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CASE REPORT

Dandy-Walker malformation and syringomyelia: a rare association

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Abstract

Purpose Dandy-Walker malformation is a rare condition due to imperforation of the Blake's pouch during intrauterine brain development, usually leading to early severe hydrocephalus. The association with holocord syringomyelia is rare, and from the Gardner's first report in 1957, only 23 cases have been described, mostly from autopsy series and pre-MRI period. Besides a worsening of clinical picture, its occurrence generates some concern about the best surgical treatment that varies widely among the literature reports.

Methods An 11-year-old girl with Dandy-Walker malformation presented with a holocord syrinx due to the herniation of the lower pole of the posterior fossa cyst through the foramen magnum.

Results After an unsuccessful shunt revision, she underwent a cystoperitoneal shunt with regression of the syrinx and of neurological symptoms at the 12-month follow-up.

Conclusions Previous literature about pathogenesis, treatment, and follow-up is discussed and summarized.

Keywords Dandy-Walker malformation · Syringomyelia · Syrinx cyst

Abbreviations

DWM	Dandy-Walker malformation
MRI	Magnetic resonance imaging
CT	Computed tomography
VPS	Ventriculoperitoneal shunt
CPS	Cystoperitoneal shunt
CSF	Cerebospinal fluid
IVH	Intraventricular hemorrhage
C1M-LM	Chiari 1 malformation-like mechanism

Introduction

Dandy-Walker malformation (DWM) is a rare congenital malformation involving the posterior fossa, affecting 1/25000–30,000 live births [3, 4, 20]. Diagnosis is based on classical radiological features, which include (1) upward and backward rotation of hypoplastic cerebellar vermis and (2) posterior fossa cyst [4]. At the initial diagnosis, 80% of the patients present hydrocephalus, which represents the most common complication of this malformation [20]. The co-existence of DWM and syringomyelia is rare, and only 23 cases have been reported so far [1, 5–11, 13–19, 21–23]. According to the available literature, pathogenic hypotheses, best surgical approach, and outcome remain poorly defined.

We report on a girl with DWM and concomitant holocord syringomyelia that presented with severe cervicalgia. The clinical course, surgical approach, and outcome are described, and possible mechanisms of syrinx formation are discussed.

Case presentation

A 2-month-old girl underwent head CT for progressive macrocrania without other symptoms. Neuroimaging diagnosed a DWM with mild supratentorial ventricular dilation that was clinically and radiologically monitored. At the age of 2, a VPS was placed for symptomatic hydrocephalus with good postoperative outcome. In 2013, an MRI was performed to investigate a growth deficiency associated to low levels of serum somatomedine C: neuroimaging showed a holocord syrinx associated with herniation of the posterior fossa cyst through the foramen magnum (Fig. 1a, b).

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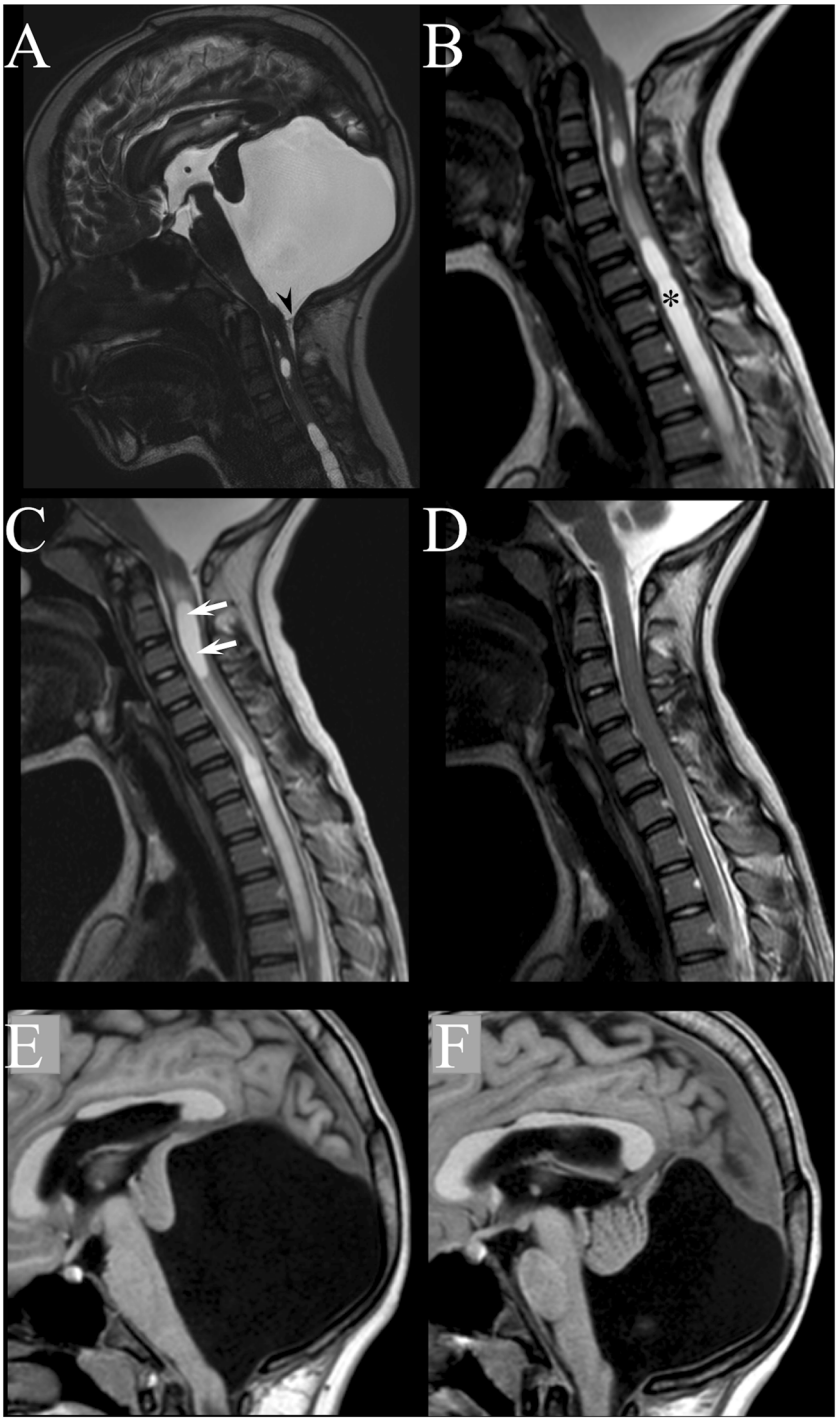

Fig. 1 MRI exams in our DWM girl. **a** Sagittal T2-weighted image of the brain showing the upward and backward rotation of the hypoplastic cerebellar vermis; a large cyst occupies the posterior cranial fossa and its wall is protruding in the foramen magnum (black arrowhead). **b**–**d** Midsagittal T2-weighted images of the cervical spine showing the syrinx at diagnosis (**b**; *), its cervical progression after ventriculo-peritoneal shunt revision (**c**; white arrows) and its complete regression after cyst-peritoneal shunting (**d**). **e**, **f** Midsagittal T1-weighted images of the posterior cranial fossa disclosing the infratentorial cyst after ventriculo-peritoneal shunt revision (**e**) and its detension after cyst-peritoneal shunting (**f**)

Surgery

Despite the absence of supratentorial hydrocephalus progression, a shunt malfunction was considered and the VPS was revised. During surgery, a distal malfunction of the system was found and both distal catheter and the programmable valve were changed. After few weeks, she started to complain about a neck pain, which progressively impaired daily activities. The follow-up MRI showed a reduction in size of the medio-dorsal part of the syrinx but an increase of the cranial portion of its cervical part (Fig. 1c), prompting for a CPS connected to VPS beyond the programmable valve with a Y-connector, deferring a more invasive surgery such as a suboccipital decompression with C1 laminectomy and cyst decompression in the case of failure.

Follow-up

Postoperatively, the girl made a good recovery and there were no complications related to the surgery. The neck pain progressively disappeared. The follow-up MRI showed the reduction in size of the syrinx that completely regressed 1 year after surgery (Fig. 1d). After a 4-year follow-up, she is neurologically intact without recurrence of the syrinx.

Literature review

The association between DWM and syringomyelia has been reported in 23 subjects [1, 5–11, 13–19, 21–23]. Among them, nine were reported in autopsy series (Table 1) and 14 in clinical series (Table 2). Only the latter were considered in our literature review, whenever there was direct causal relationship between DWM and syrinx. The syrinx was cervicothoracic in six [1, 5, 9, 14, 19, 21] and cervical in two cases [6, 18]; in four, the syrinx was extensive (at least 10 myelomers) [5, 14, 19, 21]. A direct communication with the ventricular system was found in five cases [6–8], while a CIM-LM was observed in six [5, 9, 14, 18, 19, 21]. Among DWM patients undergoing MRI, neuroimaging findings showed herniation of the lower pole of the posterior fossa cyst through the foramen magnum [5, 6, 9, 14, 18, 19, 21]. The age at diagnosis varied widely from 9 months to 39 years (mean 16.6 years). Most frequent symptoms were headache, especially in the occipital region, and upper extremities sensorymotor alteration [1, 5–7, 9, 18]. Regarding the treatment for this condition, many surgical procedures have been used such as sub-occipital decompression with C1 laminectomy and duroplasty [5, 7, 8, 18], cyst decompression and plugging of obex [1], VPS and CPS [5, 9, 19, 21], and syrinx shunting [6]. Globally, postoperative outcome was favorable for all patients with a reduction in size of the syrinx in three patients [5, 9, 21] and complete regression in other three [6, 18, 19]. Among the reports without imaging, clinical improvement was observed in two cases [1, 7].

Discussion

The co-existence of syringomyelia and DWM represents a challenge both for treatment and prognosis and arises interesting questions about the pathogenesis and the best surgical approach.

Mechanisms of spinal cord cavitation

Different possible mechanisms have been proposed to explain the formation of the syrinx in patients with DWM. A first mechanism consists in dilation of a central canal (hydromyelia) anatomically connected to the fourth ventricle. In the presence of hydrocephalus, the impaired CSF circulation out of the fourth ventricle is thought to increase CSF pressure directly on the central canal (communicating syringomyelia). This type of syrinx was found in patients with DWM and Chiari-II malformation and it is also called "fifth-ventricle." A second mechanism is the impairment of CSF circulation due to obstruction at the level of the foramen magnum, which generates dilation of the central canal that is separated from the fourth ventricle (non-communicating syringomyelia). This type of syrinx was especially associated with Chiari-I malformation (CIM) [2, 12, 17]. Analyzing DWM patients with MRI, the responsible mechanism for syrinx formation was a CIM-like malformation in all cases but one [5, 9, 14, 18, 19, 21]. Also in our case, MRI and intraoperative findings were consistent with an impairment of CSF circulation acting as the abovementioned CIM-like mechanism.

Table 1 Reported cases of syringomyelia in association with DWM in autopsy series and MRI series

Authors	No. of pts	Age (years)	Sex	Syrinx extension	Treatment	Comments
Whitten et al. [23]	1	2	F	NA	Suboccipital craniectomy, excision of the cyst	The patient died due to postoperative complications. Moderate hydromyelia found at autopsy
Vuia and Pascu [22]	1	neonate	NA	Holocord	None	The patient died for hydrocephalus and respiratory disorder
Hart et al. [11]	1	12	NA	NA	NA	Previous surgery for communicating hydrocephalus
Hinokuma et al. [13]	1	9	F	C1-T8	None	A ventriculo-sagittal sinus shunt was performed to treat hydrocephalus 2 months prior to death
Milhorat et al. [16]	3	NA	NA	NA	NA	3 cases mentioned in MRI series of syringes with hindbrain lesions
Milhorat et al. [17]	2	NA	NA	NA	NA	NA

NA not available, *MRI* magnetic resonance imaging

Table 2 Reported cases of syringomyelia in association with DWM in clinical series

Authors	No. of pts	Age at surgery (years)	Sex	MRI yes/no	Syrinx extension	Clinical presentation	Treatment	Mechanism of syrinx formation	Comments
Gardner et al. [7]	1	24	F	No	NA	Severe headache, frequent falls, sensory loss in upper extremities, scoliosis	Suboccipital decompression	Communicating syrinx (fifth ventricle)	Clinical improvement at 2 months of FU
Gardner [8]	3	NA	NA	No	NA	NA	Suboccipital decompression	Communicating Syrinx (fifth ventricle)	None
Baker and Rydell [1]	1	25	F	No	Cervicothoracic	Back pain, right lower extremity weakness	Cyst decompression and plugging of obex	Not reported	Clinical improvement at 3-month FU
Harlow and Drose [10]*	1	neonate	NA	Yes	Upper cervical	NA –	Resection of the encephalocele, no treatment for DWV or syrinx	NA	DWV with hindbrain encephalocele
Tekkök and Ventureyra [21]	1	2	F	Yes	Cervicothoracic	Incidental diagnosis	VPS revision	Non-communicating Syrinx (CIM-LM)	Decrease in side of the syrinx at FU
Cinalli et al. [5]	1	7	M	Yes	Cervicothoracic	Gait disturbances	CPS revision (3 times), suboccipital decompression, C1 laminectomy and duroplasty	Non-communicating Syrinx (CIM-LM)	Decrease in side of the syrinx at 3-month FU
Hammond et al. [9]	1	39	M	Yes	Cervicothoracic	Headache, stabs of pain over the occiput with coughing	CPS revision	Non-communicating Syrinx (CIM-LM)	Decrease in side of the syrinx at 12-month FU
Erdal et al. [6]	1	21	F	Yes	Cervical	Pain and weakness in upper extremities	Syrinx shunting	Communicating Syrinx (fifth ventricle)	Complete resolution of syrinx at 3-month FU
Owler et al. [18]	1	29	F	Yes	Upper cervical	Occipital headache, upper extremities paresthesia	Suboccipital craniectomy, C1 laminectomy and duroplasty	Non-communicating Syrinx (CIM-LM)	Complete regression of the syrinx at 6-month FU
Richter and Pincus [19]	1	0.7	F	Yes	Cervicothoracic	No apparent symptomatology	CPS	Non-communicating Syrinx (CIM-LM)	Complete regression of the syrinx at 6-month FU
Kasliwal et al. [14]	1	2	M	Yes	Cervicothoracic	Delayed motor milestones, swelling along shunt track	none (CPS proposed but refused by parents)	Non-communicating Syrinx (CIM-LM)	None
Lee et al. [15]**	1	10	M	Yes	Cervicothoracic	Fecal incontinence, scoliosis	Suboccipital craniectomy and membrane excision	Non-communicating Syrinx (CIM-LM)	Thickened membrane/post-hemorragic arachnoid web. Decrease in size of the syrinx at 6-month FU
Our case	1	11	F	Yes	Holocord	Cervicalgia	VPS revision, CPS placement	Non-communicating Syrinx (CIM-LM)	Complete regression of the syrinx at 12-month FU

NA not available, *DWV* Dandy-Walker variant, *FU* follow-up, *CIM-LM* Chiari I malformation-like mechanism

*Case of Dandy-Walker variant

**A case of arachnoid web at foramen magnum as a complication of intraventricular hemorrhage during shunt revision surgery in a Dandy-Walker patient

Nonetheless, Erdal et al. observed high-flow CSF drainage after syrinx shunting despite a CIM-like mechanism at imaging, thus unveiling that the pathogenic mechanisms might vary across patients and that they cannot be easily identified before surgery.

Treatment

CPS and VPS placement/revision appears to be the treatment of choice in most cases from the 1990s. In fact, decompressing the foramen magnum by either the resolution of the hydrocephalus or the drainage of the posterior fossa cyst or both seem to be appropriate to treat the impaction at this level with a good post-operative outcome [9, 19, 21]. A direct approach to craniovertebral junction by suboccipital decompression with C1-laminectomy and duroplasty was the treatment of choice in the first attempts to treat DWM-associated syringomyelia [1, 7, 8]. Nowadays, it has become a secondary treatment when shunting is ineffective [5]. In our case, size increase and foramen magnum herniation of the posterior cranial fossa cyst was attributed to shunt dysfunction even though supratentorial hydrocephalus appeared stable. Despite intraoperative evidence of distal shunt malfunction, shunt revision showed to be inefficient and direct shunting of the posterior cranial fossa cyst was necessary. According to the literature and the present case, it seems that DWM-associated syringomyelia has a rather complex management and the treatment is still not standardized, often requiring a multicompartimental approach.

Post-operative follow-up

Despite different surgical approaches, the outcome was globally favorable with decrease in size or disappearing of the syrinx at radiological follow-up [5, 6, 9, 18, 19, 21] and/or neurological improvement [1, 7]. A few literature cases and the present report seem to confirm that posterior cranial fossa cyst shunting might represent an effective treatment for DWM-associated syringomyelia and more aggressive treatments should be restrained to cases with no clinical and imaging improvement.

Conclusions

DWM-associated syringomyelia is a rare complication with a poorly understood pathogenesis likely due to CSF circulation impairment. This condition might have a satisfactory outcome but often requires a multicompartimental approach. Longer follow-up studies are necessary to unravel whether ventriculoperitoneal shunt alone or combined with cystoperitoneal shunt can be considered valuable alternative to a more aggressive surgery at the craniocervical junction.

Compliance with ethical standards

Conflict of interest The authors report no conflict of interest concerning the materials or methods used in this study or the findings disclosed in this paper.

References

1. Baker GS, Rydell RE (1971) Dandy-Walker malformation: association with syringomyelia. Minn Med 54:889–893
2. Ball MJ, Dayan AD (1972) Pathogenesis of syringomyelia. Lancet 14:799–801
3. Bosemani T, Orman G, Boltshauser E, Tekes A, Huisman TAGM, Poretti A (2015) Congenital abnormalities of the posterior fossa. Radiographics 35:200–220. https://doi.org/10.1148/rg.351140038
4. 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
5. Cinalli G, Vinikoff L, Zerah M, Renier D, Pierre-Kahn A (1997) Dandy-Walker malformation associated with syringomyelia Case Illustration. J Neurosurg 86:571. https://doi.org/10.3171/jns.1997.86.3.0571
6. Erdal M, Plikcioglu AC, Bikmaz K, Cøsar M (2003) Dandy-Walker complex and syringomyelia in an adult: case report and discussion. Neurosurgery 52:1504–1505
7. Gardner WJ, Abdullah AF, McCormack LM (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
8. Gardner WJ (1965) Hydrodynamic mechanism of syringomyelia: its relationship to myelocele. J Neurol Neurosurg Psychiatry 28:247–259
9. 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
10. Harlow CL, Drose JA (1992) A special technique for cervical spine sonography. Illustrated by a patient with meningoencephalocele, Dandy-Walker variant, and syringomyelia. J Ultrasound Med 11:502–506
11. Hart MN, Malamud N, Ellis WG (1972) The Dandy-Walker syndrome. A clinicopathological study based on 28 cases. Neurology 22:771–780
12. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, Eidsath A, Talbot T, Morris J, Eskioglu E, Oldfield EH (1999) Elucidating the pathophysiology of syringomyelia. J Neurosurgery 91:553–562
13. Hinokuma K, Ohama E, Oyanagi K, Kakita A, Kawai K, Ikuta F (1992) Syringomyelia. A neuropathological study of 18 autopsy cases. Acta Pathol Jpn 42:25–34
14. Kasliwal MK, Suri A, Sharma BS (2008) Dandy Walker malformation associated with syringomyelia. Clin Neurol Neurosurg 110:317–319
15. Lee HC, Choi JW, Lee JY, Phi JH, Kim SK, Cho BK, Wang KC (2017) Syringomyelia caused by an arachnoid web in a patient with shunted Dandy-Walker malformation. Childs Nerv Syst 33:665–670. https://doi.org/10.1007/s00381-016-3293-x
16. Milhorat TH, MIller JI, Johnoson WD, Adler DE, Heger IM (1993) Anatomical basis of syringomyelia occurring with hindbrain lesions. Neurosurgery 32:748–754

17. 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
18. Owler BK, Halmagyi GM, Brennan J, Besser M (2004) Syringomyelia with Chiari malformation; 3 unusual cases with implications for pathogenesis. Acta Neurochir 146:1137–1143
19. Richter EO, Pincus DW (2006) Development of syringohydromyelia associated with Dandy-Walker malformation: treatment with cystoperitoneal shunt placement. Case report. J Neurosurg 104(3 Suppl):206–209
20. Spennato P, Mirone G, Nastro A, Buonocore MC, Ruggiero C, Trischitta V, Aliberti F, Cinalli G (2011) Hydrocephalus in Dandy-Walker malformation. Childs Nerv Syst 27:1665–1681. https://doi.org/10.1007/s00381-011-1544-4
21. Tekkök IH, Ventureyra EC (1997) Hydrosyringomyelia associated with Dandy-Walker malformation-is it really rare or undiagnosed? Eur J Paediatr Neurol 1:49–51
22. Vuia O, Pascu F (1971) The Dandy-Walker syndrome associated with syringomyelia in a newborn infant. Confin Neurol 33:33–40
23. Whitten CA Jr, Moyar JB, Wise BL (1962) Hydrocephalus syndrome. Obstruction of the foramina of the fourth ventricle. Am J Dis Child 103:55–60