A CASE OF DANDY-WALKER SYNDROME

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Dandy-Walker Sendromu Olgusu

ÖZET

Dandy-Walker malformasyonu embriyogenez sırasında posterior fossa'ya dördüncü ventrikülün kistik genişlemesi şeklindedir. Orta derecede solunum güçlüğü olan bir günlük kız yenidoğan hastanemize geldiğinde solunum sayısı 86/dakika idi. Göz hareketleri ve fundus muayenesi normal bulundu. Beyin omurilik sıvısı incelemesinde şeker ve protein normal düzeylerde, eritrosit ve lökosit yoktu. Kranial ultrasonografi, kompüterize tomografi ve magnetik rezonans görüntüleme sonucunda dördüncü ventrikülde kistik genişleme gösterildi.

Anahtar kelimeler: Dandy-Walker malformasyon, kompütarize tomografi, magnetik rezonans görüntüleme

SUMMARY

The Dandy-Walker malformation consists of a cystic expansion of the fourth ventricle in the posterior fossa, during embryogenesis. A one-day-old girl who was transferred in our hospital with mild respiratory distress had a 86/min respiratory rate, on admission. Eye movement and fundus examination were normal. Cerebrospinal fluid had normal protein and glucose levels with no pleocytosis. Cranial ultrasonography, computed tomography and magnetic resonance imaging showed cystic expansion of the fourth ventricle.

Key words: Dandy-Walker malformation, computed tomography, magnetic resonance imaging.

INTRODUCTION

The Dandy-Walker malformation consists of a cystic expansion of the fourth ventricle in the posterior fossa, which results from a developmental failure of the roof of the fourth ventricle during embryogenesis. Approximately 90% of patients have hydrocephalus, and a significant number of children have associated anomalies, including agenesis of the posterior cerebellar vermis and corpus callosum (1,2).

Most children have evidence of long-tract signs, cerebellar ataxia, and delayed motor and cognitive milestones, probably due to the associated structural anomalies. Lesions of malformations of the posterior fossa leading to hydrocephalus are Dandy-Walker syndrome, and the Chiari malformation (1,2).

CASE REPORT

A 1-day-old girl was transferred for evaluation of mild respiratory distress. The newborn infant was delivered by spontaneous vaginal delivery at 39 weeks' gestation to a 30-year-old woman. She weighed 3160 g at birth and had an Apgar score of 7/8 at 1 and 5 minutes.

The neonate appeared healthy until the first 15 hours of life. During the next five hours, she developed irritability, irregular respiration and tachypnea. On admission, the neonate's temperature (36.7°C) and heart rate (124/min) were normal. Respiratory rate was 86/min. Her height (50.5 cm), weight (3160 g) and head circumference (35 cm) were between the 50th and 75th percentile. Eye movements and fundus examination were normal. Complete blood count, renal and liver

function tests, and electrolytes were within normal limits. The cranial, chest and abdominal roentgenograms, and abdominal ultrasonography and echocardiography showed no abnormalities. Blood, cerebrospinal fluid had normal protein and glucose levels with no leucocytes and erythrocytes.

On day two cranial ultrasonography was performed (Fig 1), on day five cranial computed tomogram (Fig 2) and on day 35 magnetic resonance imaging (Figs 3,4) were obtained.

During the next five days, there was improvement in her respiratory pattern and clinical status. On day 75, she had a regular respiration, normal growth, and motor and mental development.

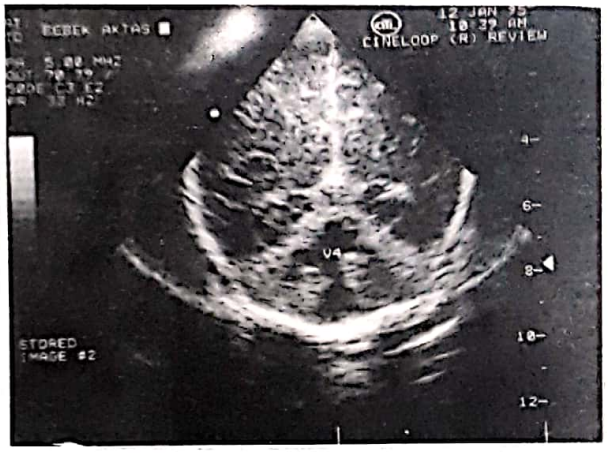

Figure 1. Coronal ultrasonographic image shows enlarged fourth ventricle and hypoplastic cerebellar hemispheres.

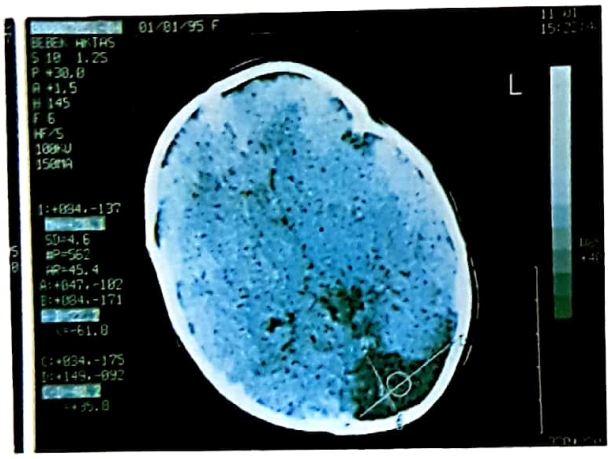

Figure 2. Computed tomographic images show enlarged fourth ventricle and hypoplastic cerebellar hemispheres. The cerebellar vermis is absent.

DISCUSSION

This abnormality consists of a malformation of the forth ventricle and cerebellum. The malformation is a developmental cerebellar defect that originates before embryologic differentiation of the foramina of the forth ventricle, and appears to be result of partial or complete agenesis of the cerebellar vermis and subsequent enlargment of the forth ventricle (3). Cerebellar hemispheres are variably hypoplastic (4). Obstruction of normal flow of cerebrospinal fluid results in dilatation of third and lateral ventricles in most cases, but these later structures are not enlarged as excepted. In milder cases, fourth ventricular dilatation is less stricking and associated cerebellar changes less dramatic (5). In our case the enlargement of forth ventricle and cerebellar changes are milder. The syndrome may be associated with maternal exposure to isotretinoin in the first trimester. Familial forms of Dandy-Walker syndrome have been described with autosomal inheritance (3). Our case is not a familial form of Dandy-Walker syndrome.

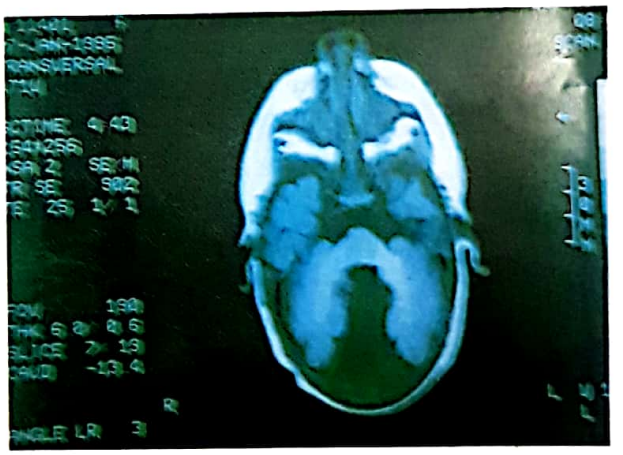

Figure 3 Magnetic resonance images show enlarged fourth ventricle with absence of hydrocephalus.

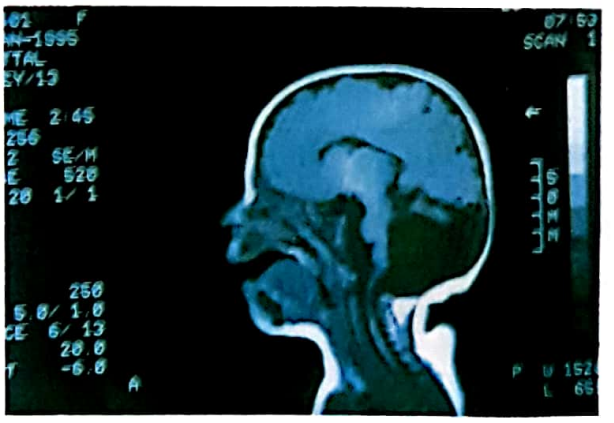

Figure 4. Magnetic resonance images show enlarged fourth ventricle with absence of hydrocephalus

Associated congenital anomalies may be present in half of children (6). Other cranial anomalies, such as agenesis of the corpus callosum, gyral abnormalities, gray matter heterotopias, microcephaly, encephalocele-meningocele, posterior fossa lymphomas and neurocutaneus melanosis (7-9). Numerous nonneural abnormalities include polydactyly, syndactyly, cleft palate, cystic renal disease (Goldstone syndrome) (3-5). In our case, associated congenital abnormalities are microcephaly and nonneural abnormalities such as syndactyly.

Clinical findings become evident during infancy. Eighty per cent of patients become symptomatic by three years of age, with 70% presenting within the first year of life. Hydrocephalus (91%), developmental delay (33%), nystagmus, spasticity, titubation and apnea are common. In our case, also apnea was manifested in the newborn period. The difficulties in older children may be manifestations of increased intracranial pressure and ataxia. Cranial ultrasonography, computed tomography and magnetic resonance imaging best demonstrate the characteristic pattern of enlargement of the fourth ventricle (3,6).

A shunt from the ventricle and/or cyst has removed the enlargement as the primary treatment for Dandy-Walker syndrome. The mortality rate is 24% (6). Gerszten et al (10) who treating the patients with shunts, conclude that there is no relationship between the cerebellar development evident on computed tomography scans and the cerebellar or the intellectual function of children with Dandy-Walker syndrome.

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