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A New Born With Dandy Walker Malformation- A Rare Presentation.

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ABSTRACT

We report a case of Dandy Walker malformation, associated with multiple congenital anomalies in a newborn. And also various imaging modalities have been done to confirm the diagnosis.

Keywords: Dandy walker malformation, associated congenital anomalies, imaging modalities

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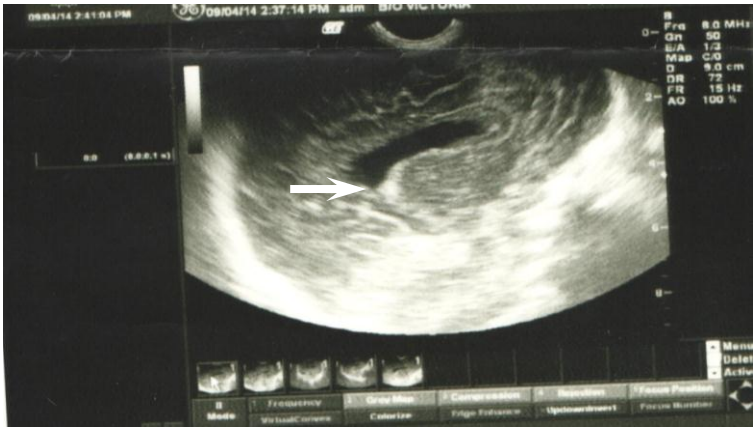
INTRODUCTION

Dandy walker malfomation(DWM) is rare congenital malformation that involves the cerebellum and fourth ventricle. Dandy walker malformation occurs in at least 1 in 5000 live born infants[1].The key components include hypoplasia of cerebellar vermis, cystic dilatation of fourth ventricle and enlargement of posterior fossa[1]. We are reporting an interesting case of Dandy walker malformation.

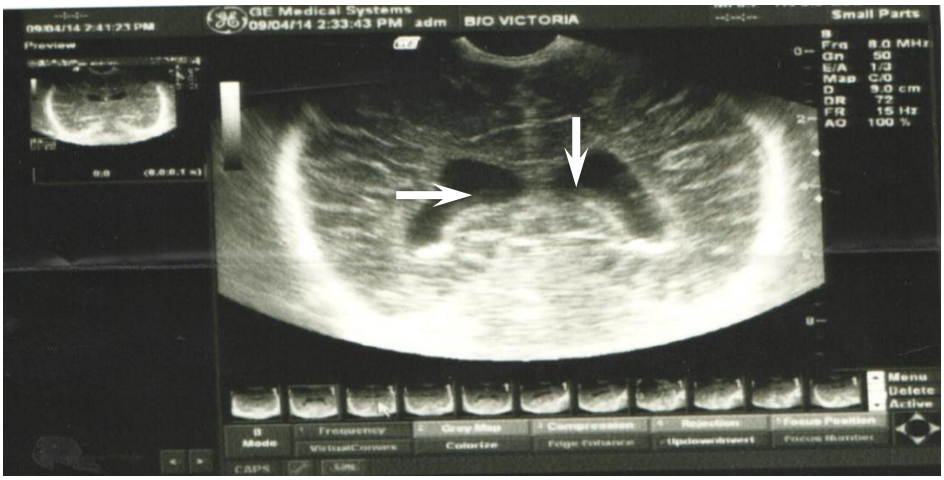
CASE REPORT

18 days old female child born to non consanguineous marriage delivered by LSCS, term, AGA, birth weight- 2.8kgs with past history of neonatal convulsions at day 10 of life.Child was admitted in our hospital for poor feeding, fever and not passed urine for more than 24 hours. On examination child was found to be lethargic with signs of shock. Head to foot examination- macrocephaly, wide open anterior and posterior fontanelles , frontal prominence, microphthalmia of right eye, hypertelorism, low set ears , polydactyly of left finger and polysyndactyly of right toe. Systemic examination revealed cardiovascular system- S1S2 + with systolic murmur, central nervous system-hypotonia of all 4 limbs, Per abdomen and Respiratory system- NAD. Septic workup and RFT was done which was within normal limits. Fluid bolus was given and child passed urine immediately. Child was started on IV fluid maintenance and IV antibiotics. USG cranium was done which showed agenesis of genu and rostrum of corpus callosum as shown in picture 1 and dilated lateral, third and fourth ventricles as shown in picture 2. MRI imaging of brain was done which showed agenesis of genu and rostrum of corpus callosum as shown in picture 3,colpocephaly as shown in picture 4, dilated lateral, third and fourth ventricles as shown in picture 5 and posterior fossa cystic lesion communicating with fourth ventricle as shown in picture 6. ECHO was done which showed small ASD measuring 5 mm left to right shunt with normal LV function as shown in picture 7.

Picture 1: Agenesis of rostrum and genu of corpus callosum



Picture 2: Dilated ventricles (Hydrocephalus)



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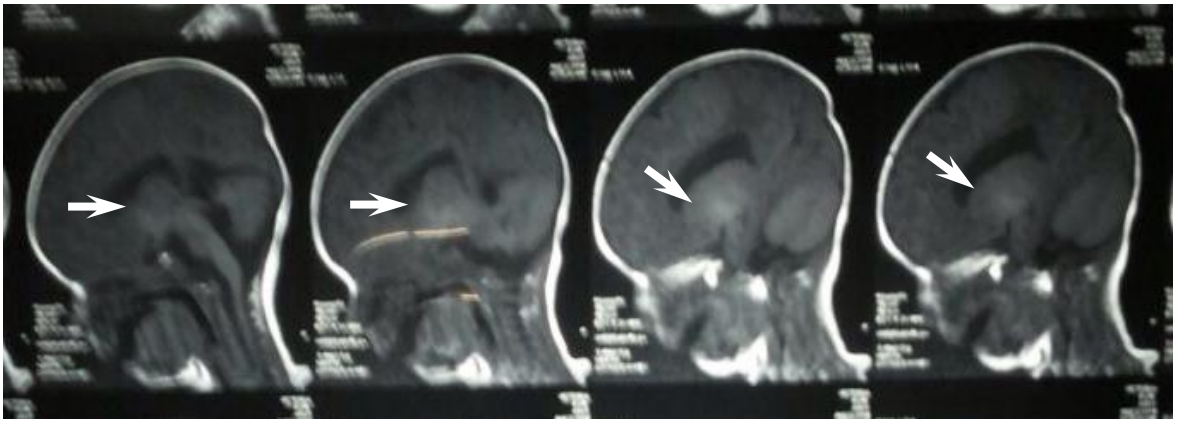
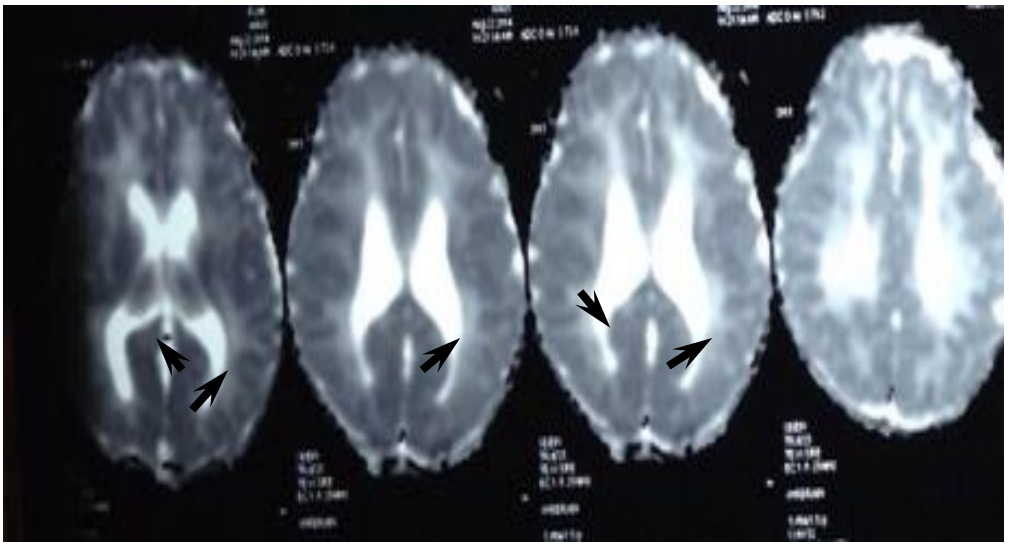
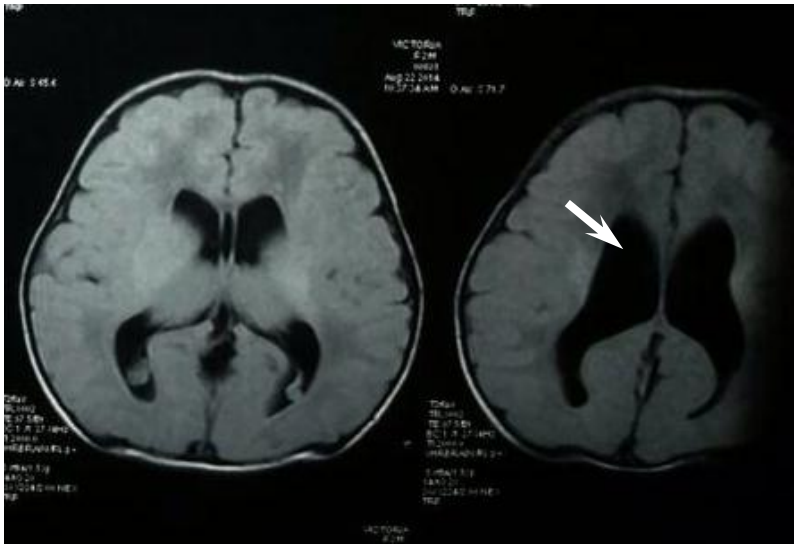
Picture 3: Agenesis of rostrum and genu of corpus callosum



Picture 4 : Colpocephaly



Picture 5: Dilated ventricles (Hydrocephalus)



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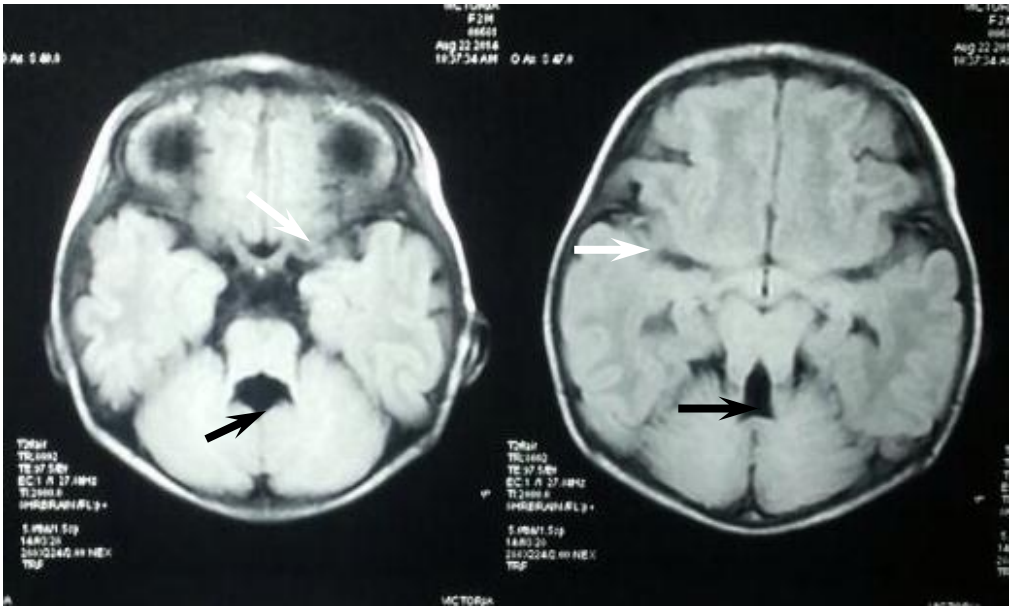
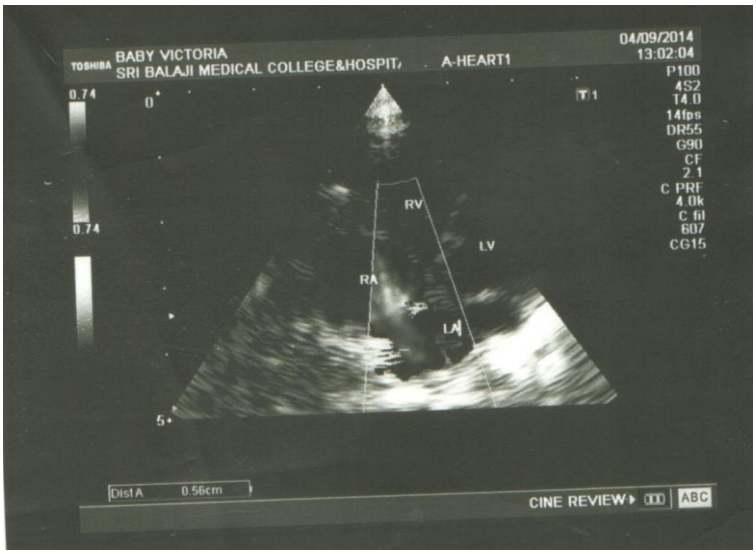
Picture 6: Posterior fossa cyst communicating with fourth ventricle



Picture 7: Small ASD measuring 5mm left to right shunt



These features were suggestive of Dandy walker malformation which is a rare congenital malformation in newborn.

DISCUSSION

Dandy walker malformation was first described in 1887 by Sutton and characterized by Dandy and Blackfan in 1914 and Taggart and Walker in 1942[2]. The components of Dandy walker malformation include complete or partial agenesis of cerebellar vermis, cystic dilatation of fourth ventricle and enlargement of posterior fossa. It is usually associated with supratentorial hydrocephalus which should be considered as a complication rather than part of malformation[1,3]. In our child similar features were present.

Dandy walker malformation, Dandy walker variant and mega cisterna magna are believed to represent a continuum of developmental anomalies on a spectrum that has been termed as Dandy walker complex[4,5].

Associated central nervous system abnormalities of Dandy Walker malformation are reported in 70% of children. The commonest malformations are dysgenesis or agenesis of corpus callosum (20-25%), holoprosencephaly (25%), malformation of cerebellar folia (25%). Other least common manifestations of central nervous system include occipital encephalocele, polymicrogyria, heterotropia, colpocephaly and schizencephaly[2,6,7]. In our child CNS malformation present were agenesis of corpus callosum ,colpocephaly and polymicrogyria. Non CNS malformations were reported in 20-33% children which includes oro-facial deformities, cleft lip/palate, congenital heart disease, malformation of urogenital tract, extra fingers/toes (polydactyly) and fused fingers/toes (syndactyly)[2,6,7]. In our child non CNS malformation present were macrocephaly, wide open anterior and posterior fontanelles, frontal prominence, micropthalmia of right eye, hypertelorism, low set ears ,polydactyly of left finger , polysyndactyly of right toe and small ASD.

The clinical manifestations may occur in early infancy which includes slow motor development, apnea, seizures, developmental delay and mental retardation which are highly variable. On examination they tend to have congenital hypotonia[2]. In older children there may be signs of increased intracranial pressure such as irritability, vomiting and seizures. There may be cerebellar signs such as ataxia, nystagmus and in co-ordination.

The diagnosis of Dandy walker malformation can be made by ante natal ultrasound[8]. Though the diagnosis can be made postnataly by USG, it is operator dependent. MRI is usually performed for detailed evaluation of Dandy walker malformation after USG cranium and CT. MRI can best define the relationship between the cyst and the fourth ventricle and it can detect vermian rotation and signs of vermian dysgenesis.

Treatment for Dandy walker malformation generally consists of reducing raised ICT by various shunt procedures and treating the associated symptoms if necessary[9]. Children with Dandy walker malformation may never have normal intellectual development even when the hydrocephalus is treated early. The presence of multiple congenital defects may shorten life span.

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