

# Syringomyelia caused by an arachnoid web in a patient with shunted Dandy-Walker malformation

Hee Chang Lee<sup>1</sup> · Jung Won Choi<sup>1</sup> · Ji Yeoun Lee<sup>1</sup> · Ji Hoon Phi<sup>1</sup> · Seung-Ki Kim<sup>1</sup> ·  
Byung-Kyu Cho<sup>1</sup> · Kyu-Chang Wang<sup>1</sup>

Received: 8 August 2016 / Accepted: 26 October 2016 / Published online: 7 November 2016  
© Springer-Verlag Berlin Heidelberg 2016

## Abstract

**Introduction** Dandy-Walker malformation (DWM) is a congenital brain anomaly characterized by dysgenesis of the cerebellar vermis and the presence of a posterior fossa cyst. The association of syringomyelia with DWM is extremely rare.

**Case report** A 10-year-old patient who was diagnosed with DWM in infancy presented with progressive scoliosis and fecal incontinence. He had been treated with cystoventriculoperitoneal shunting with a Y-connection during infancy, which was followed by a revision 6 years later. During the revision surgery, intraventricular bleeding occurred and was managed conservatively. Imaging studies for the current visit revealed syringomyelia along the cervicothoracic spinal cord and a membranous structure around the cervicomedullary junction. Phase-contrast cine magnetic resonance imaging (MRI) revealed disturbed cerebrospinal fluid (CSF) flow across the membrane. We excised the arachnoid web that was tethering the brainstem and blocking CSF flow. Postoperatively, the patient experienced symptom relief, and the follow-up imaging study demonstrated a dramatic decrease in the size of the syringomyelia.

**Discussion** We suggest that syrinx formation in this patient was possibly caused by disturbed CSF flow and tethering of the brainstem.

**Conclusion** We experienced an unusual case of DWM with syringomyelia which was caused by an arachnoid web blocking CSF flow and tethering the brainstem. The arachnoid

web seems to be formed by previous bleeding which occurred at the time of shunt revision. After excision of the arachnoid web, the patient showed good outcome.

**Keywords** Dandy-Walker malformation · Syringomyelia · Arachnoid web · Membrane excision

## Introduction

Dandy-Walker malformation (DWM) is a congenital brain defect characterized by complete or partial agenesis of the cerebellar vermis and the presence of a posterior fossa cyst [1]. Children diagnosed with DWM frequently present with hydrocephalus, and the majority of patients require a shunt operation [2, 3]. Although a number of abnormalities involving various organ systems are associated with DWM, the co-occurrence of DWM and syringomyelia is extremely rare, and only a few cases have been reported in the literature [3–7]. Here, we report a patient with DWM and concomitant syringomyelia that presented with progressive scoliosis and fecal incontinence. The clinical course and surgical outcomes are described. The possible mechanisms of syrinx formation will be discussed.

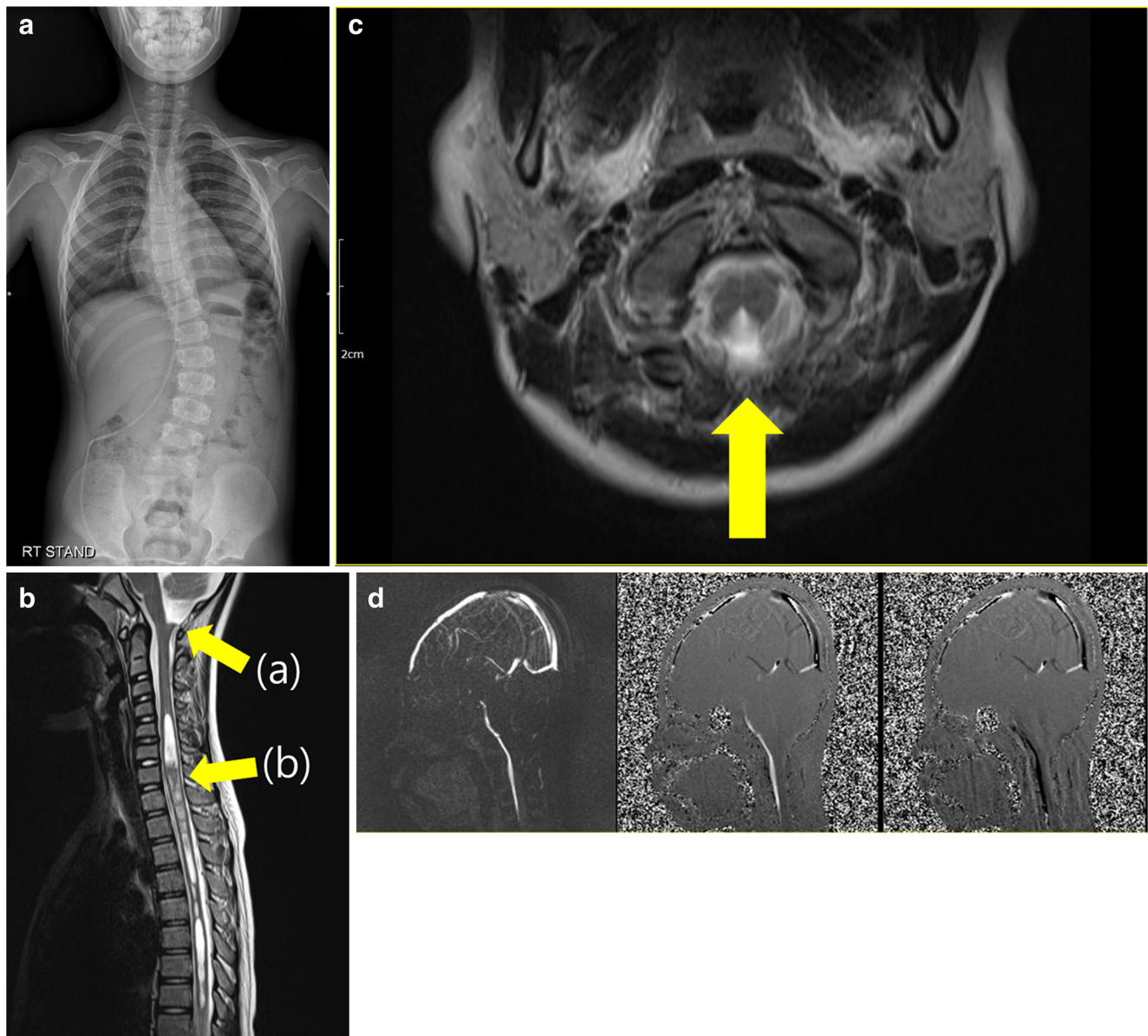
## Case report

### Clinical presentation

A 10-year-old boy previously diagnosed with DWM presented with new onset fecal incontinence that had persisted for several weeks. Additionally, he presented with a tilting posture, and a plain radiograph of his spine demonstrated scoliosis (Fig. 1a). Physical examination revealed no other neurological deficits.

✉ Kyu-Chang Wang  
kcwang@snu.ac.kr

<sup>1</sup> Division of Pediatric Neurosurgery, Department of Neurosurgery, Seoul National University Children's Hospital and Seoul National University College of Medicine, 101 Daehak-ro, Jongno-gu, Seoul 110-744, Republic of Korea



**Fig. 1** **a** A whole spine radiograph, A-P view, showing scoliosis of the thoracolumbar spine. **b** T2-weighted sagittal spine MRI showing the membranous structure (*a*) tethering the lower brainstem and concomitant syringomyelia (*b*) along the cervicothoracic spinal cord. **c** T2-weighted axial MRI demonstrating the tethered point of the lower brainstem. **d** CSF flow study with phase-contrast cine MRI. Brain

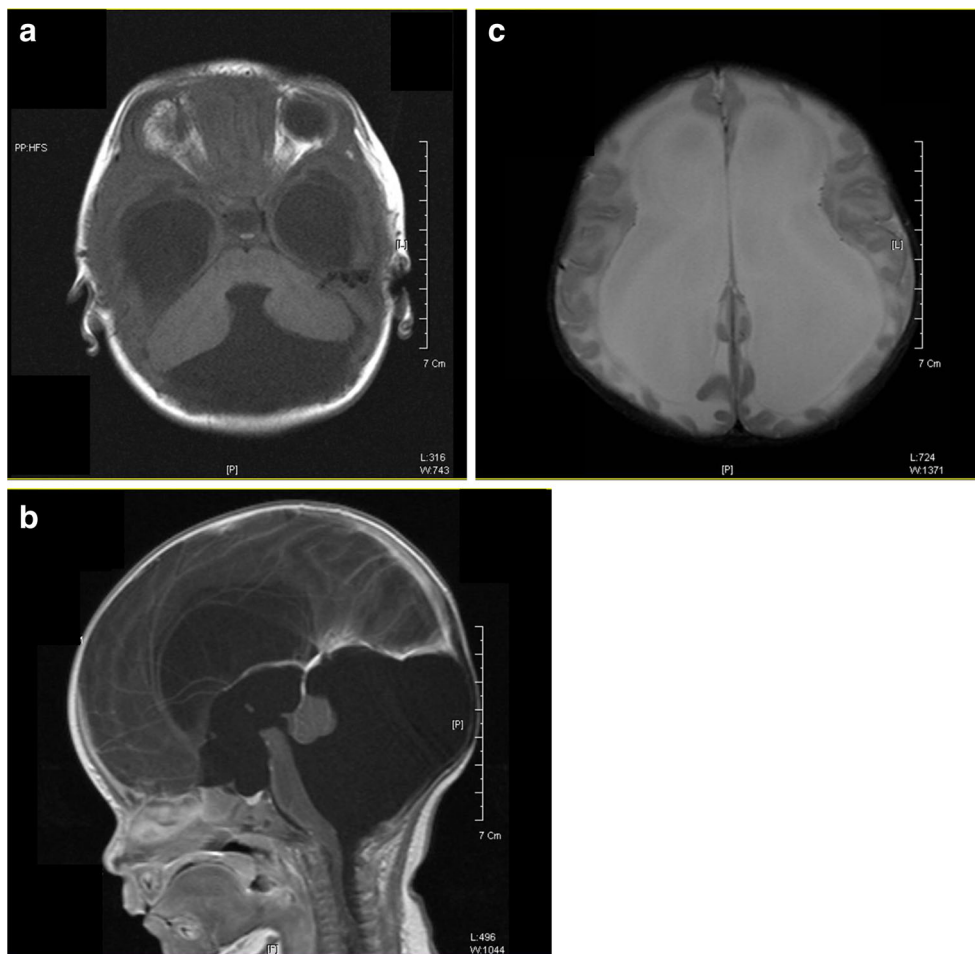
pulsation results in caudal and cephalic flow of CSF across the foramen magnum. Reversal in the direction of flow appears as alternating light and dark signals. Note the intact CSF flow in front of the cervicomedullary junction and the complete absence of CSF flow behind the cervicomedullary junction

### Past medical history

At 1 month of age, the patient was diagnosed with DWM at another hospital after the evaluation of macrocephaly using brain MRI (Fig. 2). He underwent two surgical interventions at the same hospital. The patient exhibited severe hydrocephalus during the neonatal period and underwent the surgical placement of a cystoperitoneal (CP) shunt. At the age of 5 months, the patient required the additional insertion of a ventriculoperitoneal (VP) shunt, and the

ventricular catheter was connected to the existing CP shunt via Y connection. Over the next 7 years, there was no need for shunt revision, and the patient was doing well. When he turned 7 years old, the patient presented with chronic headaches and was referred to our clinic for further evaluation. Suspicious of a shunt malfunction, we performed a shunt revision. The malfunctioning VP shunt was removed, and a new one was inserted. The revision was complicated by intraventricular hemorrhage in both lateral ventricles which was managed conservatively (Fig. 3). After the revision,

**Fig. 2** **a** Precontrast axial and **b** postcontrast sagittal T1-weighted brain MR images showing a posterior fossa cyst with vermian hypoplasia and high insertion of tentorium and an anterior displacement of the brainstem with hydrocephalus. **c** T2-weighted axial brain MRI showing ventriculomegaly before the insertion of a VP shunt



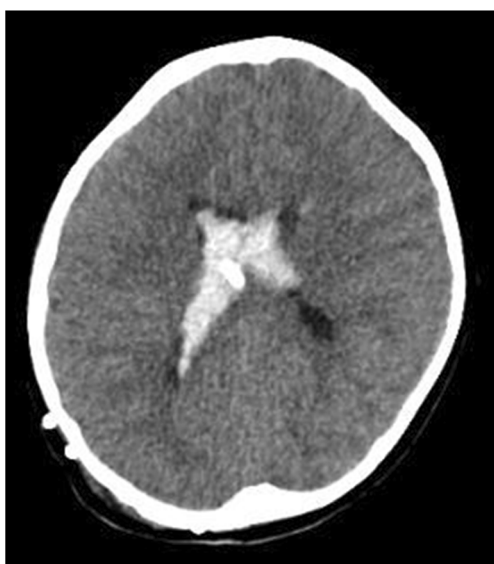
there were no shunt-related complications, and the patient was asymptomatic until his current visit to our clinic.

**Radiological findings**

To evaluate the cause of the fecal incontinence and scoliosis, a spine MRI was conducted. The spine MRI revealed tethering of the lower brainstem by a thickened membranous structure and the development of a syringomyelia along the cervicothoracic spinal cord (Fig. 1b, c). The tethering was not evident on the former radiologic exams, including the MRI image obtained at the initial diagnosis (Fig. 2). To evaluate for a possible CSF flow obstruction around the abnormally thickened membrane, phase-contrast cine MRI was conducted [8]. The cine MR images showed decreased CSF flow in the cerebral aqueduct and the dorsal aspect of the medulla and the high cervical spinal cord (Fig. 1d).

**Operation**

During the current visit, because there were no signs of increased intracranial pressure or changes in the ventricular size shown in the serial images, the possibility of a shunt malfunction was excluded. Scoliosis and fecal incontinence are common characteristic features of syringomyelia; therefore, we hypothesized that the patient’s clinical symptoms could be

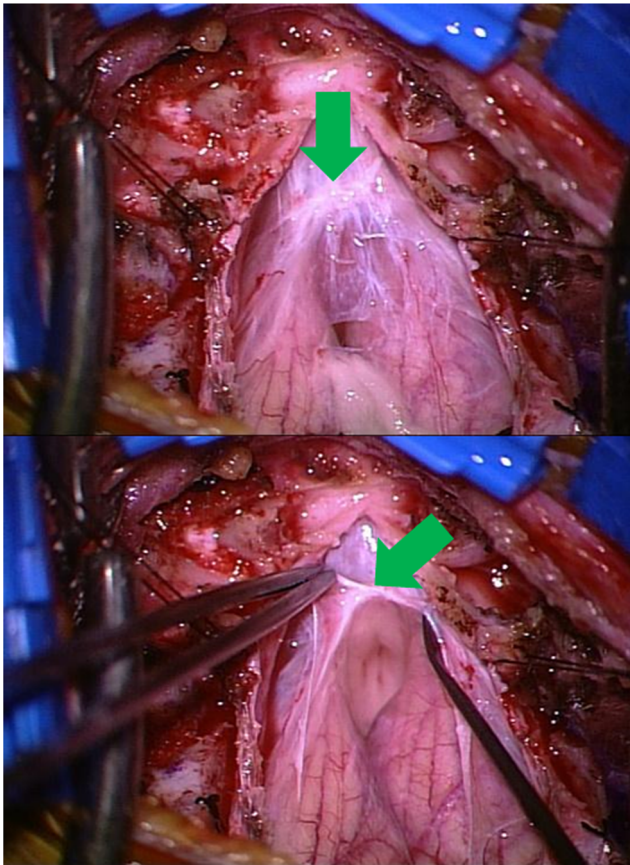


**Fig. 3** Computed tomography showing postoperative intraventricular hemorrhage

caused by syringomyelia. Under the clinical suspicion that syringomyelia was due to the thickened membranous structure around the brainstem, a suboccipital craniectomy and membrane excision were performed. Intraoperatively, as seen in the imaging studies, a thickened membranous structure was found tethering the brainstem (Fig. 4). The gross finding of the lesion was a thickened arachnoid membrane containing many adhesions to the dura mater and the medulla oblongata. There was a tight band-like adhesion which was thought to be the tethering point observed on the MR image. Careful dissection and microscopic resection of the adhesions were performed. Histological findings indicated that the lesion was composed of fibrous tissue with collagen deposition and degenerative changes. This intradural extramedullary band of arachnoid tissue is referred to as an “arachnoid web.”

### Postoperative course

Postoperatively, the patient made a good recovery, and there were no complications. The patient no longer experienced fecal incontinence. Scoliosis was also improved according to the 1-month postoperative follow-up spine radiographs (Fig. 5a). A 6-month follow-up MR image showed that



**Fig. 4** The arrow indicates the thickened arachnoid membrane tethering the lower brainstem

syringomyelia had decreased in size and revealed the resected state of the previously existing arachnoid web around the cervicomedullary junction (Fig. 5b, c).

### Discussion

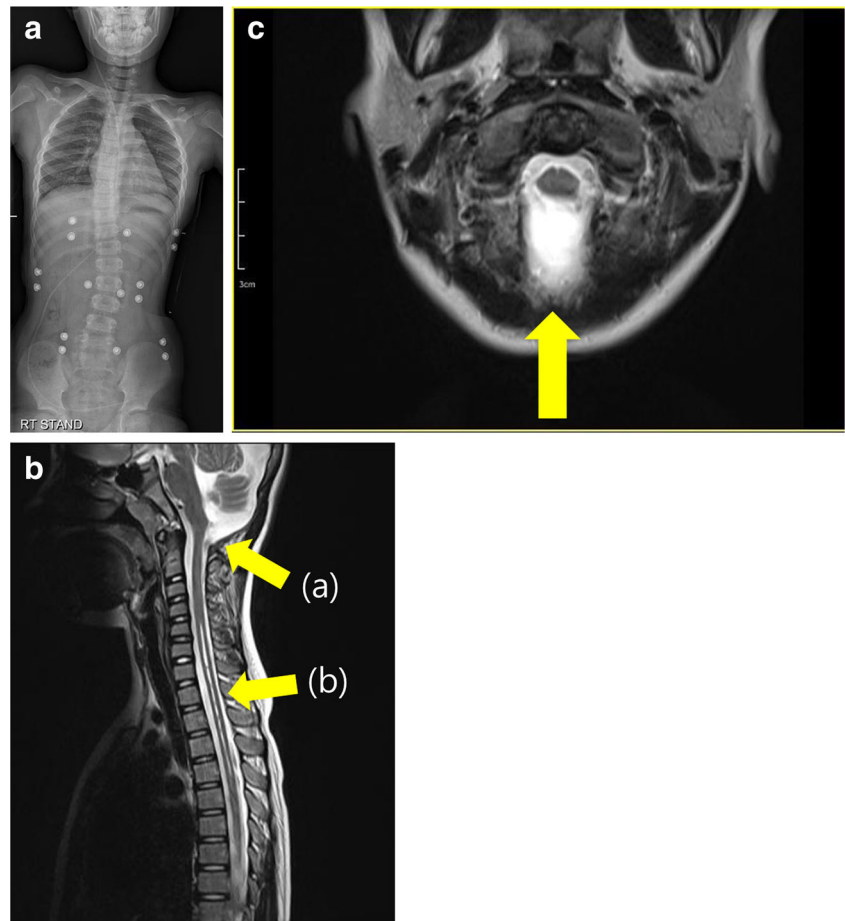
We report a case of a young child with DWM in whom syringomyelia developed due to an arachnoid web that was tethering the brainstem and blocking the flow of CSF. We performed a suboccipital craniectomy and excised the arachnoid web to release the tethered brainstem and normalize CSF flow. Clinical and radiological improvement was observed after we performed surgical excision of the arachnoid web. Our case exhibited a different mechanism of syrinx formation than previously reported cases, in that there was no sign of herniation of the lower pole of the cyst and no uncontrolled hydrocephalus because he was a shunted DWM patient [9, 10].

There are two suggested mechanisms of syringomyelia formation in patients with a DWM in the literature. The first one is the herniation of the lower pole of the fourth ventricular cyst, which mimics the Chiari-like mechanism [9]. The second proposed mechanism is the evolution of the central canal into a “fifth ventricle” when uncontrolled hydrocephalus is present due to an obstruction distal to the fourth ventricle [10]. However, our patient had a shunt, and no herniated cyst or sign of occipital foramen magnum impaction was detected. Furthermore, because the size of the ventricles and the Dandy-Walker cyst in our patient were not enlarged, the cystoventriculoperitoneal shunt was considered functional, and hydrocephalus was well controlled. Therefore, our case is not well explained by the previously suggested mechanisms of syrinx formation. On the other hand, the obstruction of CSF flow has been identified as a major predisposing factor for syrinx formation [11–13]. Because our patient exhibited CSF flow obstruction around the arachnoid web, according to the results of phase-contrast cine MRI, we postulate that syringomyelia in our patient was caused by disturbed CSF flow around the arachnoid web.

We believe that the arachnoid web also exhibited a tethering effect on the cervical spinal cord, as described in another study [14]. However, the association between high cervical or brainstem tethering and syringomyelia is not well established. An experimental study suggests that high cervical level tethering could also cause syringomyelia. Klekamp et al. investigated the effects of focal arachnoid scarring in the cervical spinal canal of animal models and its association with syringomyelia [15]. In the experimental group, the authors artificially induced a focal arachnoid scar at the high cervical level using placement of a kaolin-soaked fibrin sponge. They demonstrated that arachnoid scarring at the cervical level could induce the initial development of syringomyelia below the area of scarring.

We then considered whether arachnoid web formation in this patient was a manifestation of DWM. If the membrane

**Fig. 5** **a** The 1-month postoperative whole spine radiograph showing improvement of scoliosis. **b** T2-weighted sagittal MR images obtained 6 months postoperatively showing the resected state of the previously existing arachnoid web around the cervicomedullary junction (*a*) and decreased syringomyelia size (*b*). **c** A T2-weighted axial MR image obtained at the same time showing the untethered state of the lower brainstem



was a part of the Dandy-Walker cyst wall, the pathology findings would reveal two distinct layers: an inner layer of ependymal-glia tissue and an outer pia-arachnoid layer. However, the results of pathologic examination in our patient showed only fibrous tissue and no ependymal or glial component. Therefore, we concluded that the thickened arachnoid membrane was not a specific feature of DWM. Rather, we believed that it was a complication of the previous intraventricular hemorrhage that occurred during the patient's shunt revision. The intraventricular hemorrhage of the fourth ventricle could have flowed into the cisterna magna resulting in regional arachnoiditis and adhesions, which could then be converted into an arachnoid web [16].

Therefore, we concluded that syringomyelia formation in our patient is caused by an arachnoid web complicated by the postsurgical intraventricular hemorrhage and is not directly related to DWM. However, because the syringomyelia formation as a complication of the arachnoid web and intraventricular hemorrhage is not a common situation, we initially thought that there would be some correlation between syringomyelia formation and DWM in our case. We took many considerations about any possible mechanisms of syringomyelia formation which might directly be related to DWM. Now, we want to emphasize the importance of thorough evaluation

to find out unexpected pathophysiological mechanisms in a specific clinical setting.

Surgical strategies used for the treatment of syringomyelia associated with DWM include posterior fossa decompression, plugging the obex with muscle, VP shunt revision, CP shunt revision, and placement of a syringoperitoneal shunt [4]. If the posterior fossa cyst impaction into the foramen magnum is responsible for syringomyelia formation in patients with a DWM, a CP or VP shunt could be a first choice for treatment [17]. However, as in our case, if a functioning shunt is not adequate for the resolution of syringomyelia, posterior fossa decompression would be a reasonable second choice [9]. Moreover, when an abnormal structure is demonstrated by radiologic exam, the correction of the abnormality to restore normal anatomy is a reasonable treatment option.

## Conclusion

This unusual case shows that the arachnoid web can alter CSF flow dynamics and can result in the formation of syringomyelia. After thorough evaluation of the pathophysiological mechanisms of syringomyelia in a patient, the proper surgical procedure should be undertaken when indicated. Craniectomy

with membrane excision could be an effective treatment option depending on the specific clinical situation.

**Acknowledgments** This research was supported by Basic Science Research Program through the National Research Foundation of Korea (NRF) funded by the Ministry of Science, ICT, and Future Planning (2015R1A2A1A15055781).

#### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

#### References

1. Brodal A, Hauglie-Hanssen E (1959) Congenital hydrocephalus with defective development of the cerebellar vermis (Dandy-Walker syndrome): clinical and anatomical findings in two cases with particular reference to the so-called atresia of the foramina of Magendie and Luschka. *J Neurol Neurosurg Psychiatry* 22:99–108
2. Bindal AK, Storrs BB, McLone DG (1990) Management of the Dandy-Walker syndrome. *Pediatr Neurosurg* 16:163–169
3. Hirsch JF, Pierre-Kahn A, Renier D, Sainte-Rose C, Hoppe-Hirsch E (1984) The Dandy-Walker malformation. A review of 40 cases. *J Neurosurg* 61:515–522
4. Richter EO, Pincus DW (2006) Development of syringohydromyelia associated with Dandy-Walker malformation: treatment with cystoperitoneal shunt placement. Case report. *J Neurosurg* 104:206–209
5. Gardner WJ, Abdullah AF, McCormack LJ (1957) The varying expressions of embryonal atresia of the fourth ventricle in adults: Arnold-Chiari malformation, Dandy-Walker syndrome, arachnoid cyst of the cerebellum, and syringomyelia. *J Neurosurg* 14:591–605
6. Kasliwal MK, Suri A, Sharma BS (2008) Dandy Walker malformation associated with syringomyelia. *Clin Neurol Neurosurg* 110:317–319
7. Hammond CJ, Chitnavis B, Penny CC, Strong AJ (2002) Dandy-Walker complex and syringomyelia in an adult: case report and discussion. *Neurosurgery* 50:191–194
8. Quencer RM (1992) Intracranial CSF flow in pediatric hydrocephalus: evaluation with cine-MR imaging. *Am J Neuroradiol* 13:601–608
9. Cinalli G, Vinikoff L, Zerah M, Renier D, Pierre-Kahn A (1997) Dandy-Walker malformation associated with syringomyelia. Case illustration. *J Neurosurg* 86:571
10. Milhorat TH, Capocelli AL Jr, Anzil AP, Kotzen RM, Milhorat RH (1995) Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. *J Neurosurg* 82:802–812
11. Milhorat TH (2000) Classification of syringomyelia. *Neurosurg Focus* 8:E1
12. Klekamp J (2002) The pathophysiology of syringomyelia—historical overview and current concept. *Acta Neurochir* 144:649–664
13. Greitz D (2006) Unraveling the riddle of syringomyelia. *Neurosurg Rev* 29:251–263 discussion 264
14. Klekamp J, Batzdorf U, Samii M, Bothe HW (1997) Treatment of syringomyelia associated with arachnoid scarring caused by arachnoiditis or trauma. *J Neurosurg* 86:233–240
15. Klekamp J, Volkel K, Bartels CJ, Samii M (2001) Disturbances of cerebrospinal fluid flow attributable to arachnoid scarring cause interstitial edema of the cat spinal cord. *Neurosurgery* 48:174–185 discussion 185–186
16. Caplan LR, Norohna AB, Amico LL (1990) Syringomyelia and arachnoiditis. *J Neurol Neurosurg Psychiatry* 53:106–113
17. Tekkok IH, Ventureyra EC (1997) Hydrosyringomyelia associated with Dandy-Walker malformation—is it really rare or undiagnosed? *Eur J Paediatr Neurol* 1:49–51