Case Report

Huge Anterior Cranial Fossa Neurenteric Cyst with Unusual Ocular Presenting Symptoms without Other Neurologic Symptoms: A Case Report and Literature Review

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Objective: *To report a patient with large neurenteric cyst located in the anterior cranial fossa presented with uncommon presenting symptoms.*
Case Report: *A 47-year-old healthy Thai male presented with gradually decreasing vision in the past six months and esotropia for one year. Other neurological examinations and pituitary hormone tests were unremarkable.*
Results: *Orbital magnetic resonance imaging (MRI) depicted a large lobulated cystic mass with the size of 7.1x8.0x7.5 cm, abutted on the sellar, suprasellar, anterior cranial fossa regions, and bilateral small size optic nerves. Aspiration and incisional biopsy of the cyst were performed. Tissue pathology was compatible with neurenteric cyst.*
Conclusion: *Neurenteric cyst is rarely located in the anterior cranial fossa. Although this patient had a very large lesion located in anterior cranial fossa and sellar area, he had only ophthalmic symptoms without headache, nausea, or any neurologic deficits. It is important not to overlook this rare cyst with an unusual presentation.*

Keywords: *Neurenteric cyst, Anterior cranial fossa, Endodermal cyst, Intracranial, Visual loss*

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Neurenteric cysts, also called enterogenous cyst[1], are uncommon benign congenital epithelial cysts of endodermal origin[2]. These cysts tend to be found in the spine and rarely present in an intracranial location. For intracranial lesions, cysts are typically found in the posterior fossa[1,3], uncommon in the anterior cranial fossa. From a literature review, most cases presented with ocular symptoms together with systemic symptoms. To our knowledge, there are few neurenteric cyst patients who presented with solely ocular symptoms. Herein, we reported a neurenteric cyst that was located in the anterior cranial fossa and manifested with unusual presenting symptoms. The study adhered to the principles of the Declaration of Helsinki. The Institutional Review Board of the Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand, had exempted this current study (IRB. 584/58) and informed consent was obtained.

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Case Report

A 47-year-old Thai male presented with gradually decreasing vision in the past six months and esotropia for one year. He denied any significant underlying diseases. He had no ocular pain, nausea, vomiting, or headache. He was afebrile with normal vital signs. He had good consciousness, no history of seizure, no behavioral change, and no other neurological deficit. His visual acuity was counting finger two feet in the right eye and hand motion in the left eye. He had a marked limitation of abduction in the left eye. The Krimsky test showed esotropia 90 prism diopters. Pupils were 3 mm. slightly react to light in both eyes. Relative afferent pupillary defect was negative. Optic discs were mild pallor. Systemic examinations revealed muscle power grade 5/5 in all extremities. Babinski sign was negative. Normal deep tendon reflex was demonstrated. The findings otherwise

were unremarkable.

Results

Orbital MRI demonstrated a large lobulated cystic mass, 7.1x8.0x7.5 cm, located in the anterior cranial fossa, abutted on the sellar, suprasellar region. Mass effect on the inferior frontal lobe, medial temporal lobe, pons, midbrain, optic nerves, optic chiasm, optic tract, hypothalamus, as well as cavernous sinuses and bilateral small size optic nerves were observed. Inferior extension of the lesion into the skull base, roof of the nasopharynx and posterior nasal cavity with deformed cribiform plate, basosphenoid and clivus was also noted. The pituitary gland was indistinctive with distorted pituitary stalk. Ventricles were not dilated. Atrophic optic nerves were noted with widened cerebrospinal fluid (CSF) perineural spaces and optic atrophy (Fig. 1). The results of pituitary hormone tests were within the normal range.

The patient underwent aspiration of the cyst via endoscopic transphenoidal approach and incisional biopsy was done at the aspirated cyst wall. Pathologic examination showed fragments of fibrous cyst wall, lined with non-keratinized stratified squamous epithelium. In occasional areas, ciliated simple columnar epithelium is noted on top of the squamous epithelium, compatible with an endodermal cyst (Fig. 2). Six months after aspiration of the cyst, his visual acuity was improved to 20/200 with pinhole in the right eye and counting finger three feet in the left eye. No other neurological deficits or abnormal blood hormonal tests were noted.

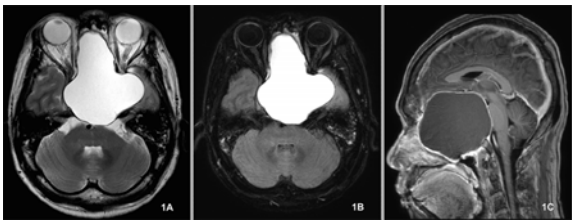

Fig. 1 1A) T2 Axial view, 1B) FLAIR axial view, 1C) T1, Post Gadolinium, Sagittal view revealed large rim enhancing cystic mass with the epicenter located at anterior-central skull base.

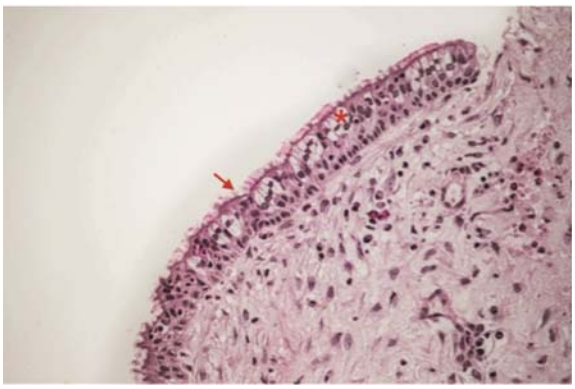

Fig. 2 This part of the cyst wall is lined by ciliated columnar cells (arrow) with occasional goblet cells (*). The underlying connective tissue consists of fibroblasts and small number of inflammatory cells.

Discussion

Neurenteric cysts, which are known by many different terms such as endodermal cyst, enterogenous, enteric, gastrocystoma, and gastroenterogenous[4-8], are rare, congenital, benign lesions lined by mucin-secreting cuboidal or columnar epithelium reminiscent of an intestinal or respiratory tract. These cysts tend to be found in the spine. Regarding intracranial lesions, these are typically found in the posterior cranial fossa[1,2,7,9], rarely in the supratentorial compartment, especially in the suprasellar region. Here, we found a large mass located in anterior cranial fossa, especially in suprasellar area, which is an uncommon location for this cyst.

To our knowledge, after reviewing the English literatures from Scopus, Pubmed database, using the terms “Intracranial neurenteric cyst” and “endodermal cyst” between 1981 and April 2016 as shown in Table 1, there were limited cases of intracranial neurenteric cysts that presented with unusual ocular symptoms. Age of onset varied. Female was predominant (11:5)[10-25]. Seven reports of oculomotor deficit symptoms were found. In all reports, lesions were small and located near the exit zone of the third cranial nerve, which was reasonable for absence of systemic symptoms. Another nine of ten reports (case 8 to 16) presented with visual disturbance. However, they had other neurological symptoms such as headache, hypopituitarism, deafness, gait disturbance, and ophthalmoplegia. Most of the lesions were located along visual pathway, especially around the sellar region. The current case showed no neurologic symptoms but ocular problems. Because of congenital nature, neurenteric cyst begins during early fetal development and can become symptomatic since childhood due to rapid expansion. However, in the current report, he stayed asymptomatic until 47-year-old. Moreover, this was the biggest lesion at presentation, compared with the others, which

Table 1. Literature review of Neurenteric cyst with abnormal ophthalmological presentations

Case	Author (year) (ref)	Age (years)	Gender	Clinical presentation	Time of onset	CT/MRI features
1	Morgan et al. (2001)[13]	30	Female	Chronic right CN III paresis	20 years	MRI: 13x8x6 mm cystic lesion originating from the right oculomotor nerve at its exit from the mesencephalon
2	Okunaga et al. (2006)[14]	16	Female	Headache, acute left CN III paresis	2 days	MRI: A well-defined cystic mass in the proximity of the left oculomotor nerve and bilobulated by the septum
3	Karikari et al. (2010)[16]	2	Female	Acute CN III paresis (pupillary involvement)	No accuracy detail	MRI: 6x5 mm nonenhancing ovoid lesion within the proximal cisternal segment of right oculomotor nerve
4	Wait et al. (2010)[18]	5	Female	Congenital CN III paresis, worsening headache	2 years	MRI: 5x10 mm homogenous the cystic lesion in the interpeduncular cistern impressing right cerebral peduncle
5	Figueiredo EG et al. (2012)[19]	20	Female	Chronic left CN III paresis with acute on top, sudden headache,	Sudden	MRI: Cyst located on the left oculomotor nerve and crural cistern
6	Turner et al. (2012)[20]	3	Female	Recurrent complete right CN III palsy	Sudden	MRI: Cystic mass in the right ambient cistern, compressing anterolateral aspect of the brainstem
7	Tanaka M et al. (2014)[24]	9	Female	Left ptosis, complete CN III paresis	Sudden	MRI: Cystic lesion at the exit of III from midbrain, impressing left cerebral peduncle, no contrast enhance
8	Palma and Celli (1983)[23]	17	Female	Visual loss, headache, hypopituitarism	9 years but rapidly progress during the last month	CT: Hyperdensity mass expanding suprasellar area
9	Scarvilli et al. (1992)[10]	36	Male	Slow progressive visual loss of left eye, left frontal headache	15 years	MRI: Well defined intrinsic mass at posterior intraorbital portion of the optic nerve
10	Leventer et al. (1994)[11]	23	Female	Visual loss, ophthalmoplegia left eye	Sudden	CT: Isointense mass at orbital apex, no contrast enhancement
11	Sampath et al. (1999)[12]	27	Male	Repeated episodes of headache, vomiting, ophthalmoplegia, decrease vision	7 years	MRI: A hyperintense, well-circumscribed parasellar lesion in the left parasellar area
12	Marchionni et al. (2008)[15]	20	Female	Headache, left deafness and blur vision	8 months	MRI: Large cystic lesion involving left temporal, peri- insular area and left cerebellopontine angle, midline shift, compression of left lateral ventricle
13	Reddy et al. (2010)[17]	20	Male	Suboccipital headache and visual loss in right eye	2 months	MRI: Extradural cyst in the left posterior fossa with extension into the temporal region across the tentorium

CN = cranial nerve; CT = computed tomography; MRI = magnetic resonance imaging

Table 1. (cont.)

Case	Author (year) (ref)	Age (years)	Gender	Clinical presentation	Time of onset	CT/MRI features
14	Caruso et al. (2013)[22]	42	Female	Worsening headache, progressive visual loss in right eye	6 months	MRI: Small cystic mass in right parasellar region which appears hypointense on T1-weighted, hyperintense on T2-weighted
15	Akimoto J et al. (2013)[21]	32	Male	Rapidly progressive decrease visual acuity and diplopia	Sudden	MRI: Suprasellar cystic mass extending to upper part of cloves, slightly higher signal intensity in T1-weighted
16	Chakraborty et al. (2015)[25]	71	Male	Progressive worsening vision in left eye	No accuracy detail	MRI: Large mass that caused severe destruction of frontal and ethmoid sinuses. Erosion of inferior part of frontal bone, top of nose and medial orbit
17	Current case	47	Male	Progressive visual loss and esotropia	1 years	MRI: A large lobulated cystic mass, 7.1x8.0x7.5 cm involves sellar, suprasellar, anterior cranial fossa region and bilateral small size optic nerve

CN = cranial nerve; CT = computed tomography; MRI = magnetic resonance imaging

supposed to have some systemic symptoms or brain stem signs due to size and location, but patient had only ocular symptoms, without any other neurological abnormalities. We hypothesized that these might be from brain adaptability responding to slow growing lesion.

Imaging

Cystic lesions are very difficult to differentiate preoperatively. There is no definite imaging finding, but it typically appears as a round and/or lobulated mass. The signal intensity varies depend on the protein content of the cyst[9,26]. Most lesions had an isointense to hyperintense signal in T1-weighted images. In T2-weighted images, most showed hyperintense signal characteristics. There was no report of gadolinium contrast enhancement[17,26]. The presenting case also had the same characteristic as described.

Pathology and location of lesion

Neurenteric cyst was highly suspected in the present case. Neurenteric cysts, Rathke's cleft cysts, and colloid cysts have similar histopathology and immunohistochemistry[8,10,12,27,28] making it difficult to differentiate these three lesions. Some investigators[14,29] have suggested that neurenteric cysts could be differentiated from the other two based on their immunoreactivity to carcinoembryonic antigen (CEA), ferritin, or cytokeratins, even though no specific marker for neurenteric cyst has been specified. Thus, these are usually classified regarding to their location[30]. Rathke's cleft cysts often result from intrasellar remnant, colloid cysts reflect remnants in the third ventricle, and cysts formed in the presellar or retrosellar location are neurenteric[26]. However, in the current case, the huge cyst was located in the anterior cranial fossa area, just abutted on the sellar, suprasellar region but had not originated in the sellar area, nor this cyst was involved in the third ventricle making it less likely to be Rathke's cleft cyst or colloid cyst.

The treatment for endodermal cyst depends on the location of the lesion. Although surgical intervention in neurenteric cysts should attempt to achieve complete resection, making complete resection is potentially dangerous in some situations. If the lesion was in a dangerous location, subtotal resection is often advocated to avoid further neurological deterioration.

However, subtotal resection has a greater chance of recurrence than total resection, thus potentially requiring further surgical intervention and risk of progressive new neurological deficits[7]. In the current case, the patient underwent aspiration and incisional biopsy of the cyst due to the location and size of lesion. Six months postoperatively, his best

corrected visual acuity was improved. No additional neurological deficit was found and the repeated imaging demonstrated no evidence of the cyst reforming at one year follow-up.

Conclusion

We report a large anterior fossa neurenteric cyst case that presented with bilateral optic atrophy, unilateral fifth (ophthalmic branch) and sixth cranial nerve palsy without systemic symptoms. The unusual presentation may lead to misdiagnosis and improper initial investigation. Early correct diagnosis and prompt surgical intervention may preserve vision. Therefore, we should be aware of this rare disease with unusual presentation.

What is already know on this topic?

From the previous study, authors knew intracranial neurenteric cyst is uncommon especially in anterior cranial fossa. Many ocular-symptom cases had other symptoms simultaneously, either systemic or neurologic symptoms. Some were rapid onset and another were gradual or repeated. In the presenting case, patient had only ocular symptoms without others. His symptoms were slowly progressive.

What this study adds?

Intracranial neurenteric cyst can be found at anterior cranial fossa. Onset of symptoms for neurenteric cyst was varied. No gender preferences for intracranial lesion of this cyst type. Huge neurenteric cyst can be presented with solely ocular symptom. Treatment options for large cyst in the brain is challenging. Pathologic evidence is still needed to prove.

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Potential conflict of interest

None.

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ถุงน้ำชนิดนิวเรนเทอริค (Neurenteric Cyst) ขนาดใหญ่บริเวณสมองส่วนหน้าซึ่งแสดงเพียงอาการทางตา: รายงานผู้ป่วยและการรวบรวมข้อมูล

วรรณกรณ์ พฤกษากร, สุภรัตน์ จริยโกศล

วัตถุประสงค์: เพื่อรายงานผู้ป่วยถุงน้ำชนิดนิวเรนเทอริคขนาดใหญ่บริเวณสมองส่วนหน้า ซึ่งมาด้วยอาการที่พบได้น้อย

รายงานผู้ป่วย: ผู้ป่วยชายไทยอายุ 47 ปี สุขภาพแข็งแรงดีมาตลอด มีอาการมองเห็นลดลงมาเป็นระยะเวลา 6 เดือน และพบตาเขเข้าในเป็นเวลา 1 ปี ผลตรวจร่างกายและการตรวจทางระบบประสาทอื่นๆ อยู่ในเกณฑ์ปกติ ไม่ตรวจระดับความผิดปกติของฮอร์โมนใดๆ เอกซเรย์คลื่นไฟฟ้าแม่เหล็กที่บริเวณสมองพบถุงน้ำขนาดใหญ่บริเวณเซลล่า เหนือต่อเซลล่า และสมองส่วนหน้า เส้นประสาทตาทั้ง 2 ข้างมีขนาดเล็ก

ผลการศึกษา: ผู้ป่วยได้รับการผ่าตัดระบายน้ำในถุงน้ำที่สมองและตัดชิ้นเนื้อบางส่วนของถุงส่งตรวจทางพยาธิ ผลทางพยาธิวิทยาเข้าได้กับถุงน้ำชนิดนิวเรนเทอริค

สรุป: ถุงน้ำชนิดนิวเรนเทอริคที่บริเวณสมองส่วนหน้าเป็นถุงน้ำชนิดที่พบได้ไม่บ่อยนัก และเมื่อพิจารณาขนาดของถุงน้ำที่มีขนาดใหญ่ที่ควรมีอาการผิดปกติทางร่างกายหลายอย่าง เป็นที่น่าแปลกใจว่าผู้ป่วยรายนี้ไม่มีอาการทางระบบประสาทหรืออาการผิดปกติทางร่างกายอื่นใด มีเพียงอาการทางตาเท่านั้น ฉะนั้น จึงควรคำนึงถึงโรคนี้ในผู้ป่วยที่มีอาการผิดปกติเพียงทางตาไว้ด้วย