Case Report

Dandy–Walker Malformation Presenting with Refractory Seizures and Unilateral Pan-ophthalmitis in Infancy: Successful Management with Dual Ventricular Shunting

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ABSTRACT:

Dandy–Walker malformation (DWM) is a congenital posterior fossa anomaly frequently associated with hydrocephalus and neurodevelopmental impairment. We report an unusual presentation of DWM in an infant with recurrent convulsions and unilateral pan-ophthalmitis leading to loss of vision, managed successfully with emergency dual ventriculoperitoneal shunting using a Y-connector. At 27 months of age, the child demonstrates normal neurodevelopment with seizure control on tapering antiepileptic therapy.

Key Words: Dandy–Walker malformation, Congenital hydrocephalus, Dual ventriculoperitoneal shunt, Y-connector shunt system, Infantile seizures, Posterior fossa malformation

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Background

Dandy–Walker malformation is characterized by cystic dilatation of the fourth ventricle, partial or complete agenesis of the cerebellar vermis, and enlargement of the posterior fossa. Hydrocephalus is a common and often early complication requiring surgical intervention. The optimal surgical strategy remains debated, particularly in complex cases with supratentorial and infratentorial CSF compartment involvement.

Case Presentation

An 8-month-old male infant presented with:

• Recurrent episodes of convulsions, beginning at 4 months of age

• History of unilateral panophthalmitis, resulting in complete loss of vision in right eye at 6 months of age

• The unilateral panophthalmitis was managed conservatively under ophthalmological supervision

• Progressive increase in head size

On examination:

• Occipitofrontal circumference (OFC): 48 cm at 8 months, consistent with macrocephaly

• Tense anterior fontanelle

• No focal motor deficits

• Developmental delay appropriate to raised intracranial pressure

Investigations

A non-contrast CT scan of the brain revealed:

• Features consistent with Dandy–Walker malformation

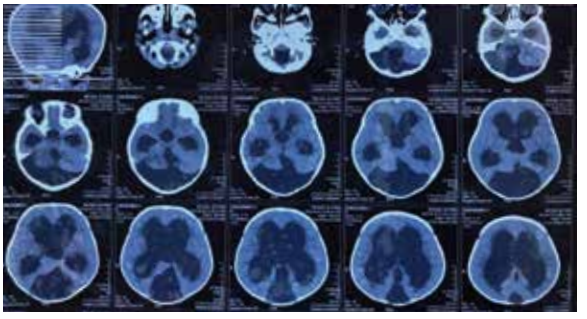
• Marked dilatation of the fourth ventricle

• Associated supratentorial hydrocephalus

The findings indicated impaired CSF dynamics involving both the lateral and fourth ventricles. [Fig-1,2]

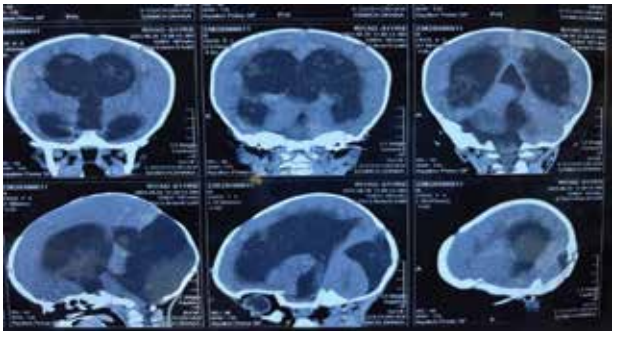
Figure Legends

Figure 1. Axial CT scan of the brain (non-contrast)



Axial sections demonstrate marked dilatation of the lateral ventricles with associated cortical thinning, consistent with severe hydrocephalus. A markedly enlarged fourth ventricle is also noted, suggestive of impaired cerebrospinal fluid outflow in keeping with Dandy–Walker malformation.

Figure 2. Coronal & Sagittal CT scan of the brain (non-contrast)



Coronal images show cystic dilatation of the fourth ventricle with partial agenesis of the cerebellar vermis and upward displacement of the cerebellar hemispheres. Associated supratentorial ventricular dilatation is evident, confirming the diagnosis of Dandy–Walker malformation with hydrocephalus. Sagittal reconstruction demonstrates hypoplastic cerebellar vermis.

Treatment

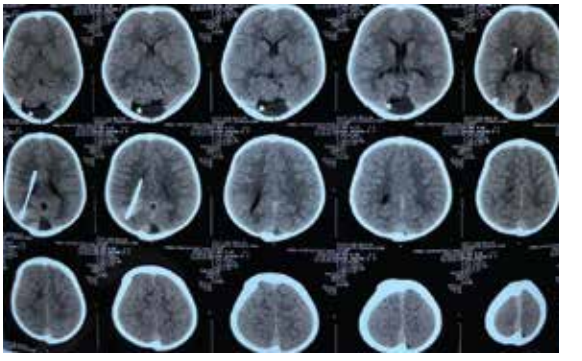
Given the severity of hydrocephalus and ongoing seizures, an emergency ventriculoperitoneal shunt was performed with:

• One catheter placed in the lateral ventricle

• A second catheter placed in the fourth ventricle

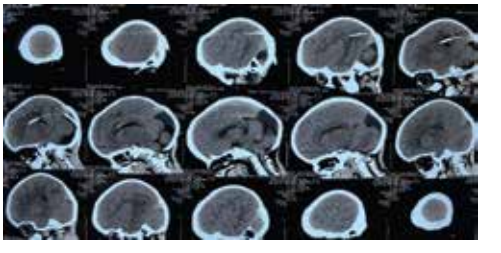
• Both catheters connected via a Y-connector to a single peritoneal distal catheter. The posterior fossa (fourth ventricular) limb was valveless, while the lateral ventricular limb incorporated a medium-pressure valve, allowing controlled supratentorial drainage with simultaneous free infratentorial decompression. [Fig-3,4]

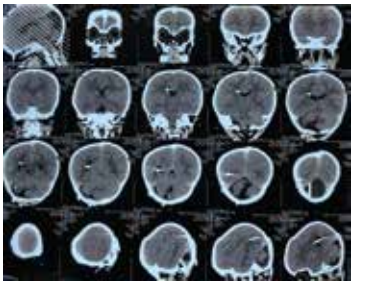
Figure 3. Post-operative axial CT scan of the brain



Post-operative axial images show satisfactory placement of ventricular catheters within the lateral and fourth ventricles, connected via a Y-connector system. There is a reduction in ventricular size, indicating effective cerebrospinal fluid diversion.

Figure 4. Post-operative sagittal & coronal CT scan of the brain





The images confirm decompression of the posterior fossa and supratentorial ventricular system, with maintained catheter position and improved cerebrospinal fluid dynamics.

Figure 5. The child shows normal neurodevelopment for age at 27 months

Postoperatively, the patient was managed with antiepileptic medication and close neurological monitoring.

Outcome and Follow-Up

The postoperative course was uneventful.

At 27 months of age:

• The child shows normal neurodevelopment for age [Fig-5]

• No recurrence of seizures

• Antiepileptic medication is being gradually tapered

• The shunt remains functioning well, with no features of raised intracranial pressure

• During follow-up, the ophthalmological evaluation was taken. Though resolution of active infection on right eye, but the vision loss remained permanent.

Discussion

Dandy–Walker malformation (DWM) is a rare congenital posterior fossa anomaly, with an estimated incidence of approximately 1 in 25,000–35,000 live births. It is classically defined by a triad consisting of cystic dilatation of the fourth ventricle, partial or complete agenesis of the cerebellar vermis, and enlargement of the posterior fossa with upward displacement of the tentorium. Hydrocephalus develops in up to 70–90% of patients, often dictating clinical presentation and prognosis. [1, 2]

Seizures are a recognized but variable manifestation of DWM and are usually secondary to raised intracranial pressure, cortical maldevelopment, or associated supratentorial anomalies. In the present case, recurrent convulsions beginning at four months of age were likely related to progressive hydrocephalus and cortical irritation, supported by the markedly enlarged occipitofrontal circumference and imaging findings. Early-onset epilepsy in DWM has been associated with poorer neurodevelopmental outcomes when CSF diversion is delayed. [3]

An unusual and noteworthy feature in this patient was the history of unilateral pan-ophthalmitis with subsequent loss of vision. Although pan-ophthalmitis is not a classical association of DWM, raised intracranial pressure, altered CSF dynamics, and potential immune vulnerability in infancy may predispose to severe ocular infections. The coexistence of DWM, hydrocephalus, and severe ocular pathology underscores the need for multidisciplinary vigilance in such complex presentations.

Management of hydrocephalus in DWM remains controversial. Surgical options include ventriculoperitoneal (VP) shunting, cysto-peritoneal shunting, combined shunting procedures, and endoscopic approaches. Isolated lateral ventricular shunting may be insufficient in cases where the fourth ventricle is massively dilated and poorly communicating, leading to persistent posterior fossa compression. [4,5]

In this patient, imaging demonstrated significant involvement of both supratentorial and infratentorial compartments. The decision to perform dual ventricular shunting—draining both the lateral ventricle and the fourth ventricle via a Y-connector into the peritoneum—was therefore anatomically and physiologically justified. This strategy allowed balanced CSF diversion from both compartments while minimizing distal hardware and reducing the risk of differential pressure gradients. Similar approaches have been reported with favorable outcomes in selected DWM cases. [6,7]

The excellent long-term outcome in this child—normal neurodevelopment at 27 months of age with successful tapering of antiepileptic medication—highlights several key principles. First, early recognition and urgent CSF diversion can reverse the effects of raised intracranial pressure even in severe malformations. Second, tailored shunting strategies based on ventricular anatomy can

significantly improve outcomes. Finally, seizure control following adequate CSF diversion supports the hypothesis that hydrocephalus was the primary epileptogenic factor rather than irreversible cortical dysgenesis.

This case adds to the limited literature demonstrating that appropriately selected dual-compartment CSF diversion in DWM can result in excellent neurological and developmental outcomes, even in the presence of early seizures and significant associated morbidity.

Learning Points

• Dandy–Walker malformation may present with early-onset refractory seizures

• Hydrocephalus in DWM may require diversion from both lateral and fourth ventricles

• Dual ventricular shunting with a Y-connector can be effective in selected cases

• Early intervention can result in normal neurodevelopmental outcomes

Patient Consent

Written informed consent was obtained from the child's parents for publication of this case and accompanying images.

Conflicts of Interest

None declared.

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