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MR AND CT IMAGING IN THE DYKE-DAVIDOFF-MASSON SYNDROME

REPORT OF THREE CASES AND CONTRIBUTION TO PATHOGENESIS AND DIFFERENTIAL DIAGNOSIS

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ABSTRACT - Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome is a condition characterized by seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, and mental retardation. These findings are due to cerebral injury that may occur early in life or *in utero*. The radiological features are unilateral loss of cerebral volume and associated compensatory bone alterations in the calvarium, like thickening, hyperpneumatization of the paranasal sinuses and mastoid cells and elevation of the petrous ridge. The authors describe three cases. Classical findings of the syndrome are present in variable degrees according to the extent of the brain injury. Pathogenesis is commented.

KEY WORDS: Dyke-Davidoff-Masson syndrome, brain atrophy, computerized tomography, magnetic resonance image.

Achados radiológicos na síndrome de Dyke-Davidoff–Masson: relato de três casos e contribuição para patogênese e diagnóstico diferencial

RESUMO – Hemiatrofia cerebral ou síndrome de Dyke-Davidoff-Masson é entidade clínica caracterizada por convulsões, assimetria facial , hemiparesia ou hemiplegia contralateral e déficit cognitivo. Estes achados estão relacionados a lesão cerebral ocorrida na infância ou *in utero*. As características radiológicas são hemiatrofia cerebral e alteracões ósseas no crânio, como espessamento, hiperpneumatização dos seios paranasais e células da mastóide e elevação do ápice da pirâmide petrosa. Descrevemos três casos com achados clássicos da síndrome discutindo aspectos fisiopatológicos.

PALAVRAS-CHAVE: síndrome de Dyke-Davidoff-Masson, atrofia cerebral, tomografia computadorizada, ressonância magnética.

Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome is a congenital, neonatal or early infantile condition characterized clinically by variable degrees of facial asymmetry, seizures, contralateral hemiplegia or hemiparesis, and mental retardation. Mental retardation is not always present[1-3] and seizures may appear months or years after the onset of hemiparesis[3]. Patients may also have speech or language disorders[1]. Radiologically, magnetic resonance (MR) and computed tomography (CT) demonstrate the parenchymal abnormalities of unilateral loss of cerebral volume and compensatory bone alterations in the calvarium, such as thickening, hyperpneumatization of the

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paranasal sinuses and mastoid cells as well as elevation of the petrous ridge and greater wing of the sphenoid bone[2,3].

CASE REPORTS

Case 1. A 39-year-old female with a history of traumatic delivery presented for routine medical evaluation for a diagnosed cerebral palsy. Physical examination revealed facial asymmetry and right hemiatrophy. She had right hemiparesis since birth, seizures at age 18 months, and mild mental retardation. CT an MR showed dilated sulci in the affected minor hemisphere (left), ipsilateral ventricular enlargement, gliosis and encephalomalacia, midline shift to the left, small left cerebral peduncle, small left anterior and middle cranial fossae, and calvarial thickening (Fig 1).

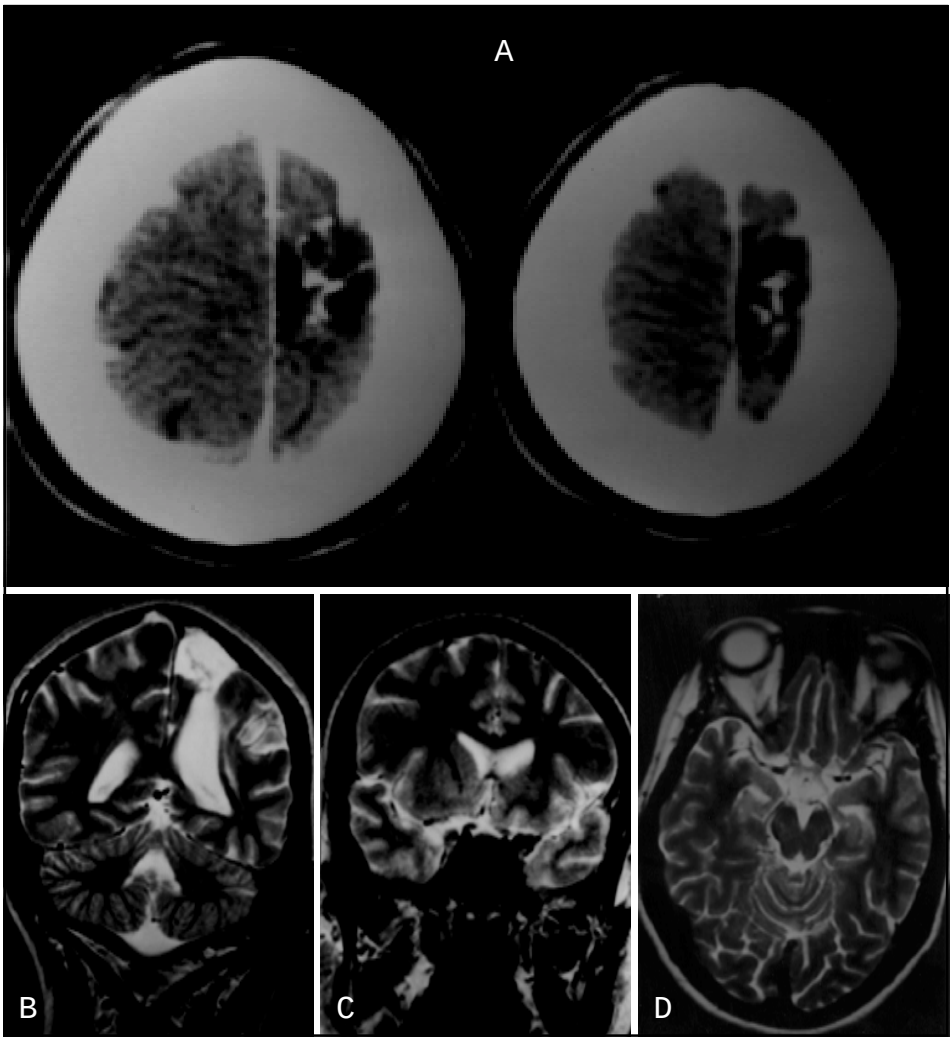

Fig 1. Case 1. Left cerebral hemiatrophy. A. Axial nonenhanced CT (NECT) shows areas of hypoattenuation or encephalomalacia with calcification in the left parietal lobe. The midline is shifted to the left. B and C. Coronal T2-weighted scans. B. The parietal encephalomalacic area is in communication with an enlarged left lateral ventricle. Adjacent cortical sulci are enlarged. C. Small anterior and middle cranial fossae with reduction in brain parenchyma. D. Axial T2-weighted scan: left cerebral peduncle is smaller; also note the atrophic giri and white substance in the temporal lobe.

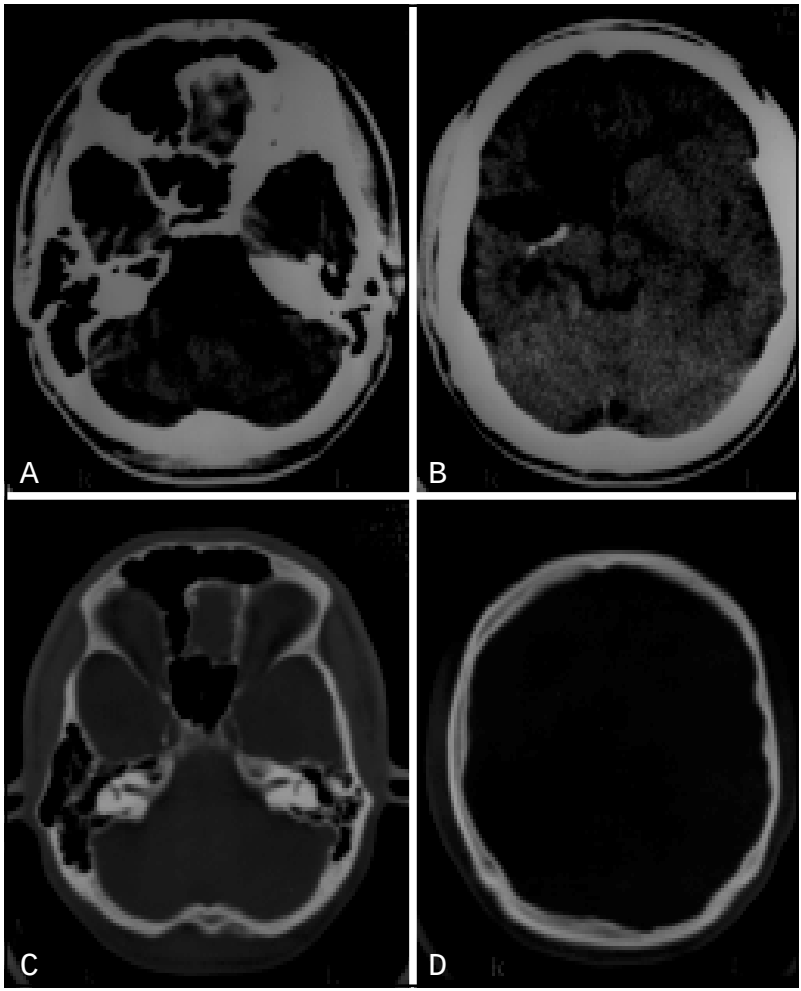

Fig 2. Case 2. Right cerebral hemiatrophy. A to D. Axial NECT scans. A. Porencephalic cyst in the right temporal fossa. B. Calcification surounding the porencephalic cyst and dilatation of the right lateral ventricle. Subarachnoid space is enlarged in the right side. C and D bone window setting. C. Enlarged frontal sinus and mastoid air cells in the right side. D. Note the diploic space thickening.

Case 2. A 19-year-old male had a normal development until age 3 months when he had meningitis. Since then, he evolved with mental retardation, spastic left hemiparesis and seizures. CT demonstrated a small right hemicranium with a small middle fossa and a porencephalic cyst in communication with a dilated lateral ventricle, ipsilateral widened sulci, hyperpneumatization of the frontal and sphenoid sinuses and mastoid air cells, and calvarial thickening (Fig 2).

Case 3. A 28-year-old female was evaluated for recurrent seizures. She was born at 36 weeks by cesarean delivery after unsuccessful forceps vaginal delivery. Facial asymmetry was noted at age 3 years, and seizures started at age 7 years. No detectable mental retardation. MRI revealed a small right hemicranium, with a hypoplasic temporal lobe, occipital porencephalic cyst in communication with a dilated lateral ventricle and adjacent prominent sulci, small cerebral peduncle, and hyperpneumatized right frontal sinus (Fig 3).

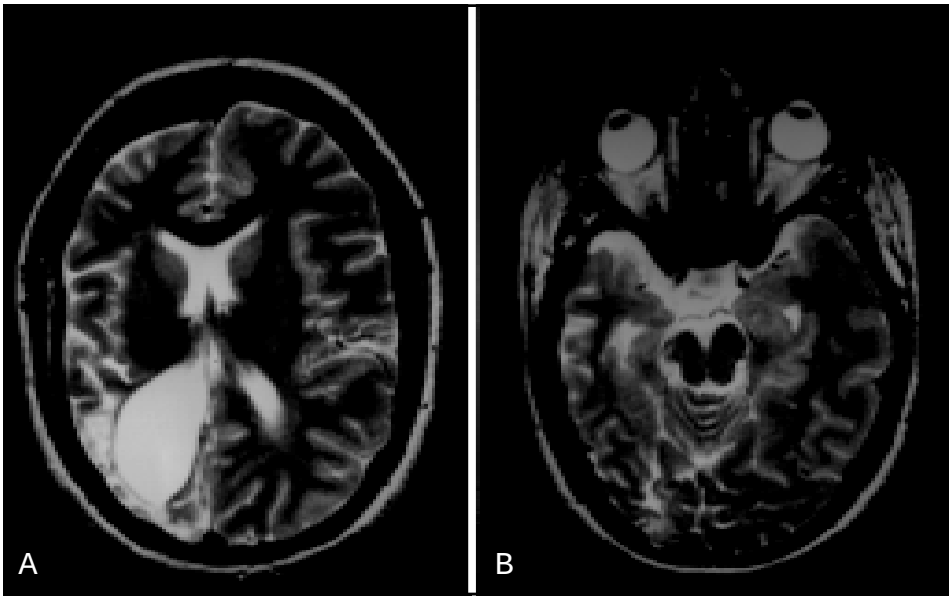

Fig 3. Case 3. Right cerebral hemiatrophy. A and B. Axial T2-weighted scans. A. Small right hemicranium. An enlarged right lateral ventricle is in communication with an occipital porencephalic cyst. Sulci are more prominent in the right hemisphere with reduction of the gray and white matter substances. B. Small right cerebral peduncle.

DISCUSSION

The etiology of cerebral hemiatrophy may be classified into two groups: congenital or primary and acquired or secondary. In the congenital type, cerebral damage which usually has a vascular origin, occurs during intrauterine life and symptoms appear at birth or shortly thereafter. In the acquired or secondary type, cerebral insults occur during the perinatal period or later. The main etiologic factors involved are trauma, infection, vascular abnormalities of the cerebral circulation, ischaemic and haemorrhagic states, and in premature infants, subependymal germinal matrix and intraventricular haemorrhage[2,3]. Coarctation of the midaortic arch has also been involved [4].

The hemiatrophic cerebral parenchyma will have prominent sulci if the vascular insult occurs after birth or after sulcation is complete. On the other hand, if the vascular ischaemia occurs during embryogenesis, when the formation of gyri and sulci is incomplete, no prominent sulci will be present. Encephalomalacia, gliosis, porencephaly, loss of white and gray matter substance, hypoplastic cerebral peduncle, thalamus and internal capsule, ventricular enlargement and midline shift toward the atrophic side may also be present in the hemiatrophic brain[2,3].

The compensatory skull changes reflect adaptations to the unilateral decrease of brain substance and consist of ipsilateral calvarial thickening (diploic space and inner table) with loss of convolutional markings of the inner table of the skull, overdevelopment of the paranasal sinuses (mainly frontal) and mastoid air cells, elevation of the petrous ridge, sphenoid wing and orbital roof, diminished size of the middle/anterior cranial fossae and displacement of falx attachment[1-5].

In the present study, the classical features of the acquired type of cerebral hemiatrophy were found in all patients. The findings of dilated sulci and encephalomalacia in the minor hemisphere reflect a late onset of brain insults which were the consequence of traumatic delivery (Cases 1 and 3) or intracranial infection (Case 2).

Vermian-cerebellar hypoplasia may be present in Dyke-Davidoff-Masson syndrome[6]. It occurs during early embryologic time and may be an occasional finding or associated with other supratentorial abnormalities and syndromes, familial or not. The causes of this condition and of Dandy-Walker malformation and variants are unknown, but there are similar predisposing factors such as gestational exposure to infection, alchool and to some drugs. In the Dandy-Walker malformation, anomalies of the posterior inferior cerebellar arteries, especially absence of the inferior vermian branches and absence of the inferior vermian vein are present[7]. These features support an early onset of insults during the intrauterine life of our patient, probably of vascular origin. Less common than ipislateral cerebellar atrophy[6], contralateral or crossed cerebellar atrophy has also been described in the cerebral hemiatrophy syndrome and it is associated with long-standing, extensive and unilateral cerebral lesions with onset during infancy or early childhood[8].

In the differential diagnosis are other conditions that are associated with cerebral hemiatrophy such as Sturge-Weber syndrome, some brain tumors[2], Silver's syndrome[1], as well as conditions that are associated with unilateral megalencephaly as in the linear nevus sebaceous syndrome. A proper clinical history will provide the correct diagnosis.

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