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HYDRANENCEPHALY SECONDARY TO CONGENITAL TOXOPLASMOSIS

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Hydranencephaly is devastating CNS malformation consisting of almost complete absence of the cerebral hemisphere. It can be developed as a consequence of intrauterine infections (such as toxoplasmosis and cytomegalovirus), other gestational insults, or acquired cerebral infarction associated with extensive meningitis, widespread intracerebral hemorrhage or ischemia. In this paper, we report a patient with hydranencephaly secondary to congenital toxoplasmosis. In the present case, affecting the cerebellum and the brain stem, and stopping the enlargement of head circumference with CSF division suggest that; (1) in the cases with hydranencaphaly secondary to congenital toxoplasmosis, progressive obstructive hydrocephalus may be associated with hydranencaphaly, and this condition may be severely affected hydranencephaly, (2) in cases with hydranencephaly and/or hydrocephalus, CSF diversion may relieve in both conditions.

Key words: *Hydranencephaly, toxoplasmosis.*

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Kongenital Toksoplazmozise Sekonder Gelişen Hidranensefali

Hidranensefali beyin hemisferlerinin tamamına yakın yokluğu ile oluşan bir SSS malformasyonudur. İntrauterin enfeksiyonların (toksoplazmosis ve sitomegalivirüs gibi), diğer gestasyonel hasarların; ya da ağır menenjit, yaygın intraserebral hemoraji veya iskeminin birliktelik gösterdiği edinsel serebral enfarktın bir sonucu olarak gelişebilir. Bu makalede kongenital toksoplazmozise sekonder gelişen bir hidranensefali olgusu sunulmaktadır. Serebellum ve beyin sapının da etkilendiği olguda BOS divizyonu ile hastanın baş çevresinin büyümesinin durması; (1) bu olgulara ilerleyici obstrüktif hidrosefalinin eşlik edebileceğini ve bu durumun mevcut hidranensefaliyi ağırlaştırabileceğini, (2) hidranensefali ve/veya hidrosefali olgularında BOS divizyonu ile rahatlama sağlanabileceğini göstermektedir.

Anahtar kelimeler: *Hidranensefali, Toxoplasmosis.*

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The term *hydranencephaly* encompasses several conditions that result in the extensive replacement of brain by the cerebrospinal fluid (CSF). This may be caused by failure of normal brain development or from an intrauterine disorders that destroys the brain paranchyma. The cranium is intact and therefore does not implicate anencephaly or hydranencephaly. Because of the nearly complete lack of cortical tissue, the children are developmentally delayed from a very early age.[1-3]

It is important to distinguish hydranencephaly from the more common condition of congenital hydrocephalus, which carries a much better prognosis with treatment. In this paper, we report a patient with hydranencephaly secondary to congenital toxoplasmosis, and the relation between progressive hydrocephalus and hydranencephaly is emphasized.

CASE REPORT

An 11-month-old male patient was referred to the Pediatric Neurology Department with a 5-month history of progressively enlarging head. As we were informed by the mother of the patient and the referring hospital, two months before admission, hydrocephalus diagnosed by computerized tomography (CT) had been noted, and for this reason ventriculoperitoneal shunt was introduced. It was realized that before the gestation of the patient the mother had aborted and not been treated for this condition. During her gestation, she experienced several periodical fever attacks. In the neonatal period the boy's appearance was normal.

On physical examination; motor and mental development was extremely retarded. The child showed grotesquely enlarged head size [head circumference was 52 cm (>98thentile)] with a weight at the 3rdcentile and length at the 10thcentile. He was noted to have a dysfunctional ventriculoperitoneal shunt on occipital area. The anterior fontanel was wide open. In the lower extremities spasticity became apparent, in the upper extremities deep tendon reflexes were hyperactive and both planter responses were extensor. The reaction of both pupils to light was decreased. Fundoscopic examination was revealed chorioretinitis. The cranial nerves were found to be normal. The rest of the physical examination was unremarkable. Chromosomal studies were normal. Cerebrospinal fluid findings were as follows: pressure 250 mm-H_2O, leukocyte count 90/mm^3 with lymphocyte predominance (85 %), protein 150 mg/dL and glucose 70 mg/dL. The CSF and serum specimens were positive for toxoplasmosis IgG antibodies (titers: 1:16 and 1:4, respectively) by the *IgG fluorescent antibody (IgG-IFA) test*, and the *IgM fluorescent antibody (IgM-IFA) test* in CSF was positive at low titers (1:10). Blood and spinal fluid cultures were sterile. *T. gondii* couldn't be isolated form blood and CSF.

On imaging studies, the brain stem was mildly atrophic. Cerebellar vermian hypoplasia was seen. Parenchymal-subependimal multiple calcifications at the diencephalic-multicentric areas and incomplete falx cerebri with cortical-subcortical structures of the occipital lobe were shown (Figures 1A and 1B). The cerebral hemispheres appeared nearly completely replaced by CSF. The thalami were preserved (Figures 2A, 2B and 2C). Electroencephalogram disclosed diffusely slow activity. The visual evoked responses and electroretinogram were abnormal, but brain stem evoked responses were preserved.

On the basis of these findings, the diagnosis ofhydranencephaly secondary to congenital toxoplasmosis was made. Probably, it was associated with progressive hydrocephalus. Intensive physical therapy was applied. Ventriculoperitoneal shunt was removed. For the first 6 mo, oral pyrimethamine (2 mg/kf/24 hr for 2 days, then 1 mg/kg/24 hr for 2 mo, then 1 mg/kg/24 hr Monday, Wednesday and Friday), sulfadiazine (100 mg/kg loading dose, then 100 mg/kg/24 hr in two divided doses), and calcium leukovorin (10 mg/24 hr Monday, Wednesday and Friday) were planned and started. Prednisone (1 mg/kg/24 hr orally in divided doses) was administrated because of active chorioretinitis involved macula. After two weeks CSF protein was found to be normal and ventriculoperitoneal shunt was introduced again. The patient was discharged in a stable clinical condition at the end of hospitalization period. In the second 6 mo this regimen was

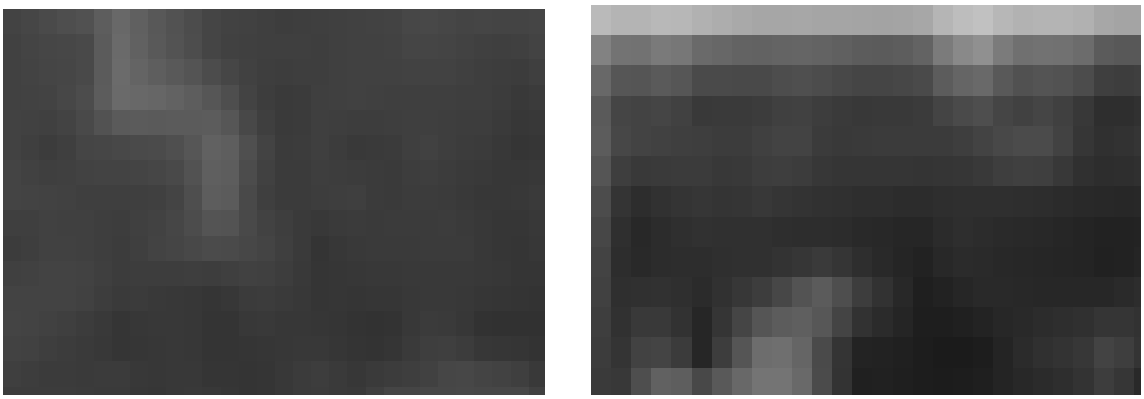

Figure 1. Hydranencephaly in an 11-month-old child. A lower axial CT scan showing he multicentric paranchymal-subependimal calcifications at the diencephalic area (A) and an upper axial CT scan showing incomplete falx cerebri with cortical-subcortical structures of the occipital lobe (B).

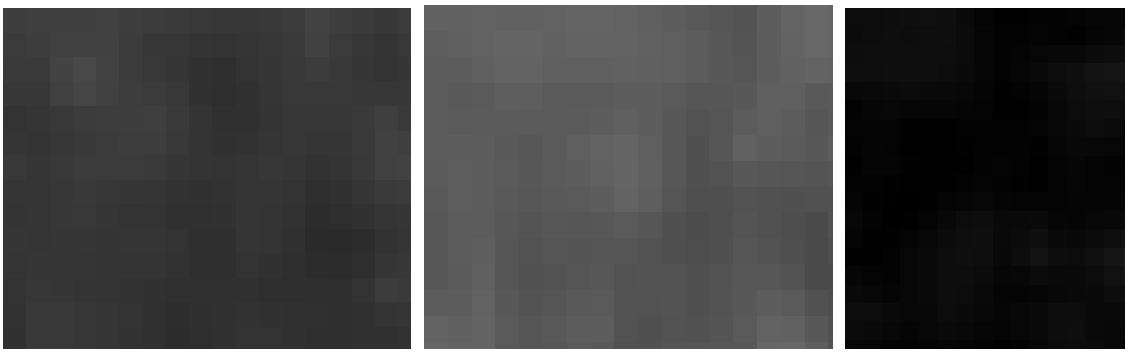

Figure 2. MR images in the same patient **(A)** T1-weighted sagittal, and **(B)** midsagittal planes showing the cerebral hemispheres appeared nearly completely replaced by CSF, and preserved thalami. **(C)** T2-weighted image showing incomplete falx cerebri with cortical-subcortical structures of the occipital lobe.

continued in alternate months with spiramycin (50 mg/kg twice a day). He had no severe complaint (including the enlargement of head circumference), and his retinal lesions were remarkably regressed during 10 months of follow-up.

DISCUSSION

The pathology of hydranencephaly implicates at least five different pathogenetic mechanisms[4,5]: (1) Some authorities have argued that hydranencephaly is a type of hydrocephalus that runs its course in utero, or progressive obstructive hydrocephalus may cause hydranencephaly if left untreated. (2) In other instances, hydranencephaly can be the consequence of intrauterine infections (such as toxoplasmosis and cytomegalovirus) or other gestational insults. (3) In other cases, the condition can represent a genetically determined defect in vascular ontogenesis or can be an outcome of vascular occlusion of both internal carotid arteries or their main branches. A proliferative vasculopathy with an autosomal recessive inheritance has also been described. (4) A few cases appear to be caused by defects in embryogenesis and subsequent cellular migration, resulting in schizencephaly and cortical agenesis. (5) Hydranencephaly may develop in neonates and older infants after widespread cerebral infarction associated with extensive meningitis, intracerebral hemorrhage, or ischemia.

In our cases, the cause of hydranencephaly was assumed to be congenital toxoplasmosis. The cerebellum and the brain stem were also affected by this process. Laure-Kaminonowska et al [5] reported that the damaging process of toxoplasmic encephalitis on newborn brains resulted in hydranencephalus finally. According to these authors, proliferation of subependimal glia and blocking the pathways of the

cerebrospinal fluid circulation may have played in this process. In the cases with hydrocephalus, excessive pressure with in the lateral ventricles destroys the midline structures and reduces the cerebral mantle to a thin membrane. It is sometimes a diagnostic problem to differentiate hydranencephaly from severe hydrocephalus.[6,7] In practice, this distinction may be purely academic because CSF diversion is dictated by an abnormally enlarged head in infants with either entity.[8] We observed that enlarging head circumference of the patient was stopped since effectively

CSF diversion. In the present case, affecting the cerebellum and the brain stem, and stopping the enlargement of head circumference with CSF division suggest that; (1) in cases with hydranencaphaly secondary to congenital toxoplasmosis, progressive obstructive hydrocephalus may be associated with hydranencaphaly, and this condition may be severely affected hydranencephaly, (2) in cases with hydranencephaly and/or hydrocephalus, CSF diversion may relieve in the both conditions.

In order to prevent CNS malformations secondary to embryofetal infections (including toxoplasmosis), in suspected cases pregnant women should be investigated for these infections as well as other gestational insults.

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