# Hydranencephaly Caused By Herpes Simplex Virus Encephalitis
## A Case Report

Etem BEŞKONAKLI, Yıldız YALÇINLAR

Department of Neurosurgery, Ankara Numune State Hospital, Ankara, TÜRKİYE

**Abstract :** Hydranencephaly is a rare anomaly of the central nervous system (CNS) which characterised by absence of the cerebral hemispheres. The etiologic hypotheses are numerous and the causal injuries have been thought to be vascular occlusion, viral encephalitis and genetic abnormalities.

In this report we present a case of hydranencephaly and suggest the etiologic factor of this anomaly is Herpes Simplex Virus Encephalitis (HSV-Encephalitis).

**Key Words :** Computed Tomography, HSV-Encephalitis, Hydranencephaly

## INTRODUCTION

Hydranencephaly is a congenital disorder characterised by absence of the cerebral hemispheres but with intact cranial vault and meninges (3,4,5, 10,13,14,23,24).

The cerebral hemispheres are replaced by cerebrospinal fluid (CSF) and necrotic debris inside a thin walled sac. There is usually preservation of the posterior fossa structures, the brain stem, the falx cerebri, and the basal temporal and occipital regions which are fed by the posterior circulation.

There are various hypotheses about the cause and pathogenesis of the disease. Vascular anomaly (4,5, 10,12,13,17,23,28), postencephalitic state (6,7,9,16,18, 20,21,25,26), and genetic anomalies (15,19) are mostly blamed.

We report a case of hydranencephaly in a 5 month-old female baby whose development had previously been normal. Although hydranencephaly is known to be a congenital anomaly (3,5,12,20, 21,26) in our patient was late in appearing. In our opinion the cause was HSV-Encephalitis. This is the only case which was progressively followed by computed tomography (CT) in the encephalitic and hydranencephalic phases.

## CASE REPORT

A 5 month-old girl was seen at the Children's Hospital because of irritability, fever and lethargy. She was said to be doing well 15 days before the developed progressive symptoms. The family history, pregnancy and delivery, had no abnormality.

At the physical examination her general condition was poor, and there were dehydration, dispnaea with rales in the lungs. The temperature was 39.2°C. Body measurements, head and chest circumference were normal pathological findings at neurological examination included bilateral optic atrophy, neck stiffness and hyperactivity in all deep tendon reflexes.

X-Ray of the skull showed no abnormality but CT showed at left temporal mass lesion which was hyperdense with a surrounding hypodense area (Figure 1). When the child was admitted to the

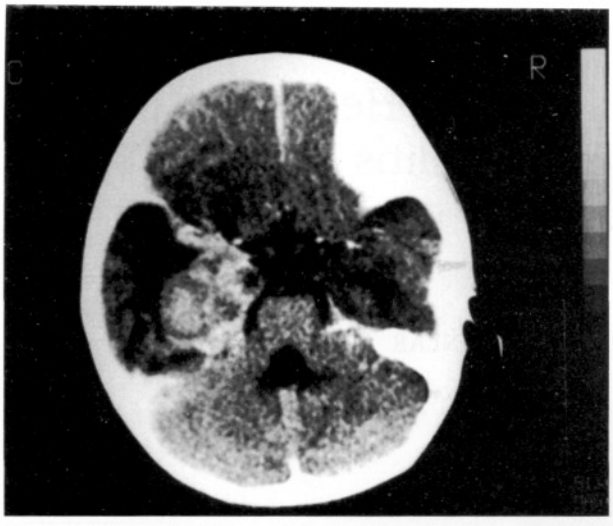

*Fig. 1 : CT showed a left temporal mass lesion (at 5 months of age).*

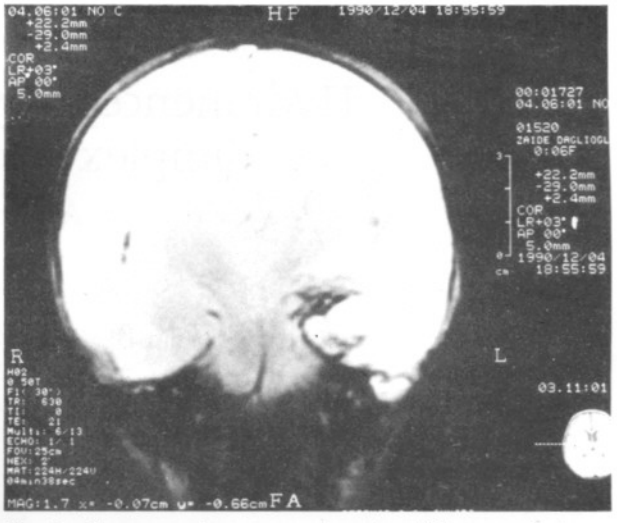

*Fig. 2 : The magnetic resonance imaging of the same lesion.*

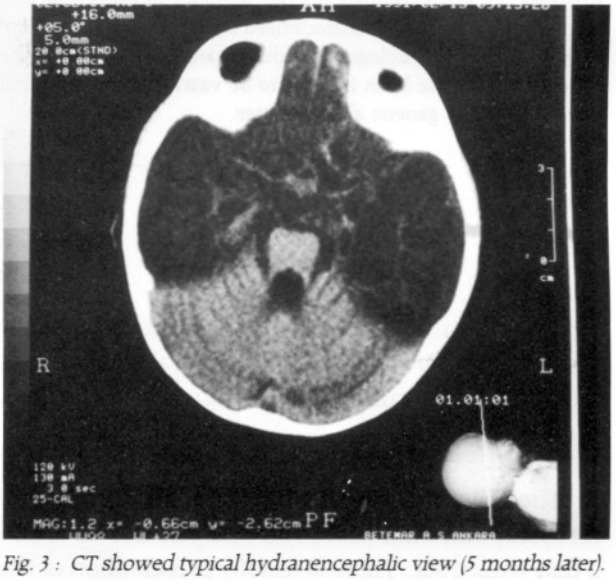

*Fig. 3 : CT showed typical hydranencephalic view (5 months later).*

hospital CSF examination were normal and the parents' serum IgG levels for HSV type-1 and type-2 were positive, the patient's blood IgG level for HSV type-1 was positive. No Igm levels HSV and No Igm and G levels for other viruses were detected.

Electroencephalogram (EEG) showed voltage suppression in all traces and slow theta waves and slow spike waves in bursts (specific for HSV-Encephalitis). After antibiotic and anticonvulsant therapy her general and neurological condition improved. After one week, oral feeding was started and at that time the left temporal lesion was confirmed by magnetic resonance imaging (MRI) (Figure 2). Although antiviral agents such as acyclovir was not used the CT after three months (at 8 months of age) showed that the lesion in the left temporal lobe had disappeared and showed absence of cerebral hemispheres but intact posterior fossa structures and brain stem. The ventricles were normal and there was typical hydranencephaly (Figure 3). Angiography could not be performed. At that SPECT with 4mci Tc HMPAO intravenously showed loss of perfussion in the frontal, temporal, parietal and occipital regions. EEG showed loss of perfussion in the frontal, temporal, parietal and occipital regions. EEG showed flat tracing without evidence of cerebral activity. Digital Subtraction Angiograply (DSA) relieved that the vascular structures were normal. At that time in the control CSF examination, only the protein content was to be high (111 mg/ml). In the Control Blood Serum examination; The parents' and the patient's Ig levels for HSV were positive but have reduced titres. Culture and isolation for HSV were not performed. The patient's condition is progressing and she is being followed.

## DISCUSSION

Hydranencephaly was first described by Cruveilher (1892) as "anencephalie hydrocephalique" or "Hydroanencephalie" (23). crome and Sylvester reviewed the disease and the defined Hydranencephaly as a congenital condition (3) characterised by absence of the cerebral hemispheres

with relatively normal-sized cranium and preservation of the posterior fossa structures, brain stem, basal ganglia and occasionally the basal temporal and occipital regions. The cerebral hemispleres are replaced by a thin-walled sac containing CSF and necrotic debris (3,5,7,13,14,22,23,27,28). There is symmetrical destruction of gray and white matter fed by the anterior and middle cerebral arteries. Why che condition only seen in the cerebral hemispheres is not clear.

For diagnosis, direct radiological examination is nonspecific and ultrasonography, CT, MRI, EEG and evoked potential studies should be performed (1,2,5,8,13,14,24,27,29).

For a long time, it was thought that the condition was congenital and the etiology and pathogenesis of this disease is unclear (3,5,7,12,20,21,26) but numerous hypotheses have been suggested. These are vascular occlusion postencephalitic state and genetic abnormalities.

It has been suggested that occlusion of the supraclinoid internal carotid artery before the 24th week of gestation results in ischaemia, oedema, autolysis, debris, cavity and loss of the cerebral hemispheres respectively (4,5,10,12,17,23,28). However some reports suggest that occlusion of the vascular structures is due to temporary spasm or compression rather that direct occlusion (13). Beside vascular disorders, some postencephalitic conditions especially HSV-Encephalitis have been blamed (6,7,9,11,15,16,18,20,21,25,26). Congenital HSV infections have a high mortality and morbidity and if seen during delivery or immediately post-partum, in utero infestations should be considered. For in utero transmission a primary maternal episode carries more risk than recurrent maternal episodes. The majority of cases of maternal HSV transmission occur during delivery by direct contact (11,20).

Patients with HSV infection may varying degrees of severity ranging from asymptomatic to systemic infection including CNS, skin, eye and mucosal anomalies. The most frequent CNS anomalies are microcephaly, epilepsy, psychomotor retardation and hydranencephaly. If dissemination occurs after birth the most frequent initial symptoms are nonspecific such as fever, lethargy, irritability and poor feeding (20). Skin lesions occur in about 50 to 92 % of nonates early in the course of the disease (11,20). Herpes specific IgM or IgG activity is positive but less helpful than a culture because of placental transfer of maternal Ig antibody (11,18,20).

At differential diagnosis, on CT appearance there are some similar entities which must be differentiated such as massive subdural effusion, hydrocephalus, alobar holoprosencephaly and postinfarctive entities (5). Hydrocephalus is the most common. Present condition differs in that (1) there is no ependyma, (2 plain skull radiography is usually normal, (3) on EEG there is no biological activity, (4) ventricles are usually normal, (5) visual evoked response (VER) is negative, (6) no improvement is seen with shunt procedures.

In summary it seems reasonable to think that hydranencephaly is the final morphological result of massive destruction caused by a variety of different etiologic agents (23).

Finally; examination of our patient confirmed typical hydranencephalic findings. We suggest the etiologic factor was HSV-Encephalitis. Although the condition was thought to be congenital the patient's symptoms appeared late and encephalitic and hydranencephalic stages were observed by CT.

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**Correspondence** : Dr. Etem Beşkonaklı
Mertler Sokak 41/6
Beştepe-Ankara, Türkiye

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