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Case Report

Dandy-Walker variant: a diagnosis by clinical and CT evaluation in a tertiary hospital in Nepal

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ABSTRACT

Dandy-Walker variant (DWV) has been used to describe a combination of cystic dilation of the fourth ventricle and hypoplastic cerebellar vermis in the absence of an enlargement of the posterior fossa. Magnetic resonance imaging (MRI) is the modality of choice for diagnosing DWV. But it can also be diagnosed by evaluating the subject clinically and by CT scans. We presented here a case report of a patient with DWV who was diagnosed by clinical evaluation and CT scans. A 15 years old girl presented with the symptoms of pain and ear discharge from both ears. She had developmental delay, mental retardation and poor nutritional status. The CT scan of the patient showed a posterior fossa cyst with hypoplastic vermis and communicating hydrocephalus without obvious enlargement of the posterior fossa. Hence, a diagnosis of DWV was made. For the ear pathology, she underwent modified radical mastoidectomy and other medical treatments.

Keywords: Dandy-Walker variant, Cystic dilation, Hypoplastic, Cerebellar

INTRODUCTION

DWV has been used to describe a combination of cystic dilation of the fourth ventricle and hypoplastic cerebellar vermis in the absence of an enlargement of the posterior fossa.[1]

Dandy-Walker complex has several forms like Dandy-Walker malformation (DWM) includes cystic dilatation of the fourth ventricle, complete or partial agenesis of the cerebellar vermis and enlarged posterior fossa; Dandy-Walker variant (DWV) comprises cystic posterior mass with variable hypoplasia of the cerebellar vermis and no enlargement of the posterior fossa; the third variant mega-cisterna magna comprises enlarged cistern magna with normal cerebellar vermis and fourth ventricle.[2] Although MRI forms the basis of diagnosing DWV; MRI is too expensive and is not easily accessible to the poor people of developing nations. So, this case study reported a case of DWV from a remote area of a developing country that was accidentally diagnosed from CT scans done for an ear pathology and from the clinical evaluation.

CASE REPORT

A 15 year old girl presented to the ENT department of a tertiary care center with a complaint of pain and ear discharge from both ears. She was diagnosed as a case of chronic otitis media squamous type on left and mucosal type on right with a mastoid abscess on the left side.

She had an ataxic gait and was mentally challenged. Her mother told that she had an uneventful pregnancy with no history of trauma or drug use. However, the child was delivered on the 7th month of pregnancy and was

accompanied by antepartum hemorrhage for about 24 hours. She had a birth weight of 1.6 kilograms and had to be kept in an incubator for 20 days. She was the first of the two children of non-consanguineous parents. No other family member had similar problems.

All her developmental milestones were delayed. She also had a history of recurrent hospital visits for various infectious diseases like pneumonia and diphtheria in the past. Personal history revealed mental retardation, dependence on others for every daily activity like eating, dressing and going to the bathroom.

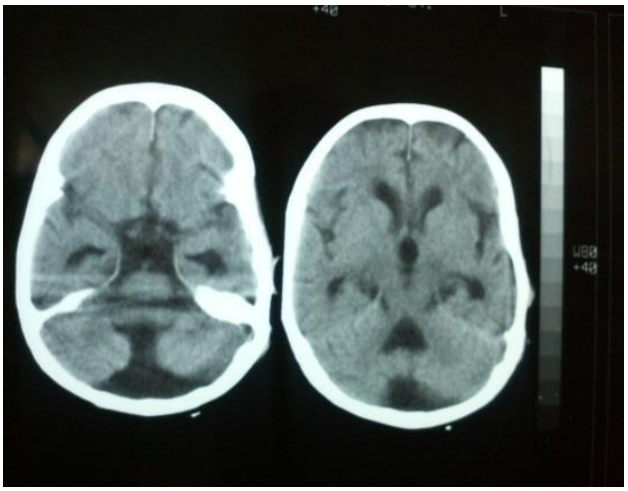

Figure 1: CT image shows posterior fossa which is communicating with the 4th ventricle in the midline and cerebellar vermis is hypoplastic.

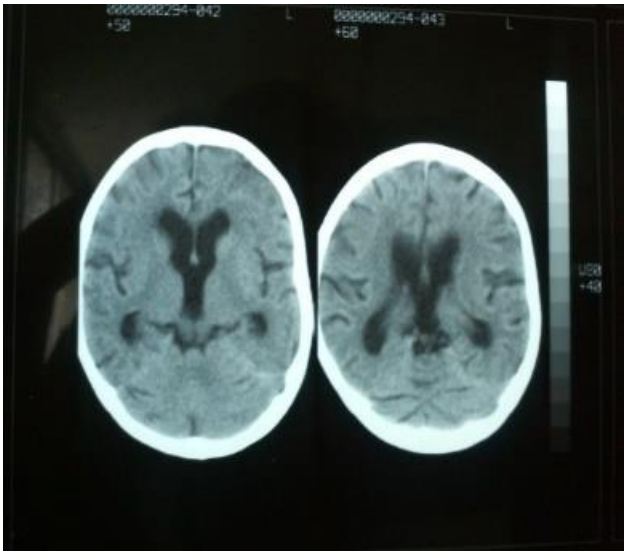

Figure 2: CT image shows rounding off of bilateral frontal horns with dilatation of 3rd and 4th ventricles.

On examination, she weighed 18 kilograms and was 120 centimeters in height with no gross congenital malformation. She had microcephaly, drooling of saliva but no other features of facial palsy had increased tone in all 4 limbs and an ataxic gait. She could hardly speak 2-3 words with meaning. Her sensory and cranial nerve examination was normal. Upon consent of her parents, her CT scan was done which showed chronic otitis media squamous type on left with mastoid abscess and mucosal type on right. Along with this the CT scan also showed a cystic lesion in the posterior fossa which is communicating with the 4th ventricle in the midline, hypoplastic cerebellar vermis and rounding of bilateral frontal horns with dilatation of 3rd and 4th ventricles without obvious enlargement of the posterior fossa (Figure 1 and 2). These findings thus directed towards the diagnosis of DWV. She was treated with modified radical mastoidectomy for the mastoid abscess.

DISCUSSION

Dandy-Walker malformation is a rare cranial abnormality and has an incidence of about one in every 25,000-35,000 live births with a slight female preponderance.[3] It accounts for 1-4% of antenatally diagnosed hydrocephalus.[3] There are various predisposing factors which include infections, cranial trauma, chronic disturbances in cerebrospinal fluid pressure, the persistence of embryonic tissue, vascular lesions, teratogenesis and maternal diabetes.[4] Previous studies showed that about 40% of children were intellectually normal, another 40% had mental retardation, 20% were borderline and our case fell in the 2nd group.[4] The gene locus for Dandy-Walker malformation is 3q24 and the presence of multiple congenital defects may shorten life span.[2]

The clinical manifestations of DWV are milder than those of Dandy-Walker malformation but may include mental retardation, cerebellar ataxia and symptoms of hydrocephalus including vomiting, irritability, convulsions. It may also be associated with schizophrenia, obsessive-compulsive disorders and bipolar disorders.[5] Out of these, the patients had mental retardation and cerebellar ataxia. The patient may present with psychomotor and growth retardation, hypotonia, strabismus, myopia, a short neck, microcephaly, brachycephaly, hypertelorism, the antimongoloid slant of palpebral fissures, globulus large nose, large mouth with down turned corners, poorly lobulated ears, high arch palate, cleft palate, small hands and feet, clinodactyly and the brachymesophalangy of the little fingers.[2] Our patient had psychomotor and growth retardation, strabismus, myopia and microcephaly.

The cerebellum plays a major role in motor control, motor learning and even cognition including the development of speech.[2] Our case had ataxic gait increased muscle tone, she had problems performing day-to-day activities and could only speak a few meaningful words. CT findings of DWV include inferior vermis hypoplasia, posterior fossa cerebrospinal fluid collection with communication with the fourth ventricle, and no obvious enlargement of the posterior fossa.[1] In our case,

the CT scan of the patient showed a posterior fossa cyst with hypoplastic vermis and communicating hydrocephalus without posterior fossa enlargement.

CONCLUSION

In conclusion, DWV is diagnosed typically by MRI findings which is very expensive and is not easily accessible to the poor people of a developing country. CT scan and thorough clinical evaluation could stand as a substitute for the more expensive MRI. Moreover, special care should be taken for such patients as they are prone to develop many infectious diseases like chronic otitis media in this case, as they are not able to maintain their basic hygiene. Due to poor access to health facilities and education even serious diseases are not given proper attention by the people and hence are either not diagnosed or are diagnosed very late and our case is no exception to this.

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