Rapid Visual Deterioration Caused by Posterior Fossa Arachnoid Cyst

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INTRODUCTION

Arachnoid cysts are benign, non-neoplastic and extra-axial lesions; most of them are clinically silent and unchanged in size. In previous studies 1,2,13,14,24, the incidence of arachnoid cysts has been estimated to range between 0.3% to 1.7%. However, the advanced neuroimaging techniques have facilitated the diagnosis of arachnoid cysts, which, in turn, has led to an increase in the number of patients 2,12,14,19.

In some cases, an arachnoid cyst arouses symptoms and its clinical presentations vary according to the size and location of the cyst. The most frequent symptom is headache, which is caused by the local mass effect, increased intracranial pressure (ICP), and/or hydrocephalus 19. Above this, seizures, cognitive dysfunction, developmental delay, and intracranial hemorrhage can be also presented.

In this report, we describe a case where a patient with a posterior fossa arachnoid cyst complained of visual field defect, visual disturbance, and mild headache; no ataxia and unsteady gait were reported. The patient's visual symptoms rapidly deteriorated in a week and, consequently, the patient's vision got worse.

CASE REPORT

A 39-year-old male patient presented at our emergency room after a traffic accident and the brain computed tomography (CT) scan indicated an incidental arachnoid cyst on posterior fossa. At that time, the patient complained only of facial abrasions and the left knee pain, but not of headache or visual disturbance. In eight months, however, he revisited our Department of Ophthalmology to check the decreased visual acuity, visual field defect, and headache. On the ophthalmologic examination, bilateral papilledema (Fig. 1A), decreased visual acuity, and visual field defect (Fig. 2A) were found. After the transfer to the Department of Neurosurgery, we rechecked brain CT scans (Fig. 3). The diameter of the patient's arachnoid cyst did not change compared to the previous CT scans; however, the slightly enlarged third ventricle was compressing the optic chiasm and the suprasellar area. Since visual complications were expected, we recommended an emergency operation. However, the patient refused the operation for personal reasons. One week later, he came back to the hospital with nearly complete visual loss. His right eye was completely blind, whereas his left eye had only 10/100 vision.
eye recognized only light (Fig. 2B).

On that day, he underwent an emergency operation. A conventional suboccipital craniectomy was performed on the patient lying on the operating table in the prone position. After the dural incision, the arachnoid cyst was exposed and cystic fluid was expelled. Arachnoid membrane was carefully dissected and the arachnoid cyst was fenestrated to cisterna magna for connecting with the cerebrospinal fluid (CSF) flows. Arachnoid membrane which covering the foramen of magendie was also fenestrated and the CSF flows was running properly (Fig. 4). Excluding the brain tumor, the collected membrane was sent to the pathologic laboratory. In high power field microscopy under the hematoxylin and eosin stain, the cystic membrane was composed by membranous fibrocollagenous tissues without tumor cells. Finally, the patient was diagnosed as arachnoid cyst (Fig. 5).

One week afterwards, we checked the brain CT scans and the removal of the cyst and the decreased ventricle size were confirmed. On the ophthalmologic examination, the papilledema and visual field were slightly improved; however, the patient’s visual acuity did not improve. Upon the tenth day after the op-

Fig. 1. Fundoscopy. A : One week before the operation. B : Five months after the operation.

Fig. 2. Visual field test. A : One week before the operation. B : Operation date. C : Five months after operation.

Fig. 3. Preoperative enhanced CT scans. A : Sagittal cutting image on the midline. Black arrow : the enlarged third ventricle is compressing the optic chiasm and sella turcica. B : Coronal cutting image on the optic chiasm. Pituitary gland is compressed by the third ventricle and sella turcica is filled with the cerebrospinal fluid. C : Axial cutting image on the cerebellar level. White arrow : a large arachnoid cyst is compressing the fourth ventricle and circumferential cerebellum.
In five months after the operation, papilledema (Fig. 1B), headache, and radiological images (Fig. 6) quickly returned to the normal ranges. Nevertheless, visual acuity and visual field improved slowly and slightly (Fig. 2C). The blind right eye improved to recognize the light and the blurred left eye improved to acknowledge the silhouette of an object; thus, it did not fully recover to the prior, normal condition.

**DISCUSSION**

Arachnoid cysts are benign, non-neoplastic and extra-axial lesions. Their prevalence has been reported to amount to approximately 1% of all intracranial mass lesions\(^ {1,12,13,24,30}\); however, advanced neuroimaging techniques and the widespread use of CT and MRI imaging have led to an even higher incidence of report of these lesions\(^ {1,2,6,12,13,19,20,30,31}\). One recent study, Al-Holou et al.\(^ {11}\), reviewed 48417 brain images and identified 661 (1.4%) arachnoid cysts. This report found that the prevalence of arachnoid cyst in children was 2.6%, slightly higher than in adults, and men (1.8%) have a high prevalence than women (1.1%). The most common locations were middle fossa (34%), retrocerebellar (33%), and cerebral convexity (14%)\(^ {1,24}\). Despite the increased identification of arachnoid cysts, the natural history and mechanism of enlargement has not been well defined\(^ {1,13,15,24}\).

Most arachnoid cysts are asymptomatic and are found incidentally in most cases\(^ {1,12,24,29}\). Nevertheless, when arachnoid cysts give rise to any symptom, headache is the most common complaint. This symptom is caused by the local mass effect and hydrocephalus. In addition to headache, gait imbalance, seizure, and visual changes can also be presented according to the location and size of the arachnoid cyst. If clinical symptoms develop, surgical interventions are recommended\(^ {3,6,15,16,21-23,27,28}\). Various operation techniques, such as cyst excision\(^ {15,17}\), stereotactic aspiration\(^ {18,25}\), endoscopic cyst fenestration\(^ {5,12}\), ventriculocystostomy\(^ {26}\), and cystoperitoneal shunt\(^ {8,10,13,27,28}\), have been introduced and the main purpose of such operations is similar, namely, a direct decompression of the arachnoid cyst or making the connection between the cyst and the CSF flows for spontaneous resolution.

Generally, visual symptoms occur in the patients with suprasellar arachnoid cysts, because the arachnoid cyst is adjacent to the optic nerves\(^ {7}\). The patients with posterior fossa arachnoid cysts usually do not complain of visual symptoms, as the cyst is not adjacent to the optic nerves. However, a posterior fossa arachnoid cyst can indirectly disrupt the CSF flows and, in turn, the disruption increases the whole ventricle size and the ICP. In our case, the enlarged third ventricle was compressing the optic chiasm with cephalocaudal direction on the CT scan (Fig. 3A, B). The compressed superior part of optic chiasm led to the defect of inferior visual field (Fig. 2). In addition, visual acuity was affected by the papilledema, which was caused by the increased ICP. Consequently, both visual acuity and visual field were influenced by the posterior fossa arachnoid cyst.

When the patient first visited our Department of Ophthalmology, his visual symptoms and headache were not severe and
the patient could walk and drive. Besides, the brain CT scan showed no changes as compared to previous CT scans, so we could not predict a risk of rapid deterioration. The patient’s refusal to have an operation and the surgeon’s wrong prediction delayed the appropriate on-time treatment; ultimately, the patient’s vision was severely damaged during the critical seven days. After the operation, the patient’s visual acuity, visual field, and papilledema were slightly improved; however, we regard that the patient’s vision will unfortunately not fully recover.

CONCLUSION

Unlike gait disturbance and ataxia, visual symptoms do not frequently occur in the patients with posterior fossa arachnoid cysts; this is so because the cyst is located away from the optic nerves. However, the disruption of CSF flows can indirectly induce visual symptoms and these symptoms can rapidly deteriorate.

Considering the results of our case, it should be emphasized that the patients with posterior fossa arachnoid cysts could develop a rapid neurologic deterioration. Furthermore, the aggravation pace of the symptoms can be neither estimated nor predicted. Therefore, in the event of the development of symptoms, an early intervention or close observation are crucial.

References

1. Al-Holou WN, Terman S, Kilburg C, Garton HJ, Muraszko KM, Maher CO: Prevalence and natural history of arachnoid cysts in adults. J Neurosurg 118: 222-231, 2013
2. Al-Holou WN, Yew AY, Boomsaad ZE, Garton HJ, Muraszko KM, Maher CO: Prevalence and natural history of arachnoid cysts in children. J Neurosurg Pediatr 5: 578-585, 2010
3. Arai H, Sato K, Wachi A, Okuda O, Takeda N: Arachnoid cysts of the middle cranial fossa: experience with 77 patients who were treated with cystoperitoneal shunting. Neurosurgery 39: 1108-1112; discussion 1112-1113, 1996
4. Banna M: Arachnoid cysts on computed tomography. AJR Am J Roentgenol 127: 979-982, 1976
5. Caemaert J, Abdullah J, Calliawu L, Carton D, Dhooge C, van Coster R: Endoscopic treatment of suprasellar arachnoid cysts. Acta Neurochir (Wien) 119: 68-73, 1992
6. Choi IU, Kim DS, Huh R: Endoscopic approach to arachnoid cyst. Childs Nerv Syst 15: 285-291, 1999
7. Choi KY, Jung S, Kang SS, Kim JY, Jung TY, Jang WY: Technical considerations to prevent postoperative endocrine dysfunction after the fenestration of suprasellar arachnoid cyst. J Korean Neurosurg Soc 49: 262-266, 2011
8. Cricillo SF, Cogen PH, Harsh GR, Edwards MS: Intracranial arachnoid cysts in children. A comparison of the effects of fenestration and shunting. J Neurosurg 74: 230-235, 1991
9. Eskandary H, Sabha M, Khajehpour F, Eskandari M: Incidental findings in brain computed tomography scans of 3000 head trauma patients. Surg Neurol 63: 550-553; discussion 553, 2005
10. Fewel ME, Levy ML, McComb JG: Surgical treatment of 95 cases of posterior fossa arachnoid cysts. Pediatr Neurosurg 25: 165-173, 1996
11. Galassi E, Tognetti F, Frank F, Fagioli L, Nasi MT, Gaist G: Infratentorial arachnoid cysts. J Neurosurg 63: 210-217, 1985
12. Gangemi M, Seneca V, Colella G, Gioffi V, Imperato A, Maiuri F: Endoscopy versus microsurgical cyst excision and shunting for treating intracranial arachnoid cysts. J Neurosurg Pediatr 8: 158-164, 2011
13. Halani SH, Safain MG, Heilman CB: Arachnoid cyst slit valves: the mechanism for arachnoid cyst enlargement. J Neurosurg Pediatr 12: 62-66, 2013
14. Hanieh A, Simpson DA, North JB: Arachnoid cysts: a critical review of 41 cases. Childs Nerv Syst 4: 92-96, 1988
15. Harsh GR 4th, Edwards MS, Wilson CB: Intracranial arachnoid cysts in children. J Neurosurg 64: 835-842, 1986
16. Helland CA, Waster K: A population-based study of intracranial arachnoid cysts: clinical and neuroimaging outcomes following surgical cyst decompression in children. J Neurosurg 105 (5 Suppl): 385-390, 2006
17. Hinojosa J, Esparza J, Muñoz MJ, Valencia J: [Endoscopic treatment of suprasellar arachnoid cysts]. Neurocirugia (Astur) 12: 482-488; discussion 489, 2001
18. Iacono RP, Labadie EL, Johnstone SJ, Bendt TK: Symptomatic arachnoid cyst at the clivus drained stereotactically through the vertex. Neurosurgery 27: 130-133, 1990
19. Katzman GL, Dagher AP, Patronas NJ: Incidental findings on brain magnetic resonance imaging from 1000 asymptomatic volunteers.

Fig. 6. Five months after operation. A: Sagittal cutting image on the midline. Black arrow: whole ventricles become smaller and there is no more compression of the optic chiasm. B: Coronal cutting image on the optic chiasm. C: Axial cutting image on the cerebellar level. White arrow: the arachnoid cyst has completely disappeared in the cerebellum and the previously compressed fourth ventricle is released.
20. Kim BS, Illes J, Kaplan RT, Reiss A, Atlas SW: Incidental findings on pediatric MR images of the brain. *AJNR Am J Neuroradiol* 23: 1674-1677, 2002
21. Levy ML, Meltzer HS, Hughes S, Aryan HE, Yoo K, Amar AP: Hydrocephalus in children with middle fossa arachnoid cysts. *J Neurosurg* 101 (1 Suppl): 25-31, 2004
22. Levy ML, Wang M, Aryan HE, Yoo K, Meltzer H: Microsurgical keyhole approach for middle fossa arachnoid cyst fenestration. *Neurosurgery* 53: 1138-1144; discussion 1144-1145, 2003
23. Oberbauer RW, Haase J, Pucher R: Arachnoid cysts in children: a European co-operative study. *Childs Nerv Syst* 8: 281-286, 1992
24. Panigrahi S, Mishra SS, Das S, Patra SK, Satpathy PC: Large intradural craniospinal arachnoid cyst: a case report and review of literature. *J Craniovertebr Junction Spine* 3: 16-18, 2012
25. Pell MF, Thomas DG: The management of infratentorial arachnoid cyst by CT-directed stereotactic aspiration. *Br J Neurosurg* 5: 399-403, 1991
26. Pierre-Kahn A, Capelle L, Brauner R, Sainte-Rose C, Renier D, Rappaport R, et al.: Presentation and management of suprasellar arachnoid cysts. Review of 20 cases. *J Neurosurg* 73: 355-359, 1990
27. Raffel C, McComb JG: To shunt or to fenestrate: which is the best surgical treatment for arachnoid cysts in pediatric patients? *Neurosurgery* 23: 338-342, 1988
28. Sato H, Sato N, Katayama S, Tamaki N, Matsumoto S: Effective shunt-independent treatment for primary middle fossa arachnoid cyst. *Childs Nerv Syst* 7: 375-381, 1991
29. Starkman SP, Brown TC, Linell EA: Cerebral arachnoid cysts. *J Neuropathol Exp Neurol* 17: 484-500, 1958
30. Weber F, Knopf H: Incidental findings in magnetic resonance imaging of the brains of healthy young men. *J Neurol Sci* 240: 81-84, 2006
31. Zada G, Krieger MD, McNatt SA, Bowen I, McComb JG: Pathogenesis and treatment of intracranial arachnoid cysts in pediatric patients younger than 2 years of age. *Neurosurg Focus* 22: E1, 2007