INTRODUCTION

Dandy-Walker malformation (DWM) lies at the most severe end of the spectrum of cystic posterior fossa anomalies. Several associated intracranial anomalies have been described including corpus callosum dysgenesis and gray matter abnormalities, as were seen in our case. Occipital cephalocele, a coexistent abnormality in this case, is a relatively remote association and has hardly been reported with DWM.

This case gives a comprehensive idea of all the commonly and infrequently described intracranial associations of DWM, all coexisting in the same patient and highlights the significance of sonography as an effective screening tool.

CASE DESCRIPTION AND MANAGEMENT

A 1-day-old female neonate was brought to the Department of Radio-diagnosis for ultrasonography of suspected caput succedaneum. Ultrasonography of the swelling revealed an anechoic cystic lesion in the scalp with intracranial communication through a gap in the occipital bone. Ultrasonography of the posterior fossa, done to ascertain the deeper extent of the lesion, revealed a large posterior fossa cyst communicating with a dilated fourth ventricle with associated vermian agenesis. The cerebellar hemispheres were hypoplastic and winged anterolaterally (FIGURE 1. I-II).

The findings were consistent with DWM. Complete sonographic evaluation of the brain was done in search of other associated intracranial anomalies demonstrating nodular projections isoechoic to gray matter along the lateral walls of both lateral ventricles with mild hydrocephalus. The splenium of corpus callosum was deficient with sulci radiating peripherally from the expected location of splenium giving a “spoke-wheel” gyral pattern (FIGURE 1. III-VI). The mother had never undergone an antenatal anomaly scan. Magnetic resonance imaging of the neonatal brain was advised for corroboration.

Magnetic resonance imaging performed after a month validated the findings of classic DWM with associated periventricular gray matter heterotopia, corpus callosal dysgenesis, hydrocephalus and occipital cephalocele.
communicating with the posterior fossa cyst. Additional important information obtained from MRI brain was the kink at the cervicomedullary junction, indicating a poor prognosis (FIGURE 1. VII-X). There was progressive increase in hydrocephalus compared to the initial scan.

The patient never underwent any definitive treatment or corrective surgery for hydrocephalus. A follow-up one month later revealed that the baby had respiratory distress and a poor suck reflex. Patient was under symptomatic and supportive treatment (oxygen support and paladai feeding) and was stable. Unfortunately, a subsequent follow up after six months she passed away one week back.
DISCUSSION

Almost all the known intracranial associations of DWM were seen in our case, including the relatively rare occipital cephalocele and no such case with all these associations in a single patient was found in existing literature. This case is a prototype of classic DWM with a natural course as described classically in literature and highlights the ominous coexistence of cervicomedullary kink. The only limitation of our report is that no definitive treatment or corrective surgery could be undertaken.

DWM is a rare spectrum of congenital anomalies with an incidence of 1 in 30,000 live births with mild female predisposition, as in our case. This spectrum of cystic posterior fossa anomalies comprises of classic DWM, Blake pouch cyst, inferior vermian hypoplasia and mega cisterna magna. DWM is the most severe anomaly in the spectrum with the classic triad of enlarged posterior fossa, upward displacement of the confluence of sinuses with lambdoid-torcular inversion and cystic dilatation of the fourth ventricle. Vermian agenesis can be complete or partial. This classic triad was seen in the index case.

The diagnosis of DWM warrants search for other extracranial anomalies due to syndromic associations like Klippel-Feil syndrome, Aicardi syndrome, Trisomy 18 and PHACE syndrome. Common intracranial associations are corpus callosal dysgenesis, gray matter heterotopia and hydrocephalus. Occipital cephalocele is a rare association, with only 33 such cases described in literature as of 2022.

Ultrasoundography and magnetic resonance imaging done one month apart revealed progressive hydrocephalus in our case. Standard surgical treatment includes ventriculoperitoneal or cistoperitoneal shunts for relief of hydrocephalus. Besides, supportive treatment like oxygen therapy and alternative feeding strategies are required, the only measures employed in our case.

Any couple who had experience of having a baby with DWM, must undergo an anomaly scan because there is a high incidence of such a condition among siblings.

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Ethical approval
Ethical approval was not sought because this is a case report. However, informed written consent was obtained from the patient for preparation of this manuscript.

Data availability statement
The data that support the findings of this study are available on request from the corresponding author.

REFERENCES


