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## Cerebral Hemiatrophy: an Associated Finding in an Epileptic Child

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Cerebral hemiatrophy is a rarely occurring condition of different etiologies that can be regarded as the final stage of a number of different disease processes. It is characterized by a marked asymmetry of the cerebral hemispheres. A 12 year old girl with history of epilepsy since infancy and psychomotor delay presented in status epilepticus, developed marked cerebral edema, bilateral

uncal herniation and bilateral infarcts of the posterior cerebral artery territories. Autopsy findings revealed left cerebral hemiatrophy as an incidental finding. The clinicopathologic features and classification of this entity are discussed.

*Key words: Cerebral hemiatrophy, Epilepsy, Seizures*

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Cerebral hemiatrophy is a rarely occurring condition of different etiologies that can be regarded as the final stage of a number of different disease processes (1) among them, epilepsy. It is characterized by a marked asymmetry of the cerebral hemispheres (1). Some cases have been associated with mesial temporal sclerosis and a history of childhood febrile seizures (2). The clinical symptoms mostly consist of seizures, contralateral hemiplegia, hemiparesis, and mental retardation (3). A 12 year old girl with history of epilepsy since infancy, psychomotor delay and associated cerebral hemiatrophy is presented and the clinicopathologic features and classification of cerebral hemiatrophy discussed.

### Case Report

A 12 year old female patient with history of seizures and psychomotor delay was admitted to a private Hospital with the complaints of exacerbation of the seizures, pharyngitis and otitis media. After being discharged home, the patient remained hypoactive, complaining of headaches and weakness, until a month later when she was brought again to the emergency room with seizures and dehydration. A head CT scan showed cerebral edema with midline shift. The patient was admitted to the Intensive Care Unit, where she was orotracheally intubated to protect

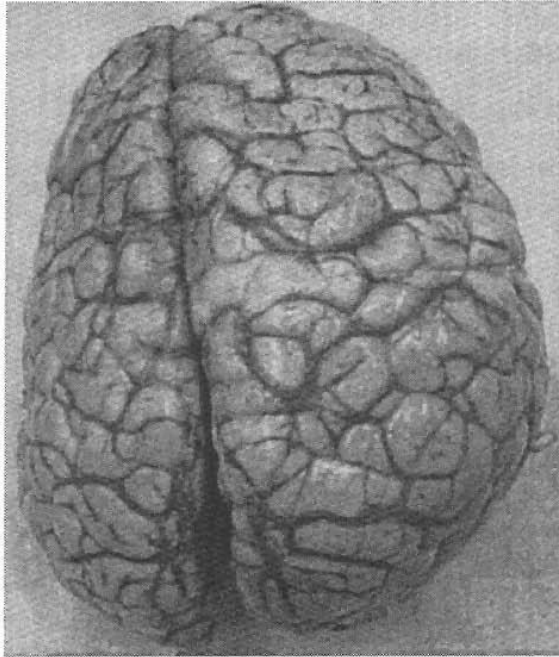
the airway in view of her deteriorating neurological status, and to prevent an increase of intracranial pressure. During her hospital stay, she became comatose, with absent corneal reflexes and no response to verbal or painful stimuli. She also developed diabetes insipidus and required support with dopamine and epinephrine drips, temperature control and 1-deamino-8-arginine vasopressin (DDAVP). A head CT scan revealed brain swelling, transtentorial herniation, right middle cerebral artery infarct and thrombosis of the circle of Willis. A day after admission, the physical exam of the patient was compatible with brain death with an isoelectric electroencephalogram. She presented hemodynamic instability requiring higher doses of epinephrine and dopamine, as well as potassium and magnesium to correct the electrolytic disturbances. After the neurologist confirmed the diagnosis of brain death, she was declared dead a day after admission. An autopsy was requested by her primary physician and a permit granted by the parents.

Postmortem examination of the brain revealed marked edema and asymmetry of the cerebral hemispheres; the left smaller than the right. (Figure 1). The fixed brain weighed 1,630 gm, when the expected is 1,351 gm for her age. At the base of the brain, there was evidence of marked bilateral herniation of the unci. The circle of Willis was grossly unremarkable with no evidence of thrombosis. The cerebellum and the brainstem were symmetrically disposed. Serial coronal sections of the cerebrum accentuated the cerebral edema and the asymmetric cerebral hemispheres. The right cerebral hemisphere was larger than the left, and presented focal effacement of the demarcation between the white and gray matter, and compression of the right lateral ventricle. The hippocampi were not

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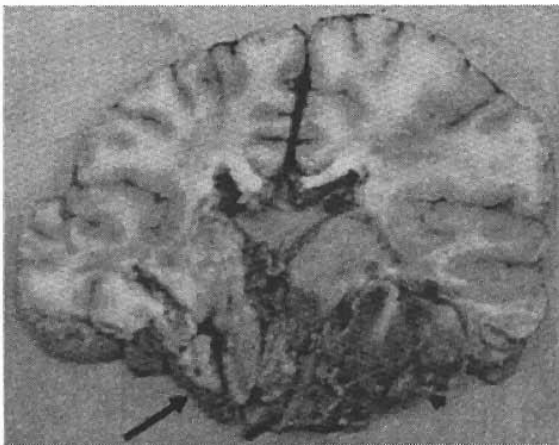
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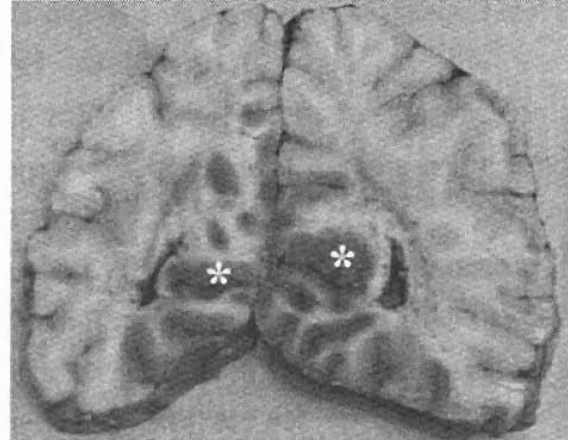


**Figure 1.** Note the marked cerebral asymmetry and flattening of the gyri in both hemispheres (edema).

identified due to bilateral uncal herniation (Figure 2). The ventricular system was asymmetric, with a slit like right lateral ventricle. Both territories of the posterior cerebral arteries presented a gray dusky discoloration with small hemorrhages within the parenchyma and softening (Figure 3). The ependymal lining was smooth and dull. Sagittal



**Figure 2.** Note cerebral asymmetry, edema and uncal herniation (arrow) with secondary hemorrhagic infarcts (arrow head).



**Figure 3.** Note the recent infarcts (\*) in the region of the posterior cerebral arteries.

sections of the brainstem and cerebellum showed no abnormalities.

Microscopic sections of the cerebral cortex showed marked acute ischemic neuronal injury. The cerebellum had loss of the Purkinje cells and necrosis of the granular cell layer. The areas in the territories of the posterior cerebral arteries revealed a recent infarct with edema, ischemic neurons and small hemorrhages within the parenchyma. The ependymal lining had focal disruption. Other significant autopsy findings included a pituitary infarction and pulmonary edema.

## Discussion

Cerebral hemiatrophy is a rarely occurring condition that can be regarded as a final stage of a number of different disease processes (1). As the name implies, it is characterized by a marked asymmetry of the cerebral hemispheres (1). The clinical symptoms mostly consist of seizures, contralateral hemiplegia, hemiparesis, and mental retardation (3). The seizures could be either generalized tonic-clonic or partial (4,5). Radiological findings include unilateral loss of cerebral volume and associated compensatory bone alterations in the calvarium, such as ipsilateral calvarial thickening and overgrowth of the sinuses (3). Clinical studies have documented that prolonged and recurrent seizures may lead to neuronal damage and loss, with subsequent synaptic reorganization (6,7,8). Some investigations support the view that cerebral hemiatrophy is the result of postictal damage, since the severity and distribution of neuronal necrosis in the immediate postictal state and cases with established cerebral hemiatrophy are strikingly similar (9). Other studies

have associated the condition to perinatal cerebral injury, which lead to lack of cerebral development (3).

Voskamper and Schachenmayr proposed a classification according to pathogenesis and etiology (1). Those cases that demonstrated symptoms of convulsions, hemiparesis and mental retardation since birth were classified as primary cerebral hemiatrophy. These were associated with cerebral malformations or ischemic brain lesions due to perinatal asphyxia or birth trauma. Secondary cerebral hemiatrophy was used for those cases which did not have cerebral or cerebrovascular malformations, lack a relevant disease history during pregnancy or birth, and show a widely normal development within at least the first four weeks of life. In the classification of Dix et al, cases were divided into those associated with MR imaging (MRI) mesial temporal sclerosis findings and a history of childhood febrile seizures, and those cases that were not (2). Most of those associated to the MRI findings correspond to the secondary cerebral hemiatrophy group in the Voskamper and Schachenmayr classification. Those cases without MRI findings are consistent with those classified as primary cerebral hemiatrophy. Neuropathological changes in hemiatrophy vary according to the pathogenesis. Cases with history of a perinatal insult demonstrate ulegyria, subcortical cystic changes or laminar cortical necrosis (1). On the other hand, cases of secondary cerebral hemiatrophy show widespread depletion of cortical neurons, gliosis and cerebellar necrosis (1).

Postmortem examination frequently fails to determine the etiology of the hemiatrophy. In our case, cerebral hemiatrophy was most likely associated to her epileptic condition. In addition, there are cases that have a combined pathogenesis. It is still controversial whether seizures are the cause or the consequence of this condition. Unfortunately, this child developed marked cerebral edema secondary to her status epilepticus, and bilateral uncal herniation leading to bilateral infarction of the posterior cerebral artery territories and death.

### Resumen

La hemiatrofia cerebral es una lesión rara de diversas etiologías que podríamos considerar como el desenlace

final de un sinnúmero de enfermedades. Se caracteriza por la presencia de una asimetría marcada de los hemisferios cerebrales. Una niña de 12 años de edad con historial de epilepsia desde su infancia y retraso sicomotor presentó con estatus epiléptico, desarrolló edema cerebral marcado, herniación uncal bilateral e infartos bilaterales en el territorio de las arterias cerebrales posteriores. La autopsia demostró hemiatrofia cerebral izquierda como un hallazgo incidental; se discute la presentación clinicopatológica y clasificación de esta entidad.

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