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Case Report

A Rare Case of Dandy Walker Malformation with Widened Anterior Fontanelle on Physical Examination

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Abstract

Dandy-walker syndrome is characterized by hypoplasia of cerebellar vermis and cystic enlargement of the fourth ventricle.

Majority of patients present with hydrocephalus which may be associated with atresia of the foramen of magendie.

Approximately 70% - 90% of patients have hydrocephalus. It may be associated with atresia of the foramen of magendie and possibly, the foramen of Luschka. Dandy walker syndrome occurs in one out of every 25,000 live births, occurs more frequently in females than males [1].

This report presents DWS in a 40 day old baby boy.

Keywords: *Dandy Walker Syndrome [DWS]; Hydrocephalus*

Introduction

Dandy-walker syndrome is characterized by hypoplasia of cerebellar vermis and cystic enlargement of the fourth ventricle.

Case Study

A 40 day old male baby boy was born prematurely at 36 weeks post cesarean section following intra uterine fetal distress.

Right after birth, the baby developed difficult in breathing and delayed crying with BWT=2.3 Kg with Apgar Score 3,9.

He was born drowsy and cyanosed SPO_2 (60% - 90%) and kept on oxygen therapy.

Resuscitation with adrenaline was done and on the 4th day post admission, he got discharged.

With a 2 week follow up, the mother reports that the baby breastfeeds well, no history of convulsion, no history of high pitch cry however on examination there is very wide anterior fontanelle.

He was referred to cranial ultrasound and the result showed cystic mass in the posterior brain fossa communicating with the 4th ventricle.

Also, the lateral ventricles were seen dilated with persistent cavum septum and the third ventricle was collapsed.

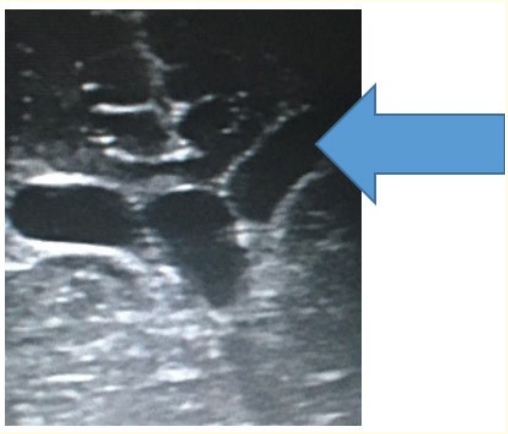

Figure 1: *The arrow above points to enlarged lateral ventricles on cranial USS.*

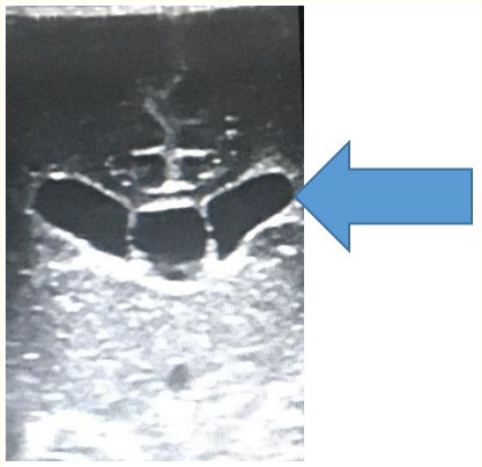

Figure 2: *The arrow above points to enlarged lateral ventricles on cranial USS.*

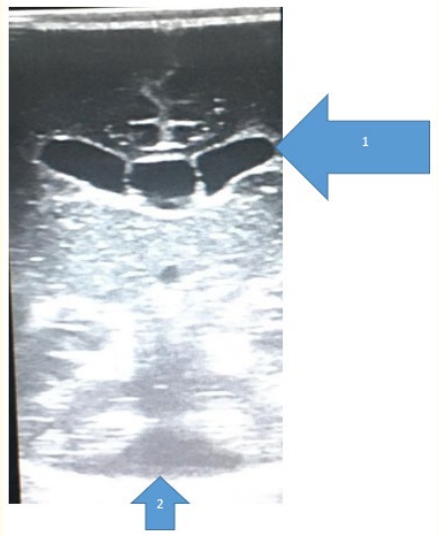

Figure 3: *The arrow 1; points to lateral ventricle on cranial USS and the arrow 2; points to enlarged posterior fossa cyst on cranial USS.*

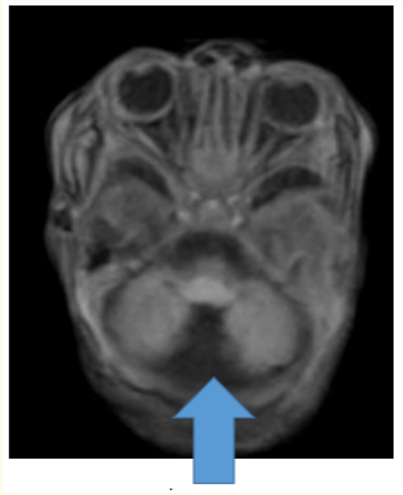

Figure 4: *The arrow above; points to enlarged posterior fossa cyst on T1WMR.*

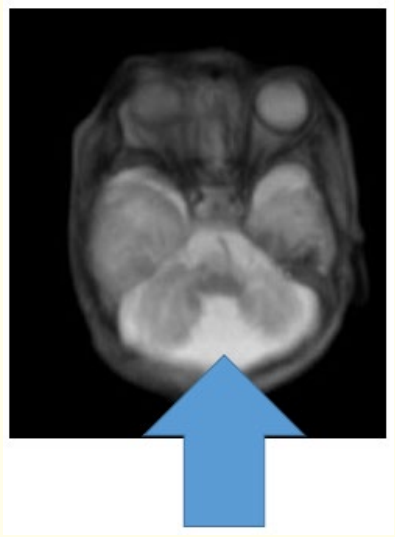

Figure 5: *The arrow above points to posterior fossa cyst on T2WMR.*

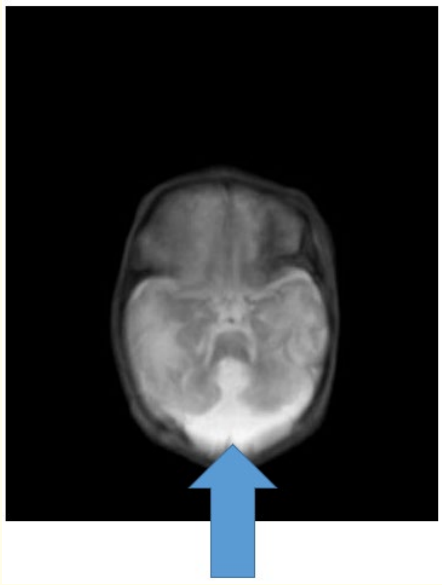

Figure 6: *The arrow above, points to enlarged posterior fossa cyst on MRI-T2.*

Discussion

It's a rare malformation in which the prevalence accounts for 1 out of 25000 - 30000 live births. This malformation is associated with posterior fossa cystic mass, absence of cerebellar vermis and ventriculomegaly [1].

This malformation in children may be associated with intellectual impairment and developmental delays [2].

Some other rare malformation associated with this Dandy Walker include agenesis of corpus callosum and encephalocele [3].

According to Sutton performed the first post mortem and revealed that hydrocephalus is not a must finding in Dandy walker malformation [4,5].

MRI is a special tool for malformations of central nervous system and can as well be used to differentiate from other posterior fossa pathologies [6].

Some of the studies show that few patients of DWM is due to genetic and environmental factors with associated inheritance [7].

Conclusion

This reports highlights the importance of early recognition by clinical grounds and radiological investigations and implementing management of hydrocephalus that was presented with widened of anterior fontanelle.

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