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Original Article

Surgical management of the fourth ventricle arachnoid cysts

Sukru Oral[1], Hanifi Bayarogullari[2], Atilla Yilmaz[1], Boran Urfali[1], Mustafa Aras[1], Yurdal Serarslan[1]

[1]Hatay Mustafa Kemal University, Tayfur Ata Sokmen Faculty of Medicine, Department of Neurosurgery, Hatay, Turkey
[2]Hatay Mustafa Kemal University, Tayfur Ata Sokmen Faculty of Medicine, Department of Radiology, Hatay, Turkey

Abstract

Aim: The arachnoid cysts are developmental masses that occur from the splitting or duplication of the arachnoid membrane. They may occur in different locations such as middle cranial fossa, retrocerebellar and convexity region, and they are usually asymptomatic. Arachnoid cysts are uncommon in association with intraventricular location especially fourth ventricle. In this study, we present six consecutive cases with arachnoid cyst in the fourth ventricle.

Material and Methods: There were four female and two male patients. The average age of patients was 37 years ranging from2 to 65 years. These patients were periodically followed-up. The follow-up period ranged from minimum 2 to 6 years. Three patients were operated by a ventriculoperitoneal shunt and surgical excision was performed in two patients. Remaining two patients were followed up because they were asymptomatic.

Results: Revision surgery was made due to shunt malfunction in a patient. The symptoms had been regressed in all patients. The arachnoid cysts were completely regressed after the operations in two patients.

Conclusion: Most of arachnoid cysts which were small and asymptomatic did not require treatment. However, the size of an arachnoid cyst typically remained stable or increased over the time. An asymptomatic cyst may become symptomatic after minor head trauma. So, the asymptomatic patients with fourth ventricle arachnoid cyst should be under periodic follow up for obstructive hydrocephalus.

Keywords: Arachnoid Cyst; Fourth Ventricle; Ventriculoperitoneal Shunt; Surgical Excision.

INTRODUCTION

Arachnoid cysts are expressed as congenital extra axial benign cystic cavities formed by the accumulation of cerebrospinal fluid (CSF) in the intrauterine life. They are localized between arachnoid membranes (1). The prevalence of intracranial arachnoid cysts is described with 0.5–1% in the general population (2). Arachnoid cysts are more often seen in the pediatric population as compared to adults. They may occur in different locations such as middle cranial fossa, retro cerebellar and convexity region, and they are usually asymptomatic. Most common location of the arachnoid cysts is middle cranial fossa (30-50%) (3,4). Arachnoid cysts are uncommon in association with intraventricular location especially fourth ventricle. To the literature, only thirteen cases have been reported to this date (5). Although various surgical treatments have been suggested for intracranial arachnoid cyst, the optimal surgical method is still a matter of debate. In this study, we report six patients who have an arachnoid cyst located in the fourth ventricle and share here our experiences in follow up periods and surgical techniques such as V-P (ventriculoperitoneal shunt) or surgical excision.

MATERIAL and METHODS

Ethical approval was not necessary for preparation of this article. Because, this is a retrospective study and no new treatment was used. We presented six consecutive cases with arachnoid cyst in the fourth ventricle. All of the patients were diagnosed in our institute during the years of 2011 and 2017. The symptoms of the patients were vertigo, headache, and vomiting. But, two patients had no obvious clinical symptoms (Figure 1). Computerized tomography (CT) and magnetic resonance imaging (MRI) were used for diagnostic evaluation of arachnoid cysts. There were three female and two male patients. The average age of patients was 37 years. ranging from 2 to 65 years. These patients were periodically followed-up. The follow-up period ranged from minimum 2 to 6 years.

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Corresponding Author: Sukru Oral, Hatay Mustafa Kemal University, Tayfur Ata Sokmen Faculty of Medicine, Department of Neurosurgery, Hatay, Turkey, **E-mail:** sukruor@yahoo.com

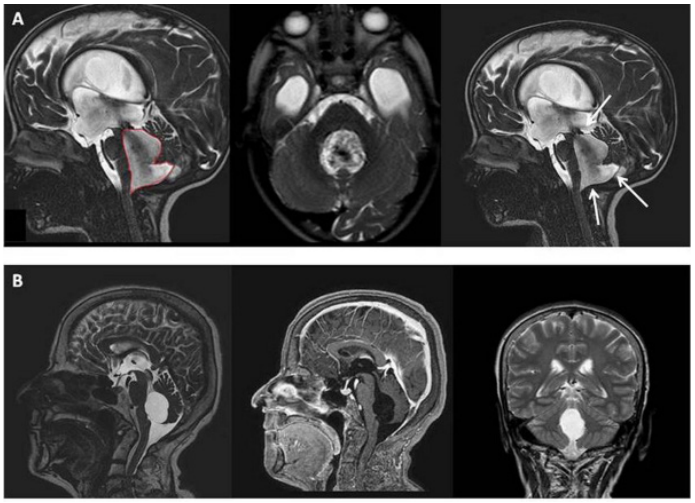

Figure 1. A) five years old/Ffemale, asymptomatic, T2 weighted sagittal and axial MRI slices showed a cyst within the fourth ventricle. B) 62 years old/female, asymptomatic, the location of the cyst in the fourth ventricle was seen in T2-sagittal, coronal and T1-sagittal with contrast images

RESULTS

One of the patients was admitted to the emergency department due to onset of acute hydrocephaly. He was treated by a ventriculoperitoneal shunt (Figure 2).

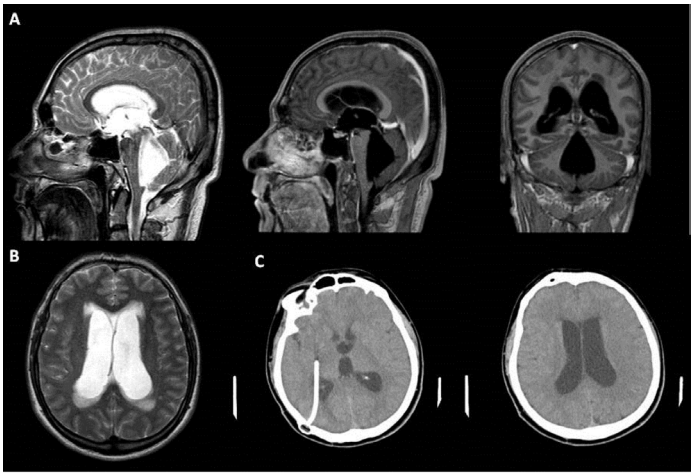

Figure 2. 65 years old and male, the arachnoid cyst is seen in the fourth ventricle and disrupt the circulation of CSF is seen in MRI images A) T2 sagittal, T1 sagittal and coronal with contrast images B) Periventricular edema is seen in T2 axial image C) V-P shunt was seen on CT images after the first operation

The symptoms due to ICP-elevation improved, after 3 days of operation. The patient was followed during two years after the operation. At the end of 2 years, he was admitted to the outpatient clinic with visual impairment complaint. He was re-operated due to ventriculoperitoneal shunt malfunction (Figure 3). Another patient was fifty one years old. He had no symptoms throughout 3 years of follow up (Figure 4). But, he was admitted to our clinic due to repeated vomiting and headache at the end of 3 years. Her physical and neurological examination results revealed no abnormality. A ventriculoperitoneal shunt was performed shortly thereafter. The symptoms had been regressed within two days. The arachnoid cyst was operated after 3 months (Figure 5).

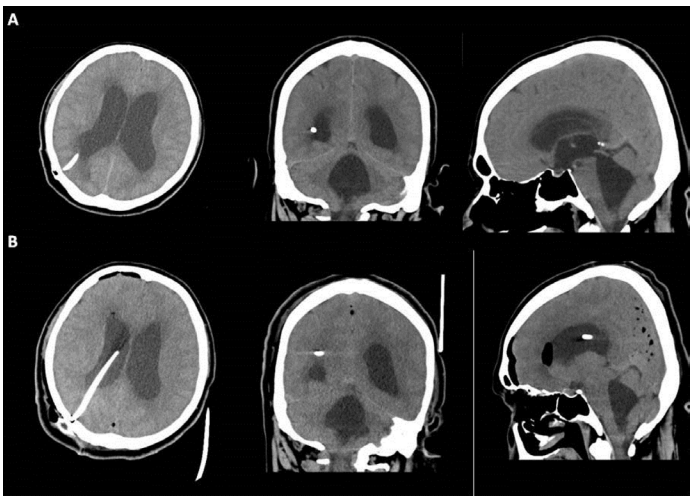

Figure 3. 65 years old male, A) Hydrocephalus and shunt malfunction are shown on CT images B: The shunt malfunction was cured

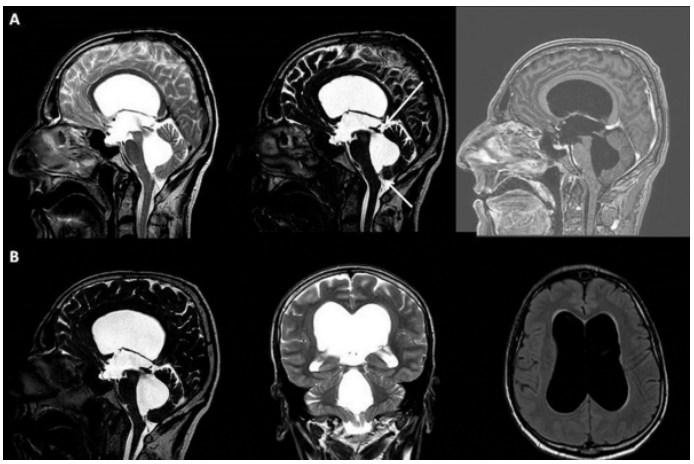

Figure 4. 51 years old and male A) Sagittal T1-weighted images showing a fourth ventricle arachnoid cyst during the asymptomatic period B) Hydrocephaly was seen in T2 sagittal, coronal and T1 flair sections after 3 years

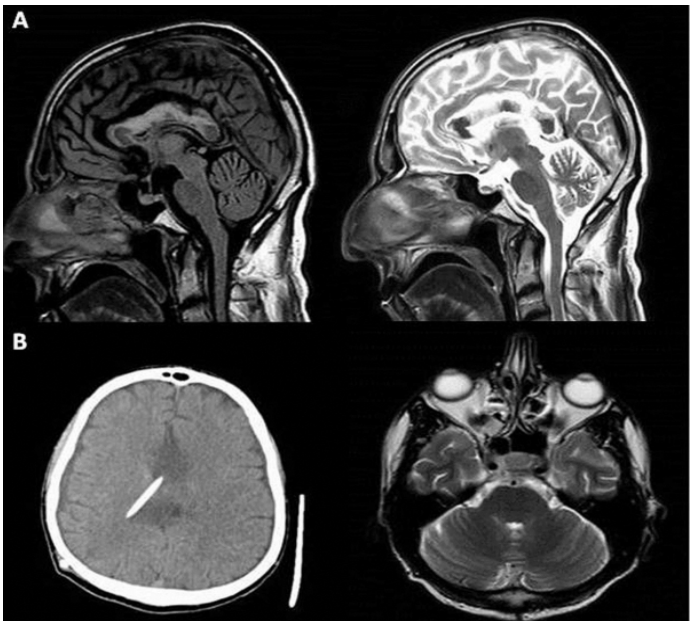

Figure 5. 51 years old and male. A) The arachnoid cyst which was excised after 3 months are shown in T1 sagittal and T2 sagittal, axial images (B). B) V-P shunt was seen on CT sections after three months of operation

Another patient who has an arachnoid cyst in the fourth ventricle was operated due to vertigo and vomiting. The cyst was excised partially via sub-occipital craniectomy (Figure 6) Postoperative Cerebrospinal Fluid (CSF) leak occurred in the patient. (Figure 6) The CSF leak was treated with external lumbar drainage for 5 days. The demographic and clinical information about the patients in this study are described in Table 1.

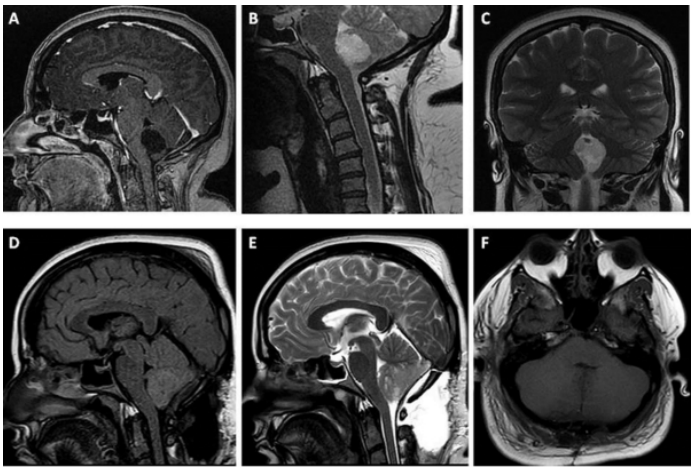

Figure 6. 33 years old and female (A,B,C) The arachnoid cyst in the fourth ventricle was seen on the MRI images (D,E,F) T1-T2 Sagittal and axial images showed the reduction in size of the cyst and CSF leak under the skin

Table 1. The demographic and clinical information about the patients

Age/sex	Symptoms	Treatment	Complication	Follow up period
5/Female	Asymptomatic	no	no	3 years
62/Female	Asymptomatic	no	no	4 years
51/Male	Vomiting Headache	v-p shunt Surgical excision	no	4 years
65/Male	Headache Vomiting Vertigo	v-p shunt	Shunt malfunction after two years and Shunt revision	6 years
2/Female	Vomiting Seizure Abnormal Increased head circumference	v-p shunt	no	2 years
33/Female	Vertigo and Vomiting	Surgical Excision	Cerebrospinal Fluid Leak	3 years

DISCUSSION

The arachnoid cysts are developmental masses that occur from the splitting or duplication of the arachnoid membrane (6). The formation of arachnoid cysts is still in debate. Intracranial arachnoid cyst accounts for approximately 1% of atraumatic intracranial mass lesions. They can cause nonspecific symptoms such as headache, nausea, vomiting, seizure, dizziness and hearing-vision problems, but there is usually no symptom in most cases. The male to female ratio is approximately 3:1 (4). Most arachnoid cysts are usually found incidentally on neuroimaging, such as CT, MRI and cine phase contrast MR imaging that was performed for a different reason. Intraventricular arachnoid cyst is used a general term for several types of cysts such as ependymal cysts, simple cysts, neuroepithelial cysts and choroid plexus cysts. These cysts have identical imaging and operative features, but they are histologically distinct. Although, there was no arachnoid tissue within the ventricular system, some studies reported that the origin of cysts was thought most likely to arise from vascular mesenchyme (7). Intraventricular arachnoid cysts are usually asymptomatic. When the arachnoid cyst becomes symptomatic, the patient may present with vomiting, headache, focal neurological deficit, seizure and symptoms of obstructive hydrocephalus (8). Similar symptoms were found in our cases such as headache, nausea, vomiting, and seizure. Furthermore, according to the location of the cyst, specific symptoms can occur. Arachnoid cysts located in the fourth ventricle have been rarely reported in the data. Until this date, only 13 cases have been reported in the literature (9-16). Numerous surgical techniques have been reported including cysto-peritoneal shunting, cyst fenestration, ventriculo-peritoneal shunting, cyst excision and marsupialization into the subarachnoid space. However, it remains unclear which is the best technique (11,17-19). A lot of researches have been done to evaluate the advantages and disadvantages of these different methods (19,20).

Bonde et al., reported that excision or marsupialization of the arachnoid cyst was curative in four patients with fourth ventricle arachnoid cyst (11). Sugimoto et al. (21) suggested that arachnoid cysts of the fourth ventricle was important to recognize them, because they cause normal pressure hydrocephalus symptoms and cerebellar or brainstem deficit. The ventriculo-peritoneal shunt was performed to the five patients, but, success was only temporary or regression of symptoms was incomplete in the data analysis. Finally, partial or complete surgical excision of arachnoid cyst was performed to several patients via a median suboccipital approach (11,14,16,22). We performed a ventriculo-peritoneal shunt to two patient and their symptoms was significantly improved after the operation whereas the size of the cyst decreased in one patient. However, Westermaier et al. (16) reported that the ventriculoperitoneal shunting procedure only provided transient improvement of symptoms in the patients. Several patients were initially treated by a V-P shunt in overview of cases published in literature. While the symptoms due to elevated ICP (Increased Intracranial Pressure) improved after V-P shunt, clinical symptoms of cerebellar dysfunction worsened such as truncal ataxia, progressive vertigo. So, partial or complete cyst excision was performed to the patients. Furthermore, there were no signs of cerebellar dysfunction in our patients and the symptoms complete recovery after the V-P shunt.

Today, endoscopic fenestration, endoscopic multiple wide fenestration and endoscopic third ventriculostomy are promoted as a safe and effective option for the treatment

of arachnoid cysts in many locations (4,16,20,23,24). But, Endoscopic procedures could perform successfully in suprasellar arachnoid cysts.

Differential diagnosis of fourth ventricle arachnoid cysts also includes Blake's pouch cyst (BPC), differentiation from which often difficult (25). The arachnoid cyst represents a CSF like fluid collection within arachnoid layers (26). In the literature, localization of BPC has been infra or retro cerebellar and these patients have minimal fourth ventricle dilatation (27). If a patient has large fourth ventricle dilatation and BPC, these patients are identified as the Dandy-Walker Syndrome variant (28). While you can see the posterior margin of BPC, in our patient, the superior, inferior and posterior margin of arachnoid cyst can be seen obviously (Figure 1A). Besides our patient has no vermian and cerebellar hypoplasia.

Another pathologic condition in the differential diagnosis is the obstruction of Magendie and Luschka's foramens. While this condition usually presents as congenital in children, in adults mostly acquired such as an inflammatory process, head trauma or hemorrhage (29,30). There was no history of infection, hemorrhage or head injury in our patient. Besides, the membrane which leads to obstruction of the foramina Luschka and Magendie could only have been seen on the sagittal T1-weighted MR images. On T2 weighted MR images, the membrane was not so evident (31). But, the borders of cyst wall were seen obviously in our patients on T2 images. (Figure 3,4).

CONCLUSION

There are several suggested surgical strategies in literature like total excision without shunt using or stereotactic cyst-ventricular shunting (32,33). In this study, we excised partially the arachnoid cysts in two patients. Finally, the goal of these techniques was a successful outcome to treat non-communicating hydrocephalus. So, any of these methods could be performed. But, due to their complications, we must follow up such these patients regularly.

The fourth ventricular arachnoid cysts are relatively infrequent lesions when compared with the other arachnoid cysts. Most of arachnoid cysts which were small and asymptomatic do not require treatment. However, the size of an arachnoid cyst typically remains stable or increases over time and an asymptomatic cyst may become symptomatic after minor head trauma. So, the asymptomatic patients with fourth ventricle arachnoid cyst should be under periodic follow up for obstructive hydrocephalus.

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Sukru Oral ORCID: 0000-0003-4328-0690
Hanifi Bayarogullari ORCID: 0000-0001-6147-5876
Atilla Yilmaz ORCID: 0000-0002-1081-3545
Boran Urfali ORCID: 0000-0003-2854-6833
Mustafa Aras ORCID: 0000-0002-8360-148X
Yurdal Serarslan ORCID: 0000-0002-3865-7206

REFERENCES

1. JA. Gosalakkal. Intracranial arachnoid cysts in children. A review of pathogenesis, clinical features, and management. Pediatr Neurol 2002;26:93-8.
2. Helland CA, Lund-Johansen M, Wester K. Location, sidedness, and sex distribution of intracranial arachnoid cysts in a population-based sample. J Neurosurg 2010;113:934-9.
3. Gandy SE, Heier LA. Clinical and magnetic resonance features of primary intracranial arachnoid cysts. Ann Neurol 1987;21:342-8.
4. Cincu R, Agrawal A, Eiras J. Intracranial arachnoid cysts: current concepts and treatment alternatives. Clin Neurol Neurosurg 2007;109:837-43.
5. Aljohani H, Romano A, Iaccarino C, et al. Pure Endoscopic management of fourth ventricle arachnoid cyst: case report and literature review. Asian J Neurosurg 2018;13:184-7.
6. Tarkman Sp, Brown Tc, Linell Ea. Cerebral arachnoid cysts. J Neuropathol Exp Neurol 1958;17:484-500.
7. Okamura K, Watanabe M, Inoue N, et al. Intraventricular arachnoid cyst--on the origin of intraventricular arachnoid cysts. No to shinkei 1996;48:1015-21.
8. Park SW1, Yoon SH, Cho KH, et al. A large arachnoid cyst of the lateral ventricle extending from the supracerebellar cistern--case report. Surg Neurol 2006;65:611-4.
9. Acar O, Kocaoğullar Y, Güney O. Arachnoid cyst within the fourth ventricle: a case report, Clin Neurol Neurosurg 2003;105:93-4.
10. Azzimondi G, Calbucci F, Rinaldi R, Arachnoid cyst in the region of the fourth ventricle mimicking clinical picture of normal pressure hydrocephalus. Neurosurg Rev 1995;18:269-71.
11. V Bonde, D Muzumdar, A Goel. Fourth ventricle arachnoid cyst, Journal of clinical neuroscience : official journal of the Neurosurgical Society of Australasia 2008;15:26-8.
12. Di Rocco C, Di Trapani G, Iannelli A. Arachnoid cyst of the fourth ventricle and "arrested" hydrocephalus. Surg Neurol 1979;12:467-71.
13. S. Gulsen. Symptomatic Fourth Ventricle Arachnoid Cyst Extending to the Upper Border of the Dens of the Axis: A Case Report and Review of the Literature, Maced J Med Sci 2014., p. 301.
14. Korosue K, Tamaki N, Fujiwara K, et al. Arachnoid cyst of the fourth ventricle manifesting normal pressure hydrocephalus. Neurosurgery 1983;12:108-10.
15. F Lockhat, L Mubaiwa, B Clarke, et al. Imaging of a fourth ventricle arachnoid cyst in a child, Clin Radiol Extra 2005;60:35-7.
16. Westermaier T, Vince GH, Meinhardt M, et al. Arachnoid cysts of the fourth ventricle - short illustrated review. Acta Neurochir (Wien) 2010;152:119-24.
17. Srinivasan US, Lawrence R. Posterior fossa arachnoid cysts in adults: Surgical strategy: Case series, Asian J Neurosurg 2015;10:47.
18. Martinoni M, Toni F, Lefosse M, et al. Endoscopic treatment of a fourth ventricle arachnoid cyst via the third ventricle: a case report. Neurosurgery 2014;10:E374-7.
19. Samii M, Carvalho GA, Schuhmann MU, et al. Arachnoid cysts of the posterior fossa, Surg Neurol 1999;51:376-82.
20. Choi JU, Kim DS, Huh R, Endoscopic approach to arachnoid cyst. Childs Nerv Syst 1999;15:285-91.
21. Sugimoto T, Uranishi R, Yamada T. Gradually Progressive Symptoms of Normal Pressure Hydrocephalus Caused by an Arachnoid Cyst in the Fourth Ventricle: a Case Reporti. World Neurosurg 2016;85:364.e19-22.

22. Nadkarni T, Hande A, Nagpal R. Arachnoid cyst within the fourth ventricle--a case report. Br J Neurosurg 1995;9:675-8.
23. Toyota S, Taki T, Oshino S, at al. A neuroendoscopic approach to the aqueduct via the fourth ventricle combined with suboccipital craniectomy. Minim Invasive Neurosurg 2004;47:312-5.
24. SJ Choi, C Ji, JG Ahn, MC, et al. Endoscopic multiple wide fenestrations of intraventricular arachnoid cyst arising in lateral ventricle. J Korean Neurosurg Soc 2004;35:614-7.
25. Ramaswamy S, Rangasami R, Suresh S, et al. Spontaneous resolution of Blake's pouch cyst. Radiol Case Rep 2015;8:877.
26. Cornips EM, Overvliet GM, Weber JW, at al. The clinical spectrum of Blake's pouch cyst: report of six illustrative cases. Childs Nerv Syst 2010;26:1057-64.
27. Azab WA, Shohoud SA, Elmansoury TM, et al. Blake's pouch cyst, Surg Neurol Int 2014;25:112.
28. Calabrò F, Arcuri T, Jinkins JR. Blake's pouch cyst: an entity within the Dandy-Walker continuum. Neuroradiology 2000;42:290-5.
29. Kasapas K, Varthalitis D, Georgakoulias N, et al. Hydrocephalus due to Membranous Obstruction of Magendie's Foramen. J Korean Neurosurg Soc 2015;57:68-71.
30. Orakdogen M, Emon ST, Erdogan B, et al. Fourth ventriculostomy in occlusion of the foramen of magendie associated with chiari malformation and syringomyelia. NMC Case Rep J 2015,2:72-5.
31. Rougier A, Ménégon P. MRI evidence of membranous occlusion of the foramen of Magendie. Acta Neurochir (Wien) 2009;151:693-4.
32. D'Angelo V, Gorgoglione L, Catapano G. Treatment of symptomatic intracranial arachnoid cysts by stereotactic cyst-ventricular shunting. Stereotact Funct Neurosurg 1999;72:62-9.
33. Kurokawa Y, Sohma T, Tsuchita H, A case of intraventricular arachnoid cyst. How should it be treated? Childs Nerv Syst 1990;6:365-7.