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SNI: Unique Case Observations

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Image Report

Adult transsphenoidal meningoencephalocele: Clinical image

Bhanu Pratap Singh Chauhan, Binita Dholakia, Daljit Singh

Department of Neurosurgery, Govind Ballabh Pant Institute of Postgraduate Medical Education and Research, New Delhi, India.

E-mail: *Bhanu Pratap Singh Chauhan - bhanupratap1819@gmail.com; Binita Dholakia - binitadholakia@gmail.com; Daljit Singh - drdaljit@hotmail.com

***Corresponding author:**
Bhanu Pratap Singh Chauhan,
Department of Neurosurgery,
GB Pant Institute of
Postgraduate Medical
Education and Research,
New Delhi, India.

bhanupratap1819@gmail.com

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ABSTRACT

Background: Encephalocele is herniation of cranial contents through a skull defect, classified according to their contents and location, and is usually seen in pediatric age group. The transsphenoidal type represents <5% of all basal meningoencephaloceles. Of them, the presentation in adulthood is even rarer.

Case Description: A 19-year-old female complaining of breathing difficulties during sleep and exertional dyspnea was diagnosed with a transsphenoidal meningoencephalocele, likely representing patent craniopharyngeal canal. On exploration through bifrontal craniotomy, the defect was identified in the sellar floor after completely reducing the contents into the cranial cavity and was repaired. She had immediate symptomatic relief and an uneventful postoperative course.

Conclusion: There can be significant symptomatic relief with minimal postoperative morbidity after transcranial repair of such large transsphenoidal meningoencephaloceles, through traditional skull base approaches.

Keywords: Adult presentation, Open repair, Transsphenoidal meningoencephalocele, Surgical repair

INTRODUCTION

Encephalocele is herniation of cranial contents through a skull defect, classified according to their contents and location, and is usually seen in pediatric age group. The transsphenoidal type represents <5% of all basal meningoencephaloceles, with an incidence of one in 700,000 live births.[1] Of them, the presentation in adulthood is even rarer. This condition requires special attention, as it can be cured completely, leading to prompt resolution of symptoms, by traditional open and minimally invasive endoscopic approaches.

CASE SUMMARY

A 19-year-old female student presented with complaints of difficulty in breathing during sleep since childhood, right vision loss (congenital cataract), and exertional dyspnea for 2 years. She was born prematurely at 7 months by cesarean section with history of neonatal intensive care unit stay. There were associated menstrual irregularities since 1 year. On general examination, she had frontal bossing, a short neck, telecanthus, and slanting palpebral fissures with normal

higher mental functions. Visual examination showed that the right pupil was dilated, not reactive, with no perception of light, with the left eye best-corrected vision measured as 6/24. Chromosomal analysis, 2D echo study, and hormonal profile were normal.

She consulted an otolaryngologist, and on finding no significant clinical cause, a contrast-computed tomography (CT) scan was performed, which was suggestive of a well-defined corticated midline defect replacing the body of sphenoid measuring 2.4 × 1.6 cm. communicating with the nasopharynx and 3rd ventricle [Figures 1a and b], likely representing a patent craniopharyngeal canal with meningocele (type 3A). These findings were corroborated on magnetic resonance imaging, with pituitary stalk and gland not being identified [Figure 2]. Incomplete cleft palate was also seen. Surgical exploration was done through a bifrontal craniotomy, with reduction of herniating meninges and pituitary stalk into cranium, excision of sac and repair of defect with pericranium, Surgicel, and glue through a subfrontal approach [Figures 3a-c]. The postoperative CT was suggestive of complete reduction of contents into the cranial cavity with air fluid level in the sphenoid sinus [Figure 4]. The patient had immediate symptomatic relief in her breathing with an uneventful postoperative course.

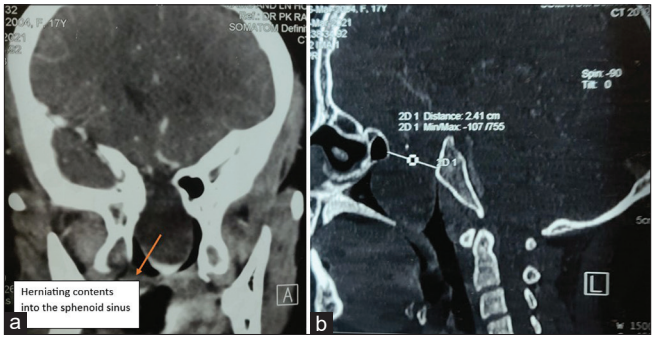

Figure 1: (a) Coronal section contrast computed tomography (CT) scan showing a nonenhancing isointense soft-tissue mass herniating into the sphenoid sinus reaching up to the floor. (b): Sagittal section bone window CT scan showing defect size of 2.4 cm in the sellar floor with herniating soft-tissue contents.

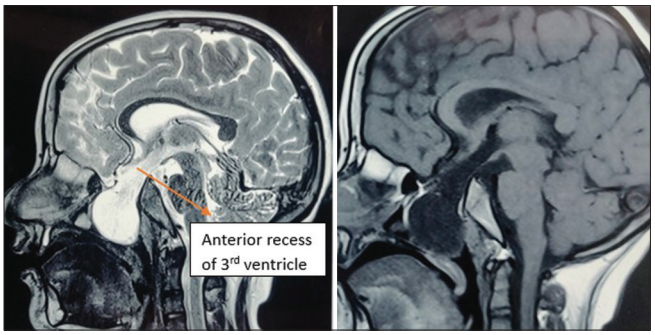

Figure 2: Sagittal section T2 and T1 sequences of magnetic resonance imaging showing herniating meninges through the sellar floor into the sphenoid sinus, including anterior recess of third ventricle, with no separately identifiable pituitary gland and stalk.

DISCUSSION

Occurrence of cephaloceles is approximately one in every 3000–5000 live births without any gender predilection, with 30% incidence of associated anomalies.[1] Encephaloceles in the frontal and basal portions of the skull are classified anatomically according to the scheme of Meyer, Safranek, and Gisselson.[2] The basal type of encephalocele occurs along the cribriform plate or through the sphenoid bone. The mass may appear in the nasal cavity, nasopharynx, epipharynx, sphenoid sinus, posterior orbit, or pterygopalatine fossa. The distinction from other types is that no external tumor is visible except in those rare instances of herniations so large that they protrude through the mouth or nares.[2]

Based on the integrity of the sphenoid sinus floor, Jabre *et al.* noted two types of transsphenoidal meningoencephaloceles: the intrasphenoidal (extending into the sphenoid sinus but confined by its) and the true transsphenoidal (traversing the floor of the sphenoid sinus and protruding into the nasal cavity or nasopharynx).[1]

Treatment options vary from minimally invasive endoscopic transsphenoidal repair to open surgical exploration and repair. Endoscopic approaches are preferred for herniations into lateral sphenoid sinus, which include transnasal, transpterygoid, and transethmoid approaches.[4] Transcranial

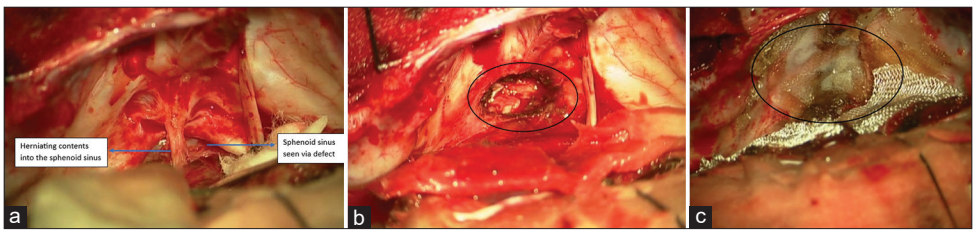

Figure 3: (a) Intraoperative microscopic view showing approximately 3 × 2 cm defect in the sellar floor with herniating meninges and pituitary stalk into the sphenoid sinus. (b) Intraoperative microscopic view showing defect (in oval) after complete reduction of contents into the cranial cavity. (c) The defect was repaired with pericranium, Surgicel, and glue (in oval).

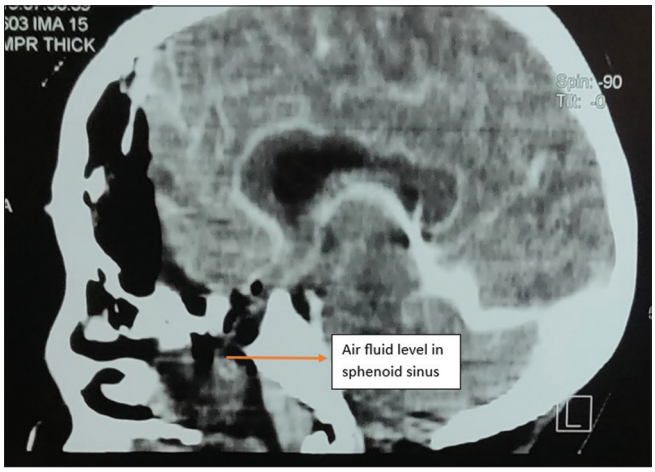

Figure 4: Sagittal section postoperative computed tomography scan demonstrating replaced contents into the cranial cavity with air fluid level in the sphenoid sinus.

approaches allow for an optimal and wide exposure of relevant anatomy, minimizing risks, and improving the possibility to achieve a strong multilayer reconstruction.[3] Both options provide good long-term outcomes, with minimal postoperative cerebrospinal fluid leak and meningitis.

CONCLUSION

There can be significant symptomatic relief with minimal post operative morbidity after transcranial repair of such large transsphenoidal meningoencephaloceles, via traditional skull base approaches.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Jabre A, Tabaddor R, Samaraweera R. Transsphenoidal meningoencephalocele in adults. Surg Neurol 2000;54:183-8.
2. Pollock JA, Newton TH, Hoyt WF. Transsphenoidal and transethmoidal encephaloceles. Radiology 1968;90:442-53.
3. Spina A, Boari N, Calvanese F, De Domenico P, Cannizzaro M, Trimarchi M, *et al*. Transcranial extradural subtemporal repair for sphenoid sinus lateral recess meningoencephalocele: Technical note. Neurosurg Rev 2022;45:903-9.
4. Tabaee A, Anand VK, Cappabianca P, Stamm A, Esposito F, Schwartz TH. Endoscopic management of spontaneous meningoencephalocele of the lateral sphenoid sinus. J Neurosurg 2010;112:1070-7.

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