A CASE OF DANDY WALKER CYST WITH PORENCEPHALY

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ABSTRACT

A 27-year-old gravida two para one mother delivered a term male baby by caesarean section. The baby was referred for enlarged head. This neonate with large head was found to have Porencephalic cyst with dilated Triventricular system. There was a posterior fossa cyst communicating with fourth ventricle. A diagnosis of Dandy Walker Cyst with Porencephaly was made. Ventriculo-Peritoneal shunt was done on the ninth day of life. The baby had an uneventful postoperative period and was subsequently discharged.

Keywords : Dandy Walker Cyst, Porencephaly.

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INTRODUCTION

The association of hydrocephalus, hypoplasia of the cerebellar vermis and posterior fossa cyst was first described by Sutton in 1887[1]. Later in 1914, Dandy and Blackfan described hydrocephalus associated with a posterior fossa cyst and atresia of the formina of Magendie and Luschka[1]. Hart in 1972, and Gardner in 1975 reported that the posterior fossa cyst which replaces the fourth ventricle may not in all cases be secondary to atresia of the outlet formina since patency of these formina was demonstrated in some cases[2]. Hence, the anatomical basis for Dandy Walker cyst is (a) cystic transformation of the fourth ventricle, (b) with or without atresia of the formina of Luschka and Magendie, (c) minimal or maximal dilatation of the cystic fourth ventricle, (d) there may or may not be dysgenesis of the inferior cerebellar vermis[3].

We report a case of Dandy Walker Cyst with porencephaly in the newborn period.

CASE REPORT

A four-day-old male term baby was referred from district hospital for progressive enlargement of head since birth. A twenty-seven year-old gravida two, para one mother delivered the baby by caesarean section. Her antenatal history was normal. Birth weight was 3600 gm. Apgar score was 4 and 7 at one and five minutes. On examination, the baby was tachypnoeic, but not anaemic or cyanosed. The anterior fontanelle was large and wide open. Head circumference: 44cm, (>97th centile.) Chest circumference: 29cm. Length was 49cm. Heart rate 136/minute, respiratory rate 52/minute, regular. No neurological deficit was made out. Other systems were normal.

On admission, investigations revealed that the level of blood glucose and calcium were 1.0mmol/l and 2.56mmol/l respectively. Serum electrolytes showed sodium 149mmol/l, potassium 5.7 mmol/l, urea 3.3mol/L, HB 15.4gm/dl, PCV 38%, RBC 4.32×10^{12}/l, total leucocyte count was 12.2×10^9/l, differential count showed polymorph 71%, lymphocytes 29%, platelet count was 192×10^9/L. Arterial blood gas analysis done on second day of life revealed PH 7.410, PCO_2 33.7mmHg, PO_2 82.5mmHg, HCO_3 20.7 mmol/L, BE -2.3. Serial blood sugar estimations done on the second and third days of life showed 3.6mmol/l and 3.2 mmol/l. Total serum bilirubin done on fourth day of life showed 252µmol/dl. The subsequent estimation of serum bilirubin done on sixth day of life showed 202µmol/dl. Screening for G6PD was normal. Cerebrospinal fluid analysis done on sixth day of life revealed normal cell count, the concentration of protein and sugar was 2.6gm/L and 1.8mmol/dl respectively. Blood group was B positive, Coombs' test was negative. Both mother and child were non-reactive for VDRL.

Chest X-ray was normal. Lateral view of the skull X-ray showed enlargement of the posterior fossa. CT of the brain (Fig 1) revealed hypoplasia of the cerebellar hemispheres and dilated triventricular system. There was a posterior fossa cyst communicating with fourth ventricle through vallecula. Tentorium appeared elevated and like inverted V shape. Besides, there was homogenous, hypodense, sharply marginated area in the left temporal lobe which was communicating with dilated lateral ventricle. No midline shift or mass effect was noticed (Fig. 2).

The baby was treated symptomatically with head box oxygen, intravenous infusion of 10% dextrose and continuous phototherapy. Intravenous ampicillin 100 mgm/kg/day and gentamycin 2.5mgm/kg/dose in divided doses were administered.

On the ninth day of life, a ventriculo-peritoneal shunt was done. The baby had an uneventful postoperative period and was subsequently discharged.

DISCUSSION

The exact cause of Dandy Walker cyst is unknown. Developmental anomaly, infection and vascular occlusion are among possible causes. Experimental data are suggestive of genetic, chemical or multifactorial causes. An autosomal recessive trait has been implicated in some of the familial cases.

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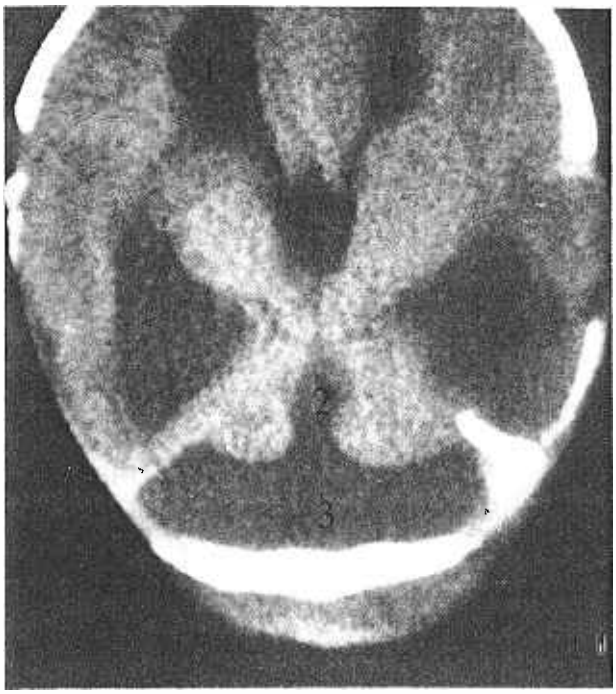
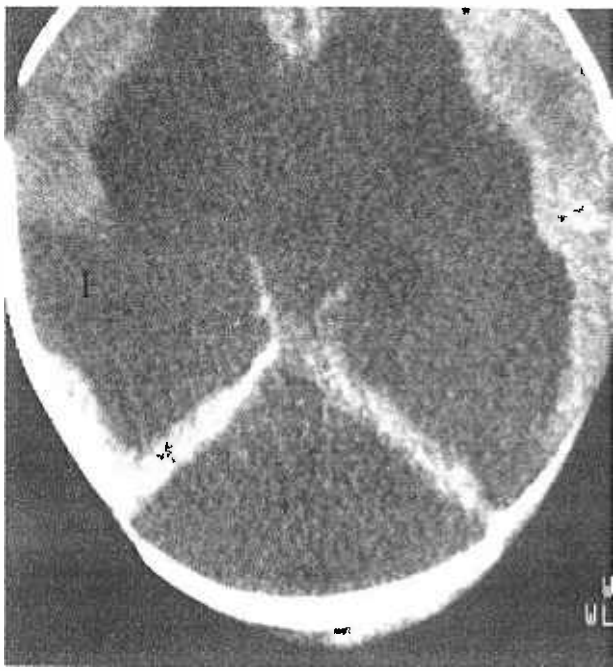
Fig 1 - CT Brain showing hypoplasia of the Cerebellar Hemispheres and dilated triventricular system. There was also a posterior Fossa Cyst communicating with fourth ventricle through vallecula.



Though Dandy Walker cyst is a well-known anomaly, its pathogenesis is obscure[1,3,4]. According to Atresia theory, the cystic transformation of the fourth ventricle is presumed to inhibit cerebellar development. While Gardner and his associates believed that impermeability of the (rhombic) roof of the fourth ventricle and persistent hydrocephalus was the cause for maldevelopment of the cerebellum, it may be associated with other cerebral abnormalities like holoprosencephaly, cerebral gyral abnormalities, malformations of the inferior olive, cerebellar folial anomalies, heterotopias, aqueductal stenosis, microcephaly, agenesis of corpus callosum, absence of bilateral bulbar pyramidal tract, occipital encephalocele. Other associated non-cereberal abnormalities are polydactyly, syndactyly, cleft palate, Klippel Feil syndrome, Cornelia De Lange Syndrome, bilateral polycystic kidney, congenital renal cyst, cardiac anomalies, diaphragmatic hernia, etc.

Fig 2 - Agenesis of the left temporal lobe with extension of the dilated lateral ventricle into it.



Porencephaly is a fluid filled cavity (presumably cerebrospinal fluid) in the brain. It occurs either due to developmental defect (Schizencephalic) or more commonly due to acquired (encephaloclastic) causes like neonatal hypoxia, infection or vascular occlusion. Porencephaly may or may not be communicating with one portion of the lateral ventricle. When this fluid filled cavity is communicating with ventricles and also associated with hydrocephaly, it is termed as a porencephalic cyst.

Porencephalic cyst has to be differentiated from the following conditions like cystic tumours and abscess which show enhancement of the contrast following intravenous contrast study, while in porencephaly, no such contrast enhancement is seen due to the absence of the vascular capsule. Arachnoidal cysts have special areas of predilection and distort the ventricular system by their mass effect. There may be thinning of the overlying contiguous bone[5]. Lipomas and epidermoids have lower attenuation value than the cerebrospinal fluid[6]. Small areas of calcification may be an associated feature.

In this neonate, CT of the brain was useful in diagnosing Dandy Walker cyst with a porencephalic cyst in the left temporal lobe in addition to the other features described earlier.

The association of porencephaly with Dandy Walker syndrome is not common. In the literature, Asai et al studied Dandy Walker syndrome in thirty-five patients over a period of twenty-three years. In that study, only one patient presented with polyporencephaly with Dandy Walker syndrome. However, 94% of them had hydrocephalus at the time of diagnosis. Raymond Sawaya et al observed that only one infant had a porencephalic cyst associated with agenesis of corpus callosum in their series of twenty-three cases of Dandy Walker syndrome[8].

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