

Dandy–Walker syndrome associated with syringomyelia in an adult: a case report and literature review

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Abstract

Dandy–Walker syndrome associated with syringomyelia is a rare condition, with few reports of adult cases. We describe an adult case of Dandy–Walker syndrome with concomitant syringomyelia. A 33-year-old man presented with a 3-month history of walking instability, numbness in the hands, memory deterioration, and urinary incontinence. A physical examination showed a positive Romberg sign. Brain computed tomography and magnetic resonance imaging showed hydrocephalus, a cyst in the posterior fossa, absence of the cerebellar vermis, hypoplasia of the corpus callosum and cerebella, and syringomyelia. All of these symptoms were consistent with the diagnosis of Dandy–Walker syndrome. Surgery involving arachnoid adhesiolysis and endoscopic third ventriculostomy was performed. At the 6-month follow-up, the symptoms were completely relieved. Magnetic resonance imaging showed that syringomyelia was greatly reduced and the hydrocephalus remained unchanged. Dandy–Walker syndrome with concomitant syringomyelia in adults is exceedingly rare. Early diagnosis and appropriate surgical treatment of this condition should be highlighted. Combined arachnoid adhesiolysis and endoscopic third ventriculostomy may be an effective approach.

Keywords

Dandy–Walker syndrome, syringomyelia, adult, surgical treatment, endoscopic third ventriculostomy, hydrocephalus

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Introduction

Dandy–Walker syndrome (DWS) is an infrequent congenital brain malformation with an incidence of 1/25,000 to 1/35,000 births.¹ This condition usually presents in pediatric patients and adult presentation of DWS is rare. Additionally, formation of syringomyelia in patients with DWS is extremely rare. Only 23 cases of DWS have been previously reported in the literature, among which three were adult-onset.^{2–4} No studies have provided a comprehensive overview regarding the pathogenesis of this coincidence, and the treatment and prognosis of this entity have yet to be well investigated.

We describe an adult case of DWS and concomitant syringomyelia. Moreover, the relevant literature was reviewed, and the possible pathogenesis, clinicoradiological characteristics, and surgical strategies of DWS with syringomyelia are discussed.

Case report

A 33-year-old man presented with a 3-month history of walking unsteadily, numbness in the hands, memory deterioration, and urinary incontinence. These symptoms were aggravated 1 week before admission. During the patient's childhood, he had a stammer and his athletic ability was poor, while his intelligence was almost normal. A physical examination showed walking unsteadily, numbness in the hands, and a positive Romberg sign. Brain computed tomography and magnetic resonance imaging (MRI) showed dilation of the lateral and third ventricles and a cyst in the posterior fossa, which was adjacent to the fourth ventricle. Furthermore, these techniques showed absence of the cerebellar vermis, hypoplasia of the corpus callosum and cerebella, and syringomyelia in the cervicothoracic spinal cord (Figure 1). A diagnosis of DWS with obstructive hydrocephalus

and concomitant syringomyelia was made. There was partial obstruction in the midbrain aqueduct and in the fourth ventricular outflow tract that leads to dilation of the supratentorial ventricles. Therefore, surgery, including first-stage arachnoid adhesiolysis and second-stage endoscopic third ventriculostomy (ETV), was scheduled. During arachnoid adhesiolysis, we found an arachnoid cyst in the posterior fossa. The local arachnoid membrane was incised and adhered to the fourth ventricular outflow. This arachnoid membrane adhesion was released and the cyst was resected, and the fourth ventricular outflow was exposed. Endoscopically, the fourth ventricular outflow tract and choroid plexus were visible in the upward view (30°), and a fissure in the central spinal cord was visible in the downward view (30°). EVT was performed 2 weeks after arachnoid adhesiolysis. Intraoperatively, we observed that the third ventricular floor was flat and thin, and a 10-mm orificium fistulae was made (Figure 2). The postoperative course was uneventful. The numbness in the hands and walking instability were greatly improved a few days later and the urinary incontinence disappeared. After a 6-month follow-up, the symptoms were completely relieved, and a neurological examination showed that the Romberg test was negative. MRI showed that the syringomyelia was considerably reduced in size and the hydrocephalus remained unchanged (Figure 1).

The study protocol was approved by the Ethics Committee of the First Affiliated Hospital of Soochow University. The patient gave consent for publication of this case report.

Discussion

DWS, also known as Dandy–Walker malformation or Dandy–Walker complex, was originally described by Sutton⁵ in 1887 in a

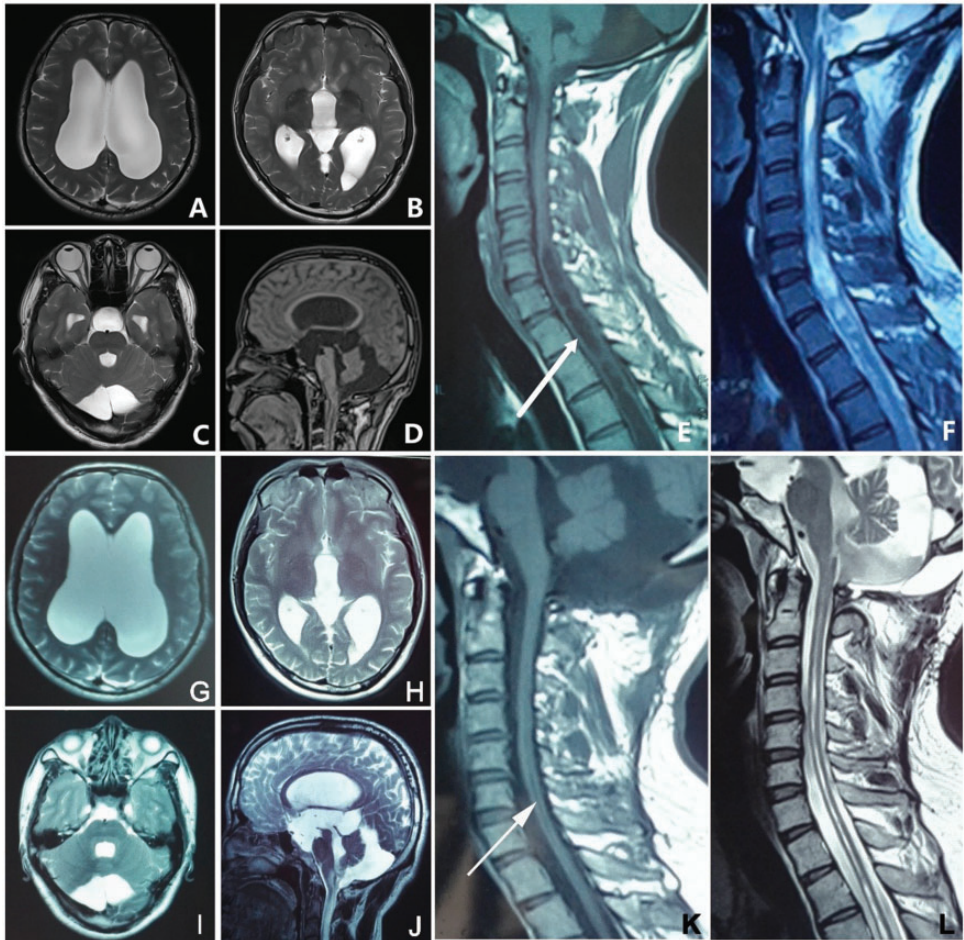


Figure 1. Preoperative and follow-up magnetic resonance imaging of the patient.

Axial brain magnetic resonance T2-weighted imaging shows that the lateral ventricle (A) and the third ventricle (B) are dilated, and the cerebellar vermis is absent (C). Sagittal brain magnetic resonance T1-weighted imaging shows a cyst in the posterior fossa and hypoplasia of the corpus callosum and cerebella, and the posterior fossa cyst is adjacent to the fourth ventricle (D). Sagittal spinal magnetic resonance T1-weighted (E) and T2-weighted (F) imaging show syringomyelia in the cervicothoracic spinal cord. Axial (G–I) and sagittal (J) brain magnetic resonance T2-weighted imaging shows ventricular dilation and the posterior fossa cyst shows no major changes. Sagittal spinal magnetic resonance T1-weighted (K) and T2-weighted (L) imaging show that syringomyelia is greatly reduced in size.

case of hydrocephalus, posterior fossa cyst, and hypoplasia of the cerebellar vermis. This triad entity was then confirmed by Dandy and Blankfan in 1914.⁶ In 1942, Taggart and Walker reported three cases of congenital atresia of foramina of

Luschka and Magendie.⁷ They proposed a hypothesis that this condition may be due to damage of the fourth ventricle and cerebellar vermis hypoplasia during embryonic development. The nomenclature “Dandy–Walker syndrome” was first introduced by

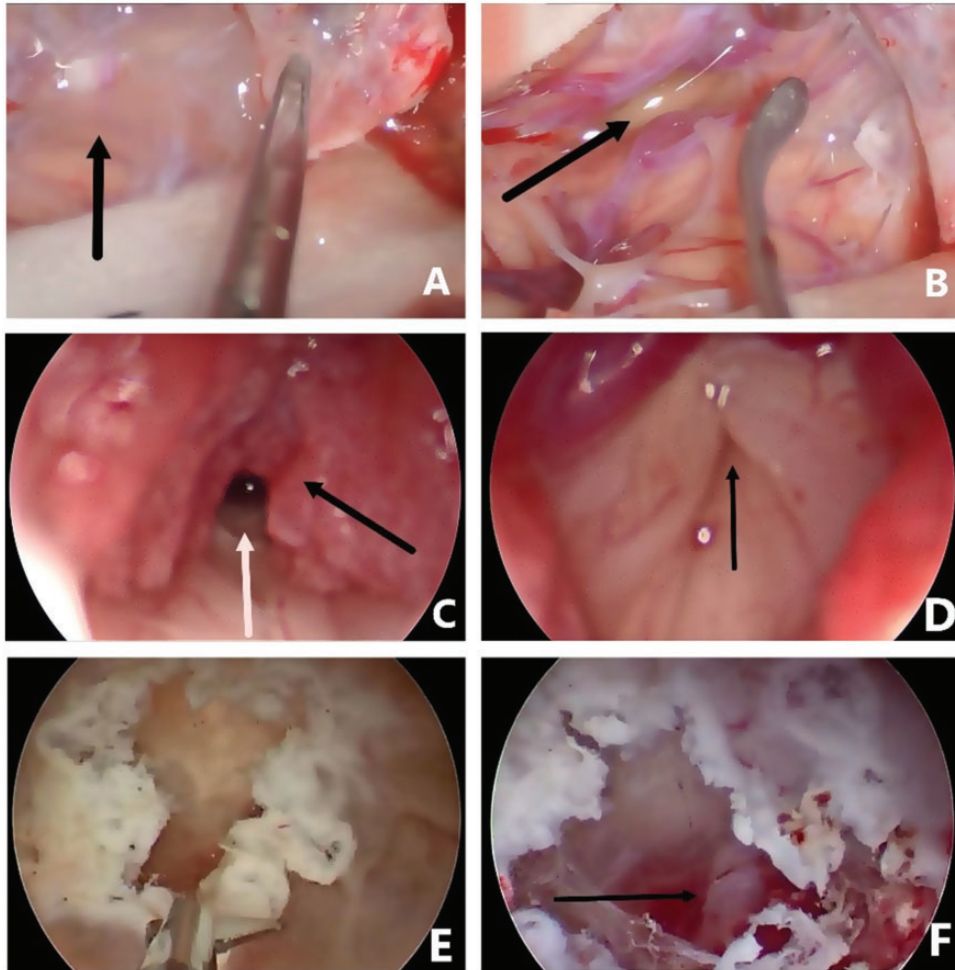


Figure 2. Intraoperative images.

(A) Once the dura mater was opened, an arachnoid cyst was visible (black arrow). The local arachnoid membrane was incassated and adhered to the fourth ventricular outflow. (B) After arachnoid membrane adhesion was released and the cyst was resected, the fourth ventricular outflow was exposed. (C) Endoscopically, the fourth ventricular outflow tract (white arrow) and choroid plexus (black arrow) can be seen in the upward view. (D) In the downward view, a fissure in the central spinal cord is visible. (E) A 10-mm orificium fistulae was made in the third ventricular floor. (F) After Lilliequist's membrane was opened, the basilar artery was exposed (black arrow).

Benda in 1954.¹ Currently, DWS is considered to be the result of maldevelopment in the region of the fourth ventricle, but is not limited to foraminal atresia.

The definitive mechanisms of DWS are not entirely clear. DWS can be characterized

as an independent deformity or associated with other congenital disorders, such as neural tube defects, congenital heart disease, cleft lip/palate, and Hirschsprung's disease. Some scholars have also proposed that DWS may be associated with genetic deficiencies,

Table 1. Current widely used surgical strategies for Dandy–Walker syndrome.

Aqueduct of the midbrain	The fourth ventricle-subarachnoid space	Surgical strategy
Obstructed	Unobstructed	Ventriculoperitoneal shunting
Obstructed	Obstructed	Ventriculoperitoneal shunting plus cystoperitoneal shunting
Unobstructed	Unobstructed or obstructed	Ventriculoperitoneal shunting or cystoperitoneal shunting

and the potential locations include 9q22-9qter and 13q22-q33.⁸

The diagnosis of DWS mainly depends on neuroimaging examinations. Prenatal ultrasound is an accurate and reliable approach for identifying DWS during gestation. Currently, MRI is the first choice of diagnosis and the typical characteristics include the following: (1) dilated fourth ventricle or huge posterior fossa cyst communicating with the fourth ventricle, and the cyst showing isointensity to cerebrospinal fluid; (2) cerebellar hypoplasia or/and absence of the cerebellar vermis; (3) enlargement of the posterior fossa with upward displacement of the lateral sinuses, sinus confluents, and tentorium; (4) with or without supratentorial hydrocephalus; (5) with or without agenesis of corpus callosum; and (6) with or without other intracranial malformations.

The clinical manifestations of DWS are non-specific, and are related to cerebellar structural defects and intracranial hypertension caused by hydrocephalus and an enlarged posterior fossa cyst. Generally, the onset of clinical symptoms of DWS occurs in infants (<2 years old), manifesting as macrocrania, headache, vomiting, cranial nerve palsy, seizures, pyramidal syndrome, and hemiparesis.¹ Adult clinical manifestations of DWS are more diverse. In addition to hydrocephalus and cerebellar signs and symptoms, there are many rare symptoms, such as diplopia, myasthenia gravis, sensorineural deafness, and

neurocutaneous melanosis. Some cases have shown that DWS might first show symptoms in the latter half of life or even be asymptomatic throughout life.^{2-4,9-11} DWS with associated syringomyelia has been described in 23 isolated case reports.^{2-4,9-11} The majority of these patients were diagnosed in an autopsy or in an incidental imaging examination.

Surgical treatment is the mainstream treatment of DWS. Nevertheless, because of low morbidity of DWS, surgical indications and operative procedures are still controversial. Cyst fenestration and posterior fossa decompression were the main choices in earlier years, but these approaches could not relieve obstruction of cerebrospinal fluid circulation. Therefore, cystoperitoneal shunting or ventriculoperitoneal shunting have been widely applied. Some authors recommended that the surgical approach should be scheduled according to the extent of obstruction of the aqueduct and subarachnoid space (Table 1).¹²

With advances in endoscopic techniques, ETV is a new option for treating DWS. Mohanty et al.¹³ retrospectively reviewed 72 pediatric patients who underwent surgical treatment for DWS. In their study, 21 patients were treated with ventriculoperitoneal shunting, 24 with cystoperitoneal shunting, and three with combined ventriculoperitoneal and cystoperitoneal shunting. A total of 21 patients underwent endoscopic third ventriculostomy, and during the follow-up period, five of them

required an additional ventriculoperitoneal shunt insertion.¹³ In 2011, Warf et al.¹⁴ conducted a retrospective review of 45 patients with DWS or DWS-associated hydrocephalus who underwent ETV and choroid plexus cauterization, with a total success rate of 74%. These authors strongly recommended endoscopic surgery as the primary management for treating DWS. ETV has significant advantages, as follows: (a) this procedure does not depend on implanted shunt devices; (b) it can allow communication of the ventricle with the subarachnoid space, restoring normal physiological mechanisms of cerebrospinal fluid circulation; and (c) ventriculoperitoneal or cystoperitoneal shunting can be a remedial measure when endoscopic surgery fails.^{12–14}

DWS associated with syringomyelia is an extremely rare entity. The pathogenesis of this coincidence is unclear. Previous studies have postulated two hypothetical theories: (1) cyst herniation through the foramen magnum may alter cerebrospinal fluid flow dynamics, causing syringomyelia; and (2) DWS may be in direct continuity with the syrinx cavity. In our case, we attempted to use arachnoid adhesiolysis and ETV for treatment. The surgical outcome was satisfactory and syringomyelia was greatly reduced in size. Therefore, combined arachnoid adhesiolysis and ETV was effective.

Conclusions

DWS with concomitant syringomyelia in adults is exceedingly rare. Combined arachnoid adhesiolysis and ETV may be an effective approach for alleviation of symptoms and syringomyelia. The surgical outcome of this condition can be favorable.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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References

1. Hirsch JF, Pierre-Kahn A, Renier D, et al. The Dandy-Walker malformation. A review of 40 cases. *J Neurosurg* 1984; 61: 515–522.
2. Erdal M, Plikcioglu AC, Bikmaz K, et al. Dandy-Walker complex and syringomyelia in an adult: case report and discussion. *Neurosurgery* 2003; 52: 1504–1505.
3. Owler BK, Halmagyi GJ, Besser M. Syringomyelia with Chiari malformation; 3 unusual cases with implications for pathogenesis. *Acta Neurochirurgica* 2004; 146: 1137–1143.
4. Hammond, CJ. Dandy-Walker complex and syringomyelia in an adult: case report and discussion: in reply. *Neurosurgery* 2003; 50: 191–194.
5. Sutton JB. The lateral recesses of the fourth ventricle; their relation to certain cysts and tumours of the cerebellum, and to occipital meningocele. *Brain* 1886; 9(3): 352–361.
6. Dandy W. Internal hydrocephalus. An experimental, clinical and pathological study. *Am J Dis Child* 1914; 8: 406–482.
7. Taggart JK, Walker AE. Congenital atresia of the Foramens of Luschka and Magendie. *Arch Neurol Psychiatry* 1942; 48: 583–612.
8. von Kaisenberg CS, Caliebe A, Krams M, et al. Absence of 9q22-9qter in trisomy 9 does not prevent a Dandy-Walker phenotype. *Am J Med Genet* 2000; 95: 425–428.
9. Richter EO, Pincus DW. Development of syringohydromyelia associated with Dandy-Walker malformation: treatment with cystoperitoneal shunt placement. Case report. *J Neurosurg* 2006; 104(3 Suppl): 206–209.
10. Kasliwal MK, Suri A, Sharma BS. Dandy Walker malformation associated with syringomyelia. *Clin Neurol Neurosurg* 2008; 110: 317–319.
11. Lee HC, Choi JW, Lee JY, et al. Syringomyelia caused by an arachnoid web in a patient with shunted Dandy-Walker malformation. *Childs Nerv Syst* 2107; 33: 1–6.

12. Ozgen T and Acikgoz B. CSF dynamics in Dandy-Walker syndrome. *Acta Neurochir (Wien)* 1990; 104: 54–58.
13. Mohanty A, Biswas A, Satish S, et al. Treatment options for Dandy-Walker malformation. *J Neurosurg* 2006; 105: 348–356.
14. Warf BC, Dewan M and Mugamba J. Management of Dandy-Walker complex-associated infant hydrocephalus by combined endoscopic third ventriculostomy and choroid plexus cauterization. *J Neurosurg Pediatr* 2011; 8: 377–383.