and the sphenoidal sinus was obliterated with fat.
Conclusion: When evaluating patients with non-traumatic CSF rhinorrhea, the otolaryngologist must consider empty sella syndrome as a diagnostic possibility.

Keywords: Empty sella syndrome; cerebrospinal fluid leakage; endonasal endoscopic approach.

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1. INTRODUCTION

"Primary empty sella is characterized by the spontaneous herniation of the subarachnoid space into the pituitary fossa. it is generally asymptomatic and rarely requires any management, however, if symptomatic, the most commonly reported signs and symptoms include headache, cerebrospinal fluid (CSF) leakage, visual deficits, and endocrine dysfunction" [1].

"But the CSF leakage and visual deficits are the only manifestations for which surgical management should be indicated" [1].

The endonasal endoscopic approach is a safe and effective method, offering a better reparation of the sinonasal cerebrospinal fluid leak and lowering morbidity [2]. This technique provides a success rate greater than 90% [3].

We report the rare case of a patient who presents cerebrospinal fluid rhinorrhea revealing empty sella turcica syndrome.

2. CASE REPORT

A 67-year-old obese woman had been oozing clear fluid from the left side of her nose for five months. The fluid was described as having a sweet taste. No nasal congestion was seen, and the leakage became worse after bending forward. The medical history found arterial hypertension, multiparity, a history of a headache without head trauma, or meningitis. No endocrine disorder, and no visual disturbances such as diplopia, blind spots, or blurring.

The examination indicated a clear fluid that could be seen flowing from the left side of the nose. The site of origin was high in the nasal cavity. There were no abnormal masses or polyps in the nose or nasopharynx.

The beta-2 transferrin level of the fluid that was dripping from the nose was 18,6 mg/l.

Computerized axial tomography (CT) indicated a decreased density in the sella, the density being similar to the density of CSF rather than that of pituitary tissue. This suggested the possibility of an empty sella Fig 1.

Magnetic resonance imaging (MRI) showed the typical herniation of the subarachnoid space into the sella Fig 2.

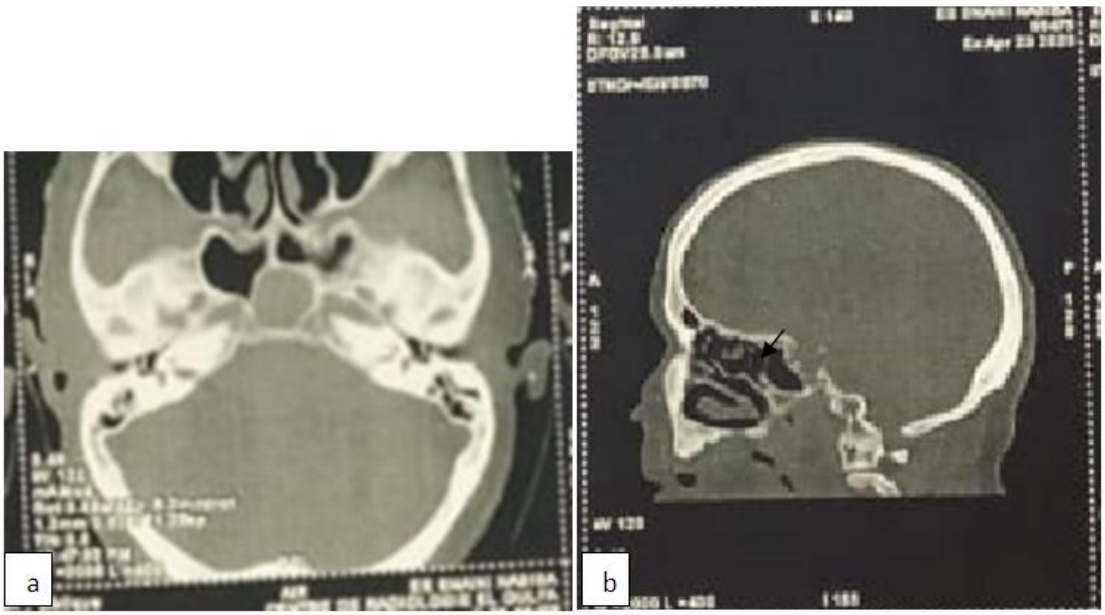

Fig. 1. Cerebral CT axial (a) and sagittal view (b): decreased density in the sella, the density is similar to the density of CSF rather than that of pituitary tissue. This suggested the possibility of an empty sella and the presence of a nibbled appearance at the level of the floor of the seller's turcique (arrow)

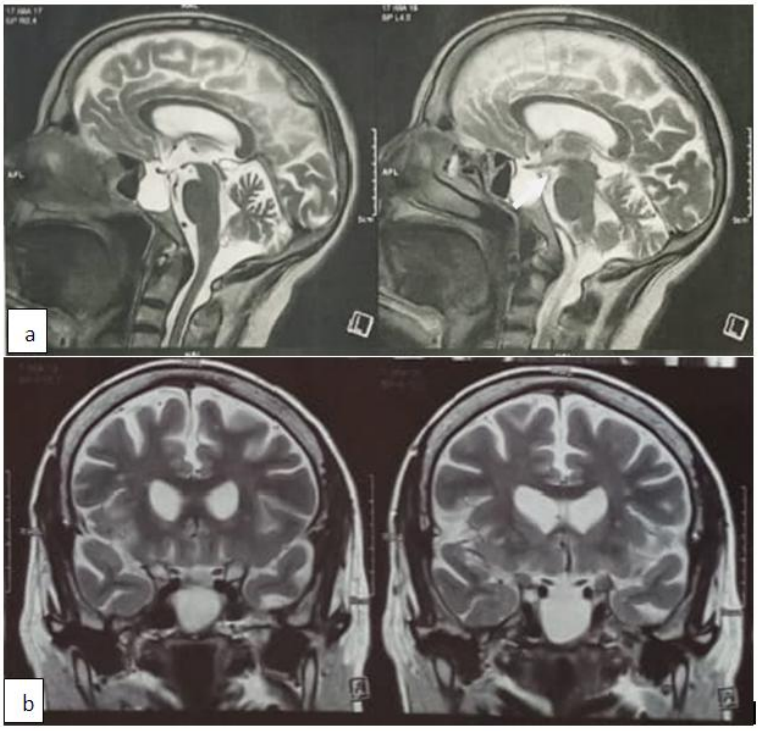

Fig. 2. Cerebral MRI sagittal (a) and coronal view (b): the presence of intra-sellar fluid formation in continuity with the chiasmatic cistern and identical signal to CSF in all sequences corresponding to an empty sella turcica without mass effect on the pituitary stalk

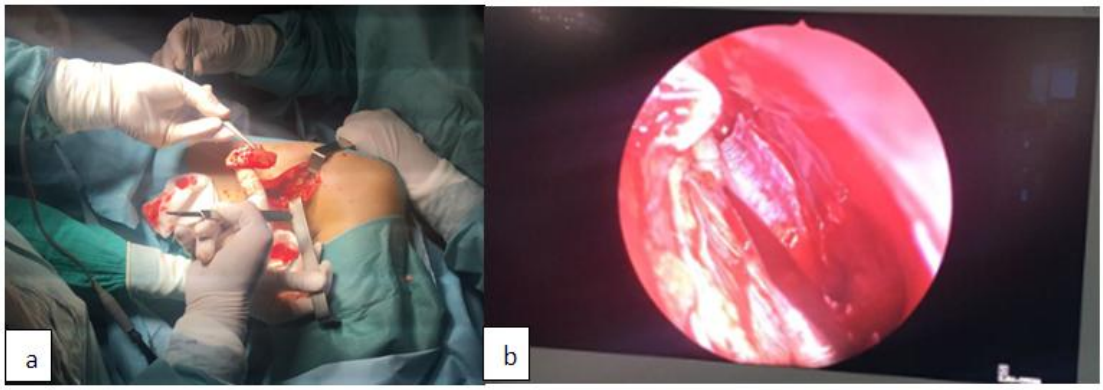

Fig. 3. Intraoperative view showing the extraction of a fascia lata graft from the right thigh (a) and its placement in the sphenoid sinus (b)

The patient underwent a full functional endoscopic sinus surgery exploration and with the use of fluorescein-soaked cottonoids, we find a minute bony defect in the floor of the sella with CSF leaking through this defect. The leak was repaired using fascia lata graft Fig 3, and the sphenoidal sinus was obliterated with fat.

The CSF leaking stopped after the surgery, but the patient developed a persistent headache. The patient was treated with oral acetazolamide and a cough suppressant and a laxative, and her symptoms improved without a recurrence of CSF leaking. No recurrence was present at 6 months of follow-up.

3. DISCUSSION

“Empty Sella is defined as a hernia of subarachnoid space in the sella turcica also called arachnoidocele. It is a term used in radiology to describe the discovery of "empty

sella space" on magnetic resonance imaging and computed tomography with a flattened pituitary gland and an elongated stalk. It can be complete if the cerebrospinal fluid fills more than 50% of space in the sella and the thickness of the gland is less than 2 mm or partial if less than 50% of the sellar space is filled with CSF" [4].

"Regarding its etiology, it can be primary if there is no pathological mechanism in the sellar region preceding the pituitary lesion or secondary if it is consecutive to a particular pathological mechanism" [4].

Primary Empty Sella is the less common of the two entities. Currently, its cause is not fully understood, but researchers have proposed several mechanisms including chronic intracranial hypertension, incompetence or complete absence of the diaphragm sellae, and temporary expansion followed by regression of the pituitary gland [5].

The primary syndrome is commonly seen in middle-aged, obese, and hypertensive women. The disorder may be a sign of idiopathic intracranial hypertension. Among the risk factors, there is obesity in women and multiparity [6]. This may be the case with our patient.

"The clinical presentation of primary empty sella is silent and asymptomatic, not requiring any management. In very rare cases, patients develop cerebrospinal fluid leaks or visual deficits that require surgical treatment" [1].

"The etiology of CSF leaks is diverse: post-surgical iatrogenic (endoscopic sinus surgery is a common cause of iatrogenic CSF leak), trauma is also a common cause, followed by congenital meningoencephalocele, meningioma/pituitary tumor" [7].

While the etiology of traumatic CSF leaks is well known, spontaneous leaks are not and they are also important causes of CSF fistulas, associated with high intracranial pressure, the causes of which are partially empty sella turcica, empty sella turcica, base cranial erosions, hydrocephalus, and intracranial arteriovenous malformation [2].

Our patient had signs of an empty sella on MRI, responsible for raised ICP. Full functional endoscopic sinus surgery exploration uncovered a tiny bone defect in the floor of the sella with CSF leaking through this defect.

"The association between the radiographic finding of an empty sella and the clinical presentation of spontaneous CSF leak is much more common than would be expected" [8]. The study of Schlosser. and al [8] and the study of Shetty and al [9] have noted "the association between spontaneous CSF leaks and an empty sella. Shetty et al16 reviewed 11 patients with spontaneous sphenoid CSF leaks and noted that 63% had completely empty sellae and an additional 27% had partially empty sellae, similar to Schlosser's study results of 67% and 33%, respectively" [8,9].

The indications for endoscopic closure are the accessibility of the leak endoscopically such as localization at (Sphenoid, Cribriform plate, Frontal sinus excepted for posterior wall, Fovea ethmoidal/Lateral lamina, Septal olfactory dural extension) after confirming it by using immunofixation of beta 2–transferring [2].

The steps of the endoscopic surgical technique require intraoperative localization of the fluorescein leak and preoperatively by magnetic resonance imaging and computed tomography (CT) [2].

"Various grafts including muscle, fascias (lata, temporalis, and rectus abdominis), bone, mucoperiosteum, mucochondrium, dural flaps, fat, and various synthetic dural substitutes may be positioned with different techniques (onlay and/or underlay, dumbbell, bath plug, and fat obliteration) to provide a stable scaffold at the site of the defect, with almost identical postoperative success rates.

Free grafts (fascia lata) are advantageous by providing an easily manageable mass allowing a satisfactory field of intraoperative vision with decreased tissue tension" [10]. In our patient, we have used a fascia lata graft.

"Various materials are used to support the secure position of a graft. Fat is a useful supplement and can act as glue to hold the fascia in place and generally render fibrin glue unnecessary." 7 "In addition, free fat is the preferred material for obliterating the sphenoid sinus and mounting the graft on the sinus wall [10], like our patient. Fibrin glue can be used as an adjuvant to improve graft adhesion and secure the graft in the correct position" [10].

Among causes of recurrent sinonasal cerebrospinal fluid (CSF) leaks after repair,

raised intracranial pressure is the most common reason [2].

4. CONCLUSION

These cases of non-traumatic CSF rhinorrhea present a diagnostic challenge.

This case demonstrates that while examining patients with non-traumatic CSF rhinorrhea, the otolaryngologist must consider empty sella syndrome as a diagnostic possibility.

In the majority of patients with sinonasal cerebrospinal fluid (CSF) leaks, endoscopic closure is currently the therapy of choice.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/86465