CASE REPORT

DANDY-WALKER MALFORMATION (DWM): CASE REPORT OF A DELAYED PRESENTATION

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ABSTRACT

Background: Dandy-Walker Malformation (DWM); a group of posterior fossa malformation characterized by a cystic enlargement of posterior fossa with high lying torcula herophili (fetal position) and a hypoplastic vermis pushed and rotated superiorly by the cyst occurs in 1 in 25,000 – 35,000 live birth and accounts for 1 – 4 % of all Hydrocephalus seen within 3 months of birth.[1] **Case report:** An 8-year-old girl from an orphanage who presented with a history of progressive head enlargement, delayed developmental milestones and poor vision. No history suggestive of raised intracranial pressure, differential limb weakness or associated congenital systemic anomalies. All fontanelles were fused. Muscle power was normal, globally spastic with brisk deep tendon reflexes. The eye examination revealed bilateral light perception only. Brain CT Scan revealed a large posterior fossa cyst, hydrocephalus and corpus callosum hypoplasia. The patient had a double shunt (ventriculoperitoneal and cysto - peritoneal). Currently on rehabilitation and can count fingers. **Conclusion:** Early identification and prompt intervention minimizes the neurological sequelae.

Keywords: Hydrocephalus, Dandy-walker, Shunt

INTRODUCTION

Dandy-Walker Malformation (DWM) is a group of posterior fossa malformations which include a large thin-walled cyst occupying an enlarged posterior fossa, with a high lying torcula herophili (fetal position) and a hypoplastic vermis rotated superiorly by the cyst.[1] It results from abnormalities of rhombencephalon formation and arrest of hindbrain development, also by secondary insults of varying severity to the cerebellum and the 4th ventricle. This results in agenesis of the cerebellar vermis with a large posterior fossa cyst communicating with an enlarged 4th ventricle.[2, 3] Both sporadic and syndromic forms do exist. Few were found to have an Autosomal recessive transmission. It occurs in 1 in 25,000 – 35,000 live birth[2] and accounts for 1 – 4 % of all Hydrocephalus seen within 3 months of birth. [1] About 50% of all patients with DWM would ultimately develop hydrocephalus. Has slight preponderance for females, with Male: female ratio of 1: 3. Common clinical findings include Macrocephaly (80%), Delayed Developmental milestones (35%), Features of raised intracranial pressure; sun-setting eyes, seizures, spasticity. Other associated anomalies include: polydactyly, cardiac, facial and rarely Neural Tube Defects (NTD).[1]

The confirmatory investigation is by Neuro-Imaging. Computed Tomography (CT) of the Brain usually shows Cystic enlargement of the posterior fossa, and hydrocephalus (75% of cases). However, Magnetic Resonance Imaging (MRI) is the imaging of choice: this shows large posterior fossa cyst which may, or may not communicate with 4th ventricle, high – lying torcular, hypoplastic corpus callosum (20%), and cortical dysplasia (10%).

Iohexol Ventriculography can be done to demonstrate if the cyst is communicating with the 4th Ventricle or not.

Management is essentially Surgery in symptomatic patients. This has changed in the last two decades from excision of the Cyst to Shunting and Endoscopic Third Ventriculostomy (ETV).

CASE REPORT

We present an 8-year-old girl from an orphanage with a history of large head, delayed developmental milestones and poor vision. However, no history suggestive of raised intracranial pressure, differential limb weakness. There were no facial, musculoskeletal or other systemic anomalies. The patient had macrocephaly with Occiput-frontal circumference (OFC) of 62 cm. All the fontanelles were fused. Eyes examination revealed light perception only with bilateral papilloedema. She was found to be globally spastic with normal muscle power, and brisk deep tendon reflexes. Computed tomography (CT) Brain with sagittal reconstruction revealed a large posterior fossa cyst, Hydrocephalus and hypoplasia of the corpus callosum. Magnetic Resonance Imaging (MRI) and Iohexol Ventriculography were not available in our center and as such were not utilized.

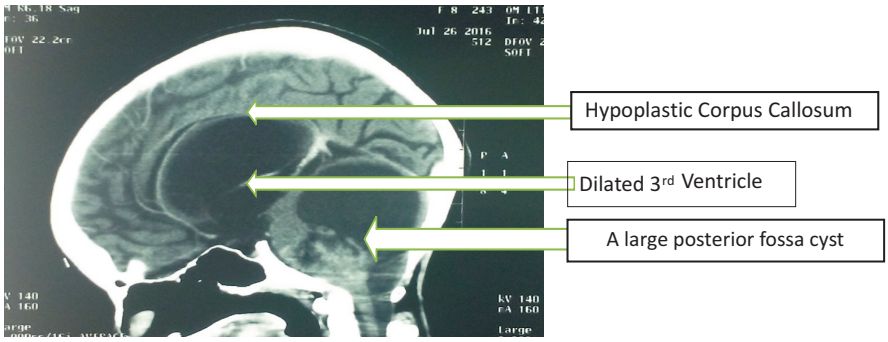


Figure 1: Brain CT Scan (Reconstruction)

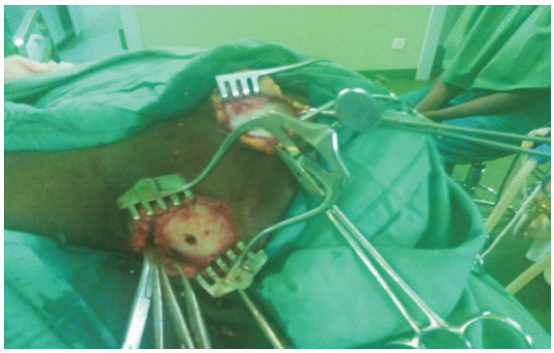
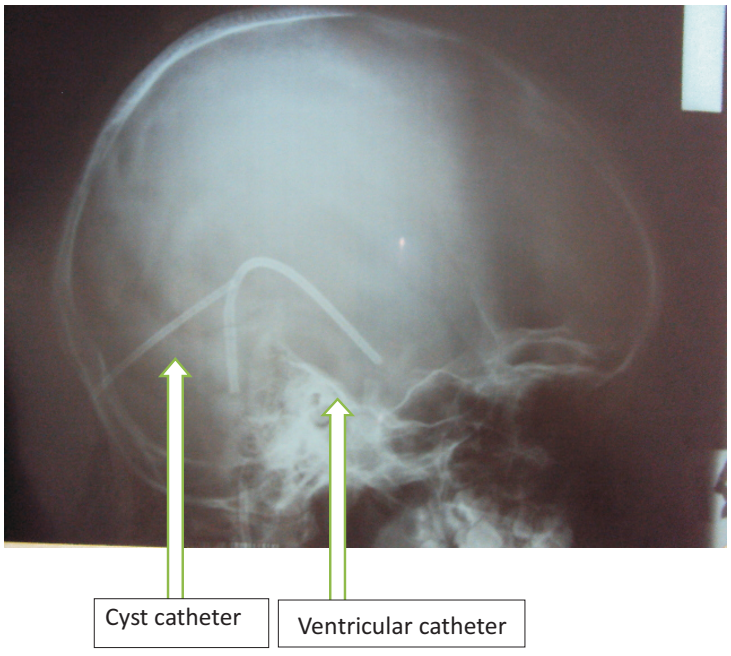


Figure 2: Intraoperative photograph showing double cranial burr hole

Patient had ventriculoperitoneal and cyst-peritoneal shunts. Postoperatively, the patient was commenced on regular physiotherapy. Currently, she has achieved a marked reduction in spasticity and can count fingers (visual improvement).



DISCUSSION:

DWM is characterized by an enlarged posterior fossa with complete or partial agenesis of the cerebellar vermis and cystic dilatation of the fourth ventricle, distorted and encased in a membrane. First described by Dandy & Blackfan in 1914, and was named Dandy-Walker malformation forty years later by Bender to acknowledge Taggart and

Walker's contribution in 1942.[2] It is essentially a large cyst containing CSF surrounded by pia and arachnoid maters. Over 85% of DWM develop hydrocephalus. Therefore, it is an important cause of post-natal hydrocephalus. Delayed developmental milestone had been reported in 70% mainly from additional CNS malformations.

Risk factors for its development include gestational exposure to infections like rubella, CMV, and toxoplasmosis. Drugs like warfarin, alcohol, and isotretinoin are also thought to be predisposing factors.[4] Generally, differential diagnosis of posterior fossa CSF containing cysts include: Dandy-Walker malformation (DWM), Dandy-Walker variant (DWV); characterized by vermian hypoplasia, normal size posterior fossa, Persistent Blake's pouch cyst (BPC): dilatation of all ventricles, communication between the posterior fossa cyst and 4th ventricle with or without hypoplastic vermis.

Associated CNS anomalies include Agenesis of the corpus callosum (17%),[5] Neural tube defects (NTD), Occipital encephalocele (7%), atresia of foramina of Magendie and Luschka.[6] etc. Other Systemic anomalies include Cardiovascular, facial, and ocular manifestations.[1,5]

Apart from Computed tomography (CT Scan) and MRI, iohexol ventriculography to demonstrate communication between the cyst and 4th ventricle is important in determining the number of cranial shunts needed for surgical intervention.

Surgical treatment options include 1) cyst excision 2) Shunting - which may be a single supratentorial shunt (Ventricle) placed if the cyst is demonstrated to be communicating with the 4th ventricle. Caution: if there is no communication, there is increased risk of upward herniation.[7] However, when the cyst is not communicating with the 4th ventricle, a double shunt (supratentorial and infratentorial) separately or joined by a "Y" connector is placed. When this communication is not demonstrable, however, putting a double cranial shunt is safe 3) Endoscopic Third Ventriculostomy (ETV) suffices if the aqueduct is patent. It is an acceptable alternative to shunting. Success with ETV depends on the age of the patient. Up to 70% success rate had been reported[7].

Prognosis/Outcome depends on severity and duration before intervention. Historically, mortality was 20 – 30 %. Bindad and colleagues – reported 14% mortality, increased cyst shunt failure, 6.5% shunt revision per patient over a 7-year period. Other notable complications determining outcome include low IQ (in 50%) from the presence of other congenital CNS anomaly, ataxia, spasticity, seizures (15%).

CONCLUSION

Despite its rarity, DWM is an important cause of hydrocephalus in infancy and therefore early decompression of ventriculomegaly and Drainage of the cyst is recommended in order to improve cognitive development in such patients.

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