

DYKE-DAVIDOFF-MASSON SYNDROME: REPORT OF TWO CASES**Monique de Souza Vitoriano Carneiro¹****Marcus Vinicius Della Coletta²****Rhea Sylvia de Souza Mota³****ABSTRACT**

The Dyke-Davidoff-Masson Syndrome results from an insult to the growing brain in utero or early infancy. It is a rare syndrome characterized by seizures, facial asymmetry, contralateral hemiplegia and mental retardation. Radiological findings include cerebral atrophy on one side. Diagnosis is established after clinical history, examination, and MRI. This report presents two cases of patients with epilepsy, deficit unilateral motor, mental retardation and findings characteristic image.

Keywords: Dyke-Davidoff-Masson syndrome (DDMS); Seizures; Cerebral hemiatrophy.

SÍNDROME DE DYKE-DAVIDOFF-MASSON: RELATO DE DOIS CASOS**RESUMO**

A Síndrome de Dyke-Davidoff-Masson resulta de um insulto ao cérebro em crescimento no útero ou primeira infância. É uma síndrome rara, caracterizada por convulsões, assimetria facial, hemiplegia contralateral e retardo mental. Os achados radiológicos incluem atrofia cerebral unilateral. O diagnóstico é estabelecido após história clínica, exame neurológico e ressonância magnética. Este relato apresenta dois casos de pacientes com epilepsia, déficit motor unilateral, retardo mental e achados de imagem característicos.

Palavras-chave: Síndrome de Dyke-Davidoff-Masson (DDMS); Epilepsia; Hemiatrofia cerebral.

INTRODUÇÃO

Dyke-Davidoff-Masson syndrome is a congenital, neonatal or early infantile condition characterized clinically by seizures, contralateral hemiplegia or hemiparesis, variable degrees of facial asymmetry, and mental retardation. Mental retardation is not always present and seizures may appear months or years after the onset of hemiparesis. Patients may also have speech or language disorders. Radiological studies demonstrate the parenchymal abnormalities of unilateral loss of cerebral volume and compensatory bone alterations in the calvarium, such as thickening, hyperpneumatization of the paranasal sinuses and mastoid cells as well as elevation of the petrous ridge and greater wing of the sphenoid bone.

¹ Médica formada pela Universidade Federal do Amazonas (UFAM) Neurologista pela Residência de Neurologia do Hospital Universitário Getúlio Vargas (HUGV). E-mail: moniquevitoriano@hotmail.com

² Formado pela Universidade Federal do Paraná Residência Médica pela Universidade Federal do Paraná Mestrado pela Universidade do Paraná Professor de Neurologia e Coordenador do Serviço de Distúrbio do Movimento da Universidade Federal do Amazonas Especialista em Doppler Transcraniano

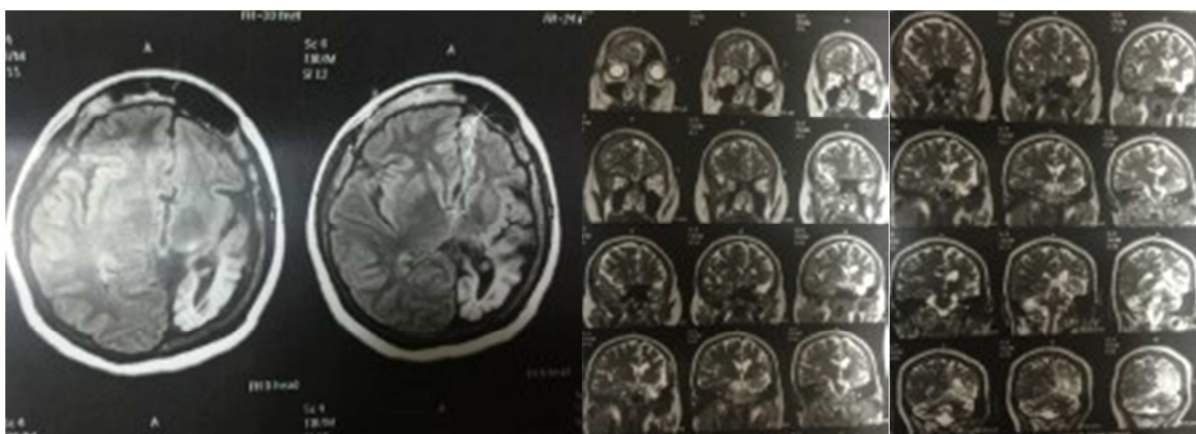
³ Médica formada pela Universidade Nilton Lins Residência Médica pelo Hospital Universitário Getúlio Vargas

CASE REPORT

Case 1. A 36-year-old female with recurrent seizures. She was born by vaginal delivery without complications and had a normal development until 01 year, when she had a febrile seizure. Since then, she evolved with recurrent seizures and developmental delay: started to walking with 02-year-old.

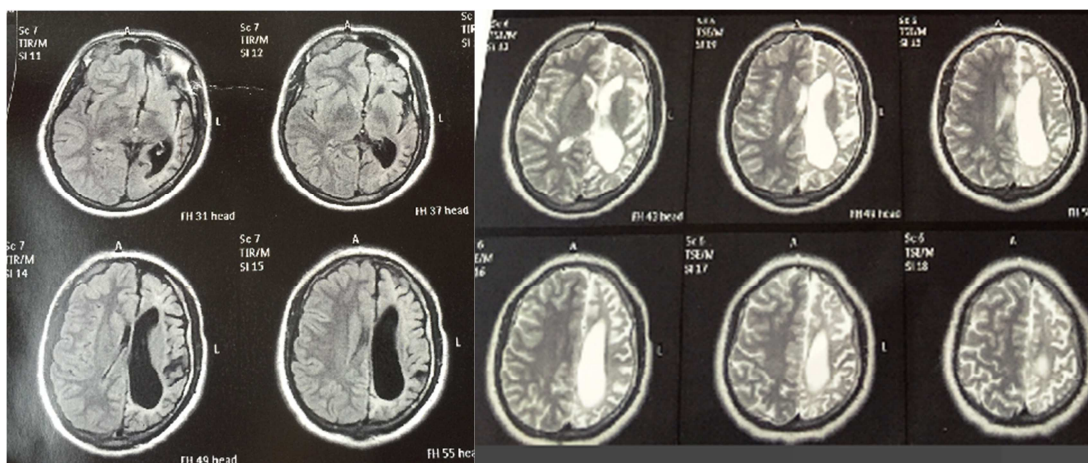
It also presents mental retardation and spastic right hemiparesis. The seizures are difficult to control. MRI revealed left-sided cortical atrophy, hyperintense in T2 and Flair of committed spins, being more evident in the left temporo-parietal lobe with restriction on broadcasting. Asymmetry of cerebellar hemispheres resulting from supratentorial impairment (impairment of cortical-point-cerebellar pathways). Asymmetry of the frontal sinus and mastoid cells predominantly on the left. Expansion former vacuum left lateral ventricle (fig. 1).

Figure 1 - MRI FLAIR and T2 shows left cerebral hemiatrophy and asymmetry of frontal sinus



Case 2. A 28-year-old male with right hemiparesis and seizures. He was born by vaginal delivery without complications and had a normal development until 03 years, when he developed a febrile seizure and delayed psychomotor development: the child already wandered and talked without limitations. The patient developed loss of developmental milestones and persistent hemiparesis and mild mental retardation. At age 18 began tonic-clonic seizures. MRI revealed hemi atrophy the left cerebral hemisphere and hyperpneumatized left frontal sinus. An associate is also compromised ipsilateral hippocampal formation. Reducing the dimensions of the left corticospinal tract along its entire length, this may be related to Wallerian degeneration. Asymmetry cerebellar hemispheres, most right, arising from supratentorial commitment (fig. 2).

Figure 2 - Case 2: MRI shows left cerebral hemiatrophy



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^(1, 2). Prenatal causes include congenital abnormalities, cerebral infarction, vascular malformations and infections. Perinatal causes include birth trauma, hypoxia and intracranial hemorrhage. Finally, cerebral hemi-atrophy can develop secondary to cerebral trauma, tumors, infections and prolonged febrile seizures after birth⁽³⁾. Generally, the etiological factor for Dyke-Davidoff-Masson syndrome has been postulated as trauma, inflammation or vascular malformations and occlusions. When the insult occurs in-utero, it could be due to gestational vascular occlusion, primarily involving the middle cerebral vascular (MCA) territory^(3, 2).

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⁽¹⁾.

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⁾.

According to Sharmal et al decreased carotid artery blood flow due to coarctation of aorta can also cause cerebral hemiatrophy⁽⁴⁾. Garg et al reported a possible etiological relation of cerebral hemiatrophy with febrile seizures. In their three cases of cerebral hemi atrophy, Sener and colleagues reported the middle cerebral artery stroke to be the cause⁽³⁾.

DDMS is seldom seen in clinical practice. When this develops early in life (during the first two years), certain cranial changes like ipsilateral hypertrophy of the skull and enlargement of sinuses occur, the elevations of the greater wing of sphenoid and the petrous ridge on the affected side. The compensatory cranial changes occur to take up the relative vacuum created by the atrophied or hypoplastic cerebral hemisphere^(3, 5).

The classical clinical presentation includes seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation. However, according to Setty et. al., mental retardation is not always present and seizures may appear months or years after the onset of hemi paresis. The clinical findings may be of variable degree according to the extent of the brain injury. Imaging studies show unilateral loss of volume of brain and calvarial changes, findings of cerebral atrophy, ex-vacuo ventricular dilatation and enlargement of sulci^(3, 6).

In the differential diagnosis are other conditions that are associated with cerebral hemiatrophy such as Sturge-Weber syndrome, Rasmussen encephalitis, Fishman syndrome, some brain tumors, Silver's syndrome, 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^(1, 5).

Differential diagnosis it is very broad and are mentioned only the most frequent entities, including: cephalohematoma calcified, subdural hematoma old calcified, internal frontal hyperostosis, dysplasia fibrous, premature synostosis, meningioma, osteomyelitis fímica, etc⁽⁶⁾.

The treatment is symptomatic, and should target convulsion, hemiplegia, hemiparesis and learning difficulties. Prognosis is better if hemiparesis occurs after the age of 2 years and in absence of prolonged or recurrent seizures. Children with intractable disabling and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases^(3, 5).

REFERÊNCIAS

- 1 Aguiar PH, Liu CW, Leitão H, Issa F, Lepski G, Figueiredo EG, et al. MR and CT imaging in the Dyke-Davidoff-Masson Syndrome. Report of three cases and contribution to pathogenesis an differential diagnosis. *Arq Neuropsiquiatr* 1998;56(4):803-807.
- 2 Zawar I, Khan AA, Sultan Tipu, Rathore AW, Dyke-Davidoff-Masson Syndrome. An unusual cause of status epilepticus. *Neurosciences* 2015; 26492121. [Pubmed]
- 3 Ndahiriwe JB, Cook E. Dyke-Davidoff-Masson Syndrome: a rare imaging case. *Rwanda Medical Journal*, March 2012;69(1).
- 4 Sharma S, Goyal D, Negi A, Sood RG, Jhobta A and Surya M. Dyke-Davidoff Masson Syndrome. *Indian Journal of Radiology & ImagingInd*, 2006;16:2:165-166.
- 5 Malik P, Garg R, Gulia AK, Kario, J. Dyke-Davidoff-Masson Syndrome – a rare cause of refractory epilepsy. *J Psychiatry*. 2014. PMC 4277607.
- 6 Cavaria BC, Briceño JC, Oro AB, Muñoz IC, Prieto AE. Atrofia de um hemisfério cerebral: syndrome de Dyke-Davidoff-Masson. Reporte de casos en el Hospital San José-Tec de Monterrey. *Anales de Radiología México* 2007;2:149-155.