Case Report

Dandy-Walker Syndrome in a Child at Rural Kelantan, Malaysia

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ABSTRACT

Dandy-Walker syndrome is a rare congenital malformation of the brain that involves the cerebellum and the fourth ventricle. It is characterised by a classical triad of hydrocephalus, cystic dilatation of the fourth ventricle and complete or partial agenesis of the vermis. Majority of cases are diagnosed during neonatal or early infantile period. In this case report, a seven-year-old boy complained of recurrent headaches for the past one year. Physical examination was unremarkable. Examination of the fundus on the same day revealed bilateral papilloedema. His subsequent computed tomography scan of the brain done at a major district hospital demonstrated features in keeping with Dandy-Walker malformation. Our case highlighted the importance of embarking on a detailed and thorough approach when dealing with a child with chronic headache, especially in rural settings where advanced medical equipment is not readily available.

Keywords: Dandy-Walker syndrome, hydrocephalus, chronic headache

INTRODUCTION

Dandy-Walker malformation represents a group of rare congenital abnormalities of the central nervous system with a reported incidence of one in 30,000 live births[1]. It is characterised by a neuro-pathological triad of hydrocephalus, cystic dilatation of the forth ventricle and complete or partial agenesis of the vermis[2]. Although commonly diagnosed in neonatal period, clinicians should have a high index of suspicion of chronic headache and vomiting and take this as a specific symptom of raised intracranial hypertension, unless proved otherwise.

CASE PRESENTATION

A seven-year-old boy presented with one year history of chronic and recurrent headache. Although unable to pinpoint the exact nature and site of headache, he commented that his symptoms occur almost on an every-other-day basis with no clear aggravating or alleviating factors. Besides, he suffers from recurrent vomiting episodes together with headache. Other associated symptoms, systems review, past histories, developmental history and social history were otherwise unremarkable. His mother expressed concerns that his headache has become increasingly disabling for the past few months as it was affecting both his sleep and learning opportunities at school due to frequent medical leaves. For the past one year, he was brought to nearby clinics on a few occasions to sought treatment where symptomatic analgesics were prescribed but to no avail.

Physical examination revealed an active and communicative child with no overt dysmorphic features. His height and weight were at the 50th centile and were consistent with his age. His head circumference was measured at 52 cm. Other parts of the examination including a thorough neurological examination were unremarkable. A fundoscopic examination of both eyes revealed presence of bilateral papilloedema (Figure 1).

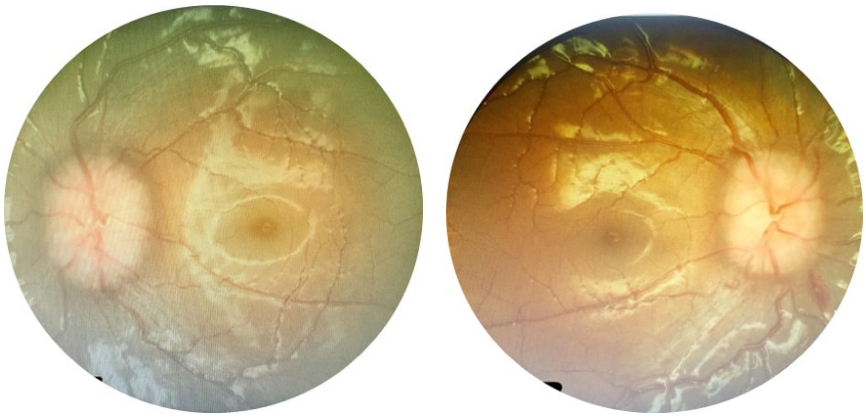

Figure 1 Bilateral fundus showing papilloedema

With a diagnosis of possible hydrocephalus or intracranial space-occupying-lesion in mind, he was sent to the nearest district hospital (approximately 90 km away) for an urgent computed tomography (CT) of the brain. CT scan report (Figure 2) demonstrated grossly-enlarged posterior fossa with agenesis of the cerebellar vermis associated with a dilated forth, third and both lateral ventricles. His corpus callosum appears to be normal with no other focal enhancing brain parenchymal lesion seen. These features are in keeping with Dandy-Walker malformation.

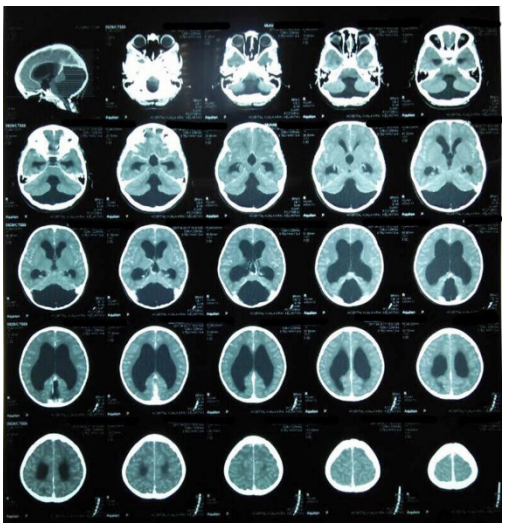

Figure 2 CT scan showing grossly-enlarged posterior fossa with agenesis of the cerebellar vermis associated with a dilated forth, third and both lateral ventricles

He was then transferred to a tertiary centre with neurosurgical support for further medical care and attention. A third ventriculostomy was subsequently performed endoscopically and went smoothly without complications, meaning that he was discharged well five days later. Both our patient and his mother reported resolution of headache during subsequent follow-up clinic visits.

DISCUSSION

Dandy-Walker syndrome refers to a rare congenital malformation of the cerebellum and the forth ventricle. It is worth mentioning that the term Dandy-Walker (or more precisely Dandy-Walker continuum) does not represent a single condition or entity, but rather several abnormalities of the posterior fossa that might

occur in isolation or might co-exist with each other. Current understanding identifies a few types of Dandy-Walker complexes, namely Dandy-Walker malformation, mega cisterna magna as well as Dandy-Walker variant[3]. Depending on the subtypes there may be a partial or complete absence of cerebellar vermis, an enlargement of the forth ventricle, as well as cyst formation adjacent to the base of the skull[4]. Various posterior fossa malformations and their association with brain anatomy are summarised in Table 1[4].

Table 1 Posterior fossa malformation type and relationship to brain anatomy[4]

	Cerebellar vermis	**Fourth ventricle**	**Brain stem**
Dandy-Walker malformation	Partially/ completely absent	Opens into large CSF-filled cyst	May be abnormal
Mega cisterna magna	Normal	Normal but large collection of CSF fluid	Normal
Dandy-Walker variant	Hypoplastic with variable-sized cyst	Mildly enlarged	Normal

Pathologically, it is believed that atresia of the foramina of Luschka and Magendie are responsible for such anomalies. An alternative explanation proposed by Benda mentioned that the syndrome represented errors in development in the region of forth ventricle but not limited to the foramina[5]. Recent researches indicated that Dandy-Walker syndrome might be related to various other conditions such as primary ciliary dyskinesia, polycystic kidney disease and Alstrom syndrome. Such conditions share a common pathology of dysfunctional ciliary motility that might come under an umbrella term named ciliopathies[6].

Patients with Dandy-Walker malformation typically exhibit symptoms during early infancy, including delayed motor development as well as progressive and disproportionate enlargement of the skull. In older children, symptoms of raised intracranial pressure such as irritability, vomiting, convulsion and headache do occur[5]. In our case, our patient was asymptomatic until the age of 6 where he starts developing recurrent headache and vomiting episodes.

Although there is no universal agreement regarding its management, some patients might benefit from neurosurgical procedures such as third ventriculostomy, or ventriculo-peritoneal shunts[7]. More importantly, the management of the patient with Dandy-Walker syndrome should be done in a holistic manner involving a multi-disciplinary team, including neurosurgeons, paediatricians, nurses, pharmacists as well as physiotherapists, occupational therapists, speech therapists, dieticians and specialised educators. Unfortunately, such specialised team might prove to be out of reach for a majority of people residing in rural areas (including our patient) facing issues with regards to assess to basic healthcare.

Moreover, it is vital to employ a systematic approach when dealing with a child with chronic headache. It is vital for clinicians to recognize that although the general approach to headaches in children is similar to that in adults, their manifestation can be less straightforward in children. Obtaining the exact description of headache might be challenging in young children. Thus, clinicians need to be more vigilant in eliciting collateral history from caretakers as well as explore the impact of headache on the child's behaviour and social capabilities. Fundoscopy remains a very important and safe procedure that should be performed in all children with symptoms suggestive of raised intracranial pressure. Chong SC (2004) in his article divides headache in children into four main clinical profiles based on clinical course[8] (Table 2). Various 'red-flags' in headache history that should alarm clinicians to a lower threshold for neuroimaging or further investigations[8] (Table 3).

Table 2 Differential diagnosis based on headache patterns[8]

Headache patterns	Possible aetiologies
Acute	*Localised* • Acute URTIs e.g. sinusitis, otitis media • Dental causes e.g. Dental abscess, temporal-mandibular joint dysfunction *Generalised* • Systemic infection e.g. meningitis *Central* • Acute intracranial haemorrhage
Acute recurrent	Migraine
Chronic, non-progressive	Psychogenic/ psychiatric causes Tension-type headache
Chronic, progressive	Space-occupying lesion Benign intracranial hypertension

Table 3 Redflags in headache history taking[8]

1.	A short history ('first' or 'worst') or recent recurrent severe headache for few weeks
2.	Headache suggesting raised intracranial pressure (vomiting in morning, pain disrobing sleep, early morning headache, headache worse with cough or Valsalva)
3.	Accelerated course, change in character over weeks or days
4.	Associated symptoms of personality changes, weakness, visual disturbances, focal weakness, confusion, seizures or fever
5.	Young age of child (less than three years old)
6.	Underlying history of neurocutaneous syndrome, history of systemic illnesses e.g. known malignancy with possible metastases, hypercoagulopathy

CONCLUSION

This case heightens the importance of doing complete neurological examination and developmental examination of a child with headache for one year with vomiting. To recommend early fundoscopic exam to rule out possible papilloedema for raised intracranial hypertension and do neuroimaging earlier.

ACKNOWLEDGEMENTS

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CONFLICT OF INTEREST

The authors declare that they have no competing interests in publishing this case.

CONSENTS

Written informed consent was obtained from the patient to publish the case. A copy of written consent is available for review by the Chief Editor.

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