

Sexual dysfunction as the initial clinical feature of a massive intrasphenoidal meningoencephalocele: A case report

Chi-Man Yip, Jui-Hsun Fu, Shu-Shong Hsu

ABSTRACT

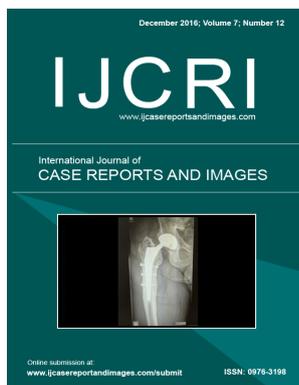
Introduction: Trans-sphenoidal encephaloceles comprise less than 5% of all basal meningoencephaloceles, which represent a rare clinical entity with wide range of symptoms in adult. Headache, endocrine alternations, visual field defects or other visual alternations, CSF leakage are the possible clinical features. CSF leakage and visual deficit are the two clinical indications for surgery.

Case Report: A 38-year-old male presented to us with the chief complaint of sexual dysfunction detected after marriage. His sellar magnetic resonance imaging (MRI) showed a large cystic lesion without obvious enhancement located in the sellar region as well as sphenoid sinus on regular T1-weighted, T2-weighted and T1-weighted with Gadolinium images. The cine phase-contrast and 3D FIESTA series disclosed the descent of anterior floor of 3rd ventricle including infundibular recess and optic recess with expansion of the descending portion of 3rd ventricle and herniation of optic chiasm into sella turcica and sphenoid sinus. A massive intrasphenoidal meningoencephalocele with panhypopituitarism, hypogonadism and hyperprolactinemia as well as optic neuropathy was diagnosed and he underwent endoscopic endonasal transsphenoidal chiasmectomy in order to release the stretching on the herniated suprasellar visual system and hormone therapy after surgery to manage his endocrinological disorders.

Conclusion: Magnetic resonance imaging (MRI) scan of sella is essential in evaluation of trans-sphenoidal encephalocele to confirm the extent of lesion, the possible associated abnormalities and to plan for the safest approach. In addition, cine phase-contrast MRI scan can differentiate the meningo-encephalocele from other cystic lesions in the sellar region and three-dimensional fast imaging employing steady-state acquisition (3D FIESTA) sequence can delineate the suprasellar visual system well before operation.



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Introduction: Trans-sphenoidal encephaloceles comprise less than 5% of all basal meningoencephaloceles, which represent a rare clinical entity with wide range of symptoms in adult. Headache, endocrine alternations, visual field defects or other visual alternations, CSF leakage are the possible clinical features. CSF leakage and visual deficit are the two clinical indications for surgery. **Case Report:** A 38-year-old male presented to us with the chief complaint of sexual dysfunction detected after marriage. His sellar magnetic resonance imaging (MRI) showed a large cystic lesion without obvious enhancement located in the sellar region as well as sphenoid sinus on regular T1-weighted, T2-weighted and T1-weighted with Gadolinium images. The cine phase-contrast and 3D FIESTA series disclosed the descent of anterior floor of 3rd ventricle including infundibular recess and optic recess with expansion of the descending portion of 3rd ventricle and herniation of optic chiasm into sella turcica and sphenoid sinus. A massive intrasphenoidal meningoencephalocele with panhypopituitarism, hypogonadism and hyperprolactinemia as well as optic neuropathy

was diagnosed and he underwent endoscopic endonasal transsphenoidal chiasmectomy in order to release the stretching on the herniated suprasellar visual system and hormone therapy after surgery to manage his endocrinological disorders. **Conclusion:** Magnetic resonance imaging (MRI) scan of sella is essential in evaluation of trans-sphenoidal encephalocele to confirm the extent of lesion, the possible associated abnormalities and to plan for the safest approach. In addition, cine phase-contrast MRI scan can differentiate the meningoencephalocele from other cystic lesions in the sellar region and three-dimensional fast imaging employing steady-state acquisition (3D FIESTA) sequence can delineate the suprasellar visual system well before operation.

Keywords: Cine phase-contrast MRI scan, Endoscopic endonasal transsphenoidal chiasmectomy, Suprasellar visual system, Three-dimensional fast imaging employing steady-state acquisition, Trans-sphenoidal meningoencephaloceles

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INTRODUCTION

Encephalocele can be congenital, spontaneous, or traumatic in cause [1]. According to the site of origin, the encephaloceles are classified to occipital, sincipital, nasofrontal, nasoethmoidal, nasoorbital and basal, sphenoorbital, sphenomaxillary, trans-ethmoidal, sphenothmoidal, trans-sphenoidal, and basioccipital cephaloceles [1–5]. Transsphenoidal meningoencephalocele is a congenital anomaly that consists of herniation through a sphenoidal bony defect of an endymal-lined sac filled with cerebrospinal fluid (CSF) and containing neural vascular tissue. It may encompass the third ventricle, hypothalamic-pituitary elements, anterior cerebral arteries and optic apparatus [5]. Transsphenoidal encephaloceles represent a rare clinical entity with wide range of symptoms in adult. Headache, endocrine alternations, visual field defects or other visual alternations, CSF leakage are the possible clinical features [1, 3–5]. We would like to share a case of massive intrasphenoidal meningoencephalocele initially presented by sexual dysfunction who underwent endoscopic endonasal transsphenoidal chiasmectomy in order to release the stretching on the herniated suprasellar visual system and hormone therapy after surgery to manage his endocrinological disorders.

CASE REPORT

In August 2015, a 38-year-old male without any past medical history and trauma history presented to us with the chief complaint of sexual dysfunction detected after his marriage. On admission, he was conscious clear and his neurological examination was essentially normal except his visual field study showed bitemporal hemianopia, but he did not notice any visual problem before this admission. His endocrine survey were as followings: cortisol 1.14 ug/dL (normal value 5–25 ug/dL), plasma ACTH 14.4 pg/mL (normal value <46 pg/mL), HS-TSH 3.76 uIU/ml (normal value 0.4–4.0 uIU/ml), T₃ 121 ng/dl (normal value 84–172 ng/dl), T₄ 3.39 ug/dl (normal value 4.5–12.5 ug/dl), free T₄ 0.496 ng/dl (normal value 0.8–1.9 ng/dl), human growth hormone 0.03 uIU/mL (normal value 0.03–1.17 uIU/mL), testosterone <0.023 ng/mL (normal value 1.42–9.23 ng/mL), FSH 0.3 mIU/mL (normal value 0.85–8.50 mIU/mL), prolactin 303.6 ng/ml (normal value 1.61–18.77 ng/ml), therefore, panhypopituitarism, hypogonadism and hyperprolactinemia were concluded.

Sellar magnetic resonance imaging (MRI) showed a large cystic lesion, about 4.14x2.04x3.21 cm in size without obvious enhancement located in the sellar region as well as sphenoid sinus on regular T₁-weighted, T₂-weighted and T₁-weighted with Gadolinium images (Figure 1). However, cystic lesions in the sella are challenging in diagnosis, especially, in this particular case, to confirm the relationship between the cystic lesion

and the third ventricle is indeed essential to plan for the safest approach and the management. After discussing with neuroradiologist, we did cine phase-contrast MRI (Figure 2) and three-dimensional fast imaging employing steady-state acquisition (3D FIESTA) sequence (Figure 3) to identify the relationship between the third ventricle and the cystic lesion and the suprasellar visual system, respectively. The cine phase-contrast and 3D FIESTA series disclosed the descent of anterior floor of 3rd ventricle including infundibular recess and optic recess with expansion of the descending portion of 3rd ventricle and herniation of optic chiasm into sella turcica and sphenoid sinus, causing compression of pituitary gland into posterior sellar floor and enlargement of sella turcica which occupied almost the whole sphenoid sinus.

Under general anesthesia, the patient was put in supine position and underwent endoscopic endonasal transsphenoidal chiasmectomy in order to release the stretching on the herniated suprasellar visual system. Under the endoscopic view and neuronavigation guidance, the rostrum of sphenoid sinus and sellar floor were identified and removed with extreme caution. Extradural dissection was performed and elevated the *intrasellar content* gently so that an extradural space was created until a tension was detected. Inserted fat grafts harvested from his abdominal wall and bone grafts harvested from his resected middle turbinate and vomer bone to occupy the extradural space in order to elevate and support his herniated suprasellar visual system. Before surgery, endocrinologist was consulted to manage his endocrinological disorders perioperatively and postoperatively. The hormone replacement regimen of this patient was as follow: cabergoline 0.5 mg per week, levothyroxine 0.1 mg per day, prednisolone 5 mg per day and nebido (1000 mg) intramuscular injection once in October 2015. After discharge, he is regularly followed at neurosurgery and endocrinology outpatient department. His latest hormone survey in April 2016 was as follows: cortisol 1.75 ug/dL, prolactin 1.32 ng/ml, free T₄ 1.22 ng/dl, testosterone 4.03 ng/mL. His vision got some improvement and his sexual performance improved a lot after surgery and hormone therapy.

DISCUSSION

Meningoencephalocele is a herniated sac of brain structures and meninges into a defect of skull and dura [4]. The incidence of congenital encephalocele is approximately 1 in 3000–5000 live births [1, 2, 5]. Trans-sphenoidal encephaloceles comprise less than 5% of all basal meningoencephaloceles [1–3, 5]. Abiko et al. believed that there are two types of trans-sphenoidal meningoencephalocele based on the integrity of the floor of the sphenoid sinus, that is, the intra-sphenoidal and the true trans-sphenoidal types. The former describes those extending into the sphenoid sinus but confined by its floor. The latter describes those traversing the

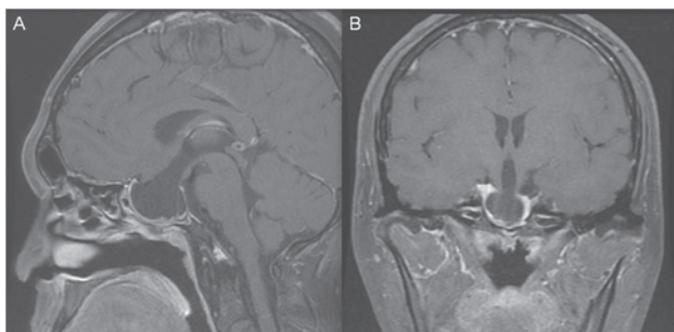


Figure 1: Preoperative sellar MRI, T1-weighted with Gadolinium. (A) Sagittal view (B) coronal view demonstrating a large cystic lesion without obvious enhancement located in the sellar region as well as sphenoid sinus.

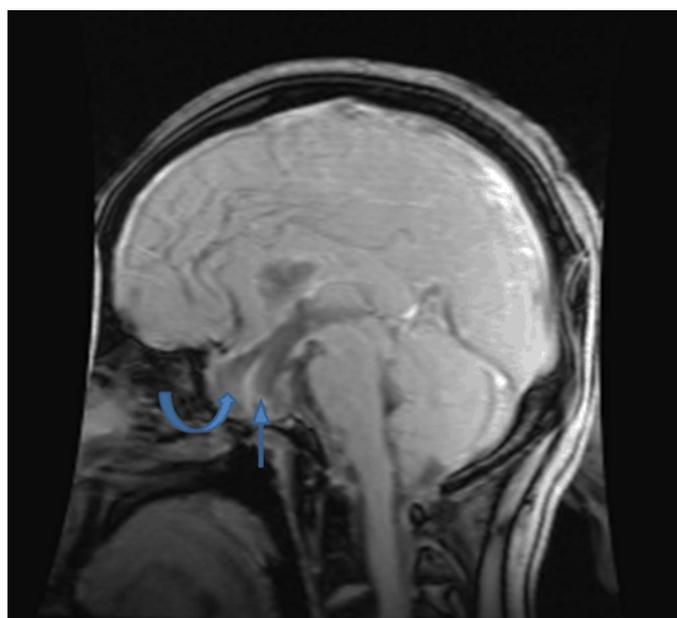


Figure 2: Preoperative sellar MRI, cine phase-contrast sequence. Sagittal view showing the descent of anterior floor of 3rd ventricle including infundibular recess (straight blue arrow) and optic recess (curve blue arrow) with expansion of the descending portion of 3rd ventricle into sellar turcica and sphenoid sinus.

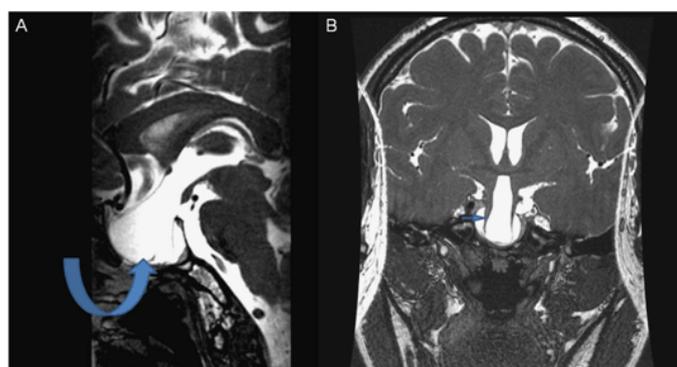


Figure 3: Preoperative sellar MRI, 3D FIESTA sequence. (A) sagittal view (B) coronal view disclosing the descent of anterior floor of 3rd ventricle including infundibular recess and optic recess with expansion of the descending portion of 3rd ventricle (curve blue arrow) and herniation of optic chiasm (straight blue arrow) into sellar turcica and sphenoid sinus.

floor of the sphenoid sinus and protruding into the nasal cavity or nasopharynx [1, 5]. Based on Abiko et al. description, the patient belonged to intra-sphenoidal type meningoencephalocele. There are multiple theories to explain the formation of the basal encephalocele, including the failure of complete closure of the neural tube leading to herniation of meninges and neural tissues, error in the development of the sphenoid bone with resultant congenital defect of the skull base, persistence of the craniopharyngeal canal allowing the descent of intracranial structures, and the failure of the neuroectoderm to separate from the surface of the ectoderm during the formation of neural tube, thus preventing the mesodermal tissue from interposing between these two germ layers causing changes in the skull ossification and allowing herniation secondarily [1, 5].

Clinical features of the patient having trans-sphenoidal meningoencephalocele are variable depending on the age [1, 3]. In infancy, trans-sphenoidal meningoencephalocele can be presented by respiratory distress, feeding problems, endocrine abnormalities, episodes of recurrent meningitis, concomitant craniofacial defects and associated congenital anomalies [2, 5]. If the diagnosis is delayed to adulthood, the common features include visual problem, endocrine abnormalities, unexplained rhinorrhea, meningitis [1, 3, 4]. The main indications of surgical intervention for trans-sphenoidal meningoencephaloceles are obstruction of respiratory pathway, CSF rhinorrhea, meningitis related to CSF rhinorrhea, progressive visual defect due to the lesion [1].

Magnetic resonance imaging (MRI) scan of sella is essential in evaluation of trans-sphenoidal encephalocele to confirm the extent of lesion, the possible associated abnormalities and to plan for the safest approach [1, 3]. The content of trans-sphenoidal meningoencephaloceles are CSF and some neural vascular structures; they may resemble other cystic lesions in the sellar region and/or sphenoid region. Cine phase-contrast MRI is a good tool to assess cerebrospinal fluid flow [6]. It can help us to identify the cystic content to be CSF; this information is essential to establish the surgical plan to a “cystic lesion” in the sellar region and /or sphenoid region. If a transsphenoidal meningoencephaloceles is inadvertently opened, CSF leakage will be resulted subsequently and sometimes it is difficult to manage without good preparation. Three-dimensional fast imaging employing steady-state acquisition (3D FIESTA), a newly developed MRI in gradient echo (GRE) sequences, has successfully delineated the anatomy and pathology of cranial nerves and spinal cord [7]. It can delineate the suprasellar visual system well and provide detailed anatomical information to the neurosurgeons before surgery.

CONCLUSION

Trans-sphenoidal encephalocele is a rare clinical entity with wide range of symptoms in adult.

Cerebrospinal fluid (CSF) leakage and visual deficit are the two clinical indications for surgery. Depend on the intactness of the floor of the sphenoid sinus, trans-sphenoidal encephalocele can be classified into two types: the intra-sphenoidal and the true trans-sphenoidal types. Magnetic resonance imaging (MRI) scan of sella is essential in evaluation of trans-sphenoidal encephalocele to confirm the extent of lesion, the possible associated abnormalities and to plan for the safest approach. In addition, cine phase-contrast MRI can differentiate the meningoencephalocele from other cystic lesions in the sellar region and three-dimensional fast imaging employing steady-state acquisition (3D FIESTA) sequence can delineate the suprasellar visual system well before operation. Most patients suffering trans-sphenoidal encephalocele have associated endocrinological disorders which can be managed by endocrinologist to achieve good clinical outcome.

Author Contributions

Chi-Man Yip – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Jui-Hsun Fu – Substantial contributions to conception and design, Analysis and interpretation of data, Revising the article critically for important intellectual content, Final approval of the version to be published

Shu-Shong Hsu – Substantial contributions to conception and design, Revising the article critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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