

SEGMENTAL MYOCLONUS AND BASILAR ARTERY GIANT ANEURYSM

CASE REPORT

J. PITÁGORAS DE MATTOS * — *ANA L. ZUMA DE ROSSO* **
EDUARDO ZAYEN ** — *SÉRGIO A. P. NOVIS* ***

SUMMARY — A 70 years-old man was admitted at our hospital because of unstable angina pectoris. He had essential hypertension and right hemiplegia from a ischemic stroke two years before admission. On neurologic examination, it was found mental disorientation, unstable emotionality, right spastic hemiparesis with right Babinski sign, and segmental myoclonus affecting the superior lip and the palate (palatal nystagmus) on the right side. On the CT scan, a giant aneurysm of the basilar artery was detected. We conclude that the segmental myoclonus could be explained by ischemic lesions in the Guillain - Mollaret triangle.

KEY WORDS: segmental myoclonus, palatal nystagmus, basilar artery aneurysm.

Mioclonia segmentar e aneurisma gigante da arteria basilar.

RESUMO — Os autores se referem ao caso de um homem de 70 anos, hipertenso e coronariopata, que apresentara, dois anos antes, hemiplegia direita como consequência de acidente vascular encefálico isquêmico. O exame neurológico, na vigência da internação hospitalar, mostrou desorientação espacial, instabilidade emocional (choro imotivado), hemiplegia com sinal de **Babinski** à direita e mioclonia segmentar comprometendo o lábio superior e o palato (nistagmo palatal) à direita. A tomografia computadorizada do crânio revelou aneurisma gigante da artéria basilar. Concluimos que a mioclonia segmentar poderia estar relacionada às lesões isquêmicas no triângulo de Guillain-Mollaret.

PALAVRAS-CHAVE: mioclonia segmentar, nistagmo palatal, aneurisma da artéria basilar.

The first reference in the literature about myoclonic jerk was made by Friedreich, in 1878, naming the abnormal movement as «paramyoklonus multiplex»¹. Presently, the myoclonus is classified according to its distribution as focal, segmental, multifocal and generalized⁵.

We report on a case of segmental myoclonus (soft palate and lips). During the investigation it was found to be associated to a basilar artery aneurysm. A discussion about probable mechanisms of the palatal myoclonus is provided.

CASE REPORT

JR (Reg 223619-7, HUCFF-UFRJ), a 70 years-old negro man, was admitted at our hospital because of unstable angina pectoris. He had essential hypertension and right hemiplegia from a ischemic stroke two years before admission. On neurologic examination, it was found mental disorientation, emotional instability, right spastic hemiparesis with right Babinski's sign, and segmental myoclonus affecting the superior and inferior lips and the palate (palatal nystagmus) on the right side. On the CT scan, a giant aneurysm of the basilar artery was detected as well as ischemic lesions and cerebral atrophy (Figs. 1 and 2). Because of his severe cardiovascular disease the angiographic study was not performed.

Department of Neurology, Clementino Fraga Filho University Hospital (HUCFF-UFRJ), Rio de Janeiro: * Associated Professor; ** Neurologist; *** Head of the Service of Neurology.

Dr. James Pitágoras de Mattos — *Av. São Sebastião 165/101 - 22291 Rio de Janeiro RJ - Brasil.*
Fax (5521) 2673996.

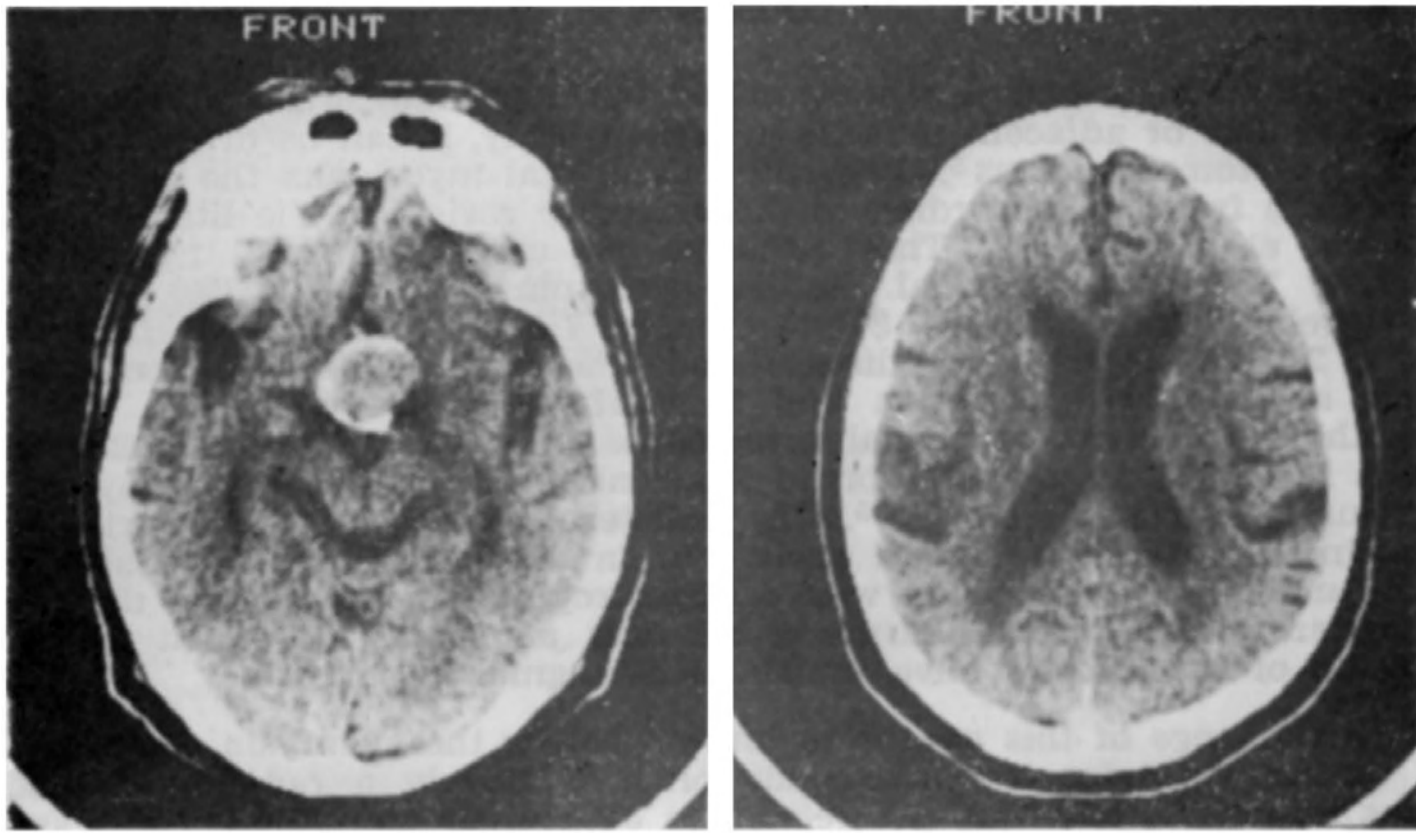


Fig. 1. Case JR. Cranial CT scan without contrast injection showing: a giant aneurysm partially calcified (A, left); cerebral atrophy and ischemic lesions (B, right).



Fig. 2. Case JR. Cranial CT scan with contrast injection showing the giant aneurysm of the basilar artery partially thrombosed.

COMMENTS

Segmental myoclonus is characterized by myoclonic jerks affecting two or more contiguous or adjacent muscles⁵. Aetiologically, it can be divided in essential and symptomatic forms⁵. Regarding to palatal myoclonus the symptomatic form is more frequent, according to the extensive review of the literature made by Deuschl et al.¹ in 1990. They reviewed 287 cases of palatal nystagmus of which only 27% were essential. In the symptomatic group (73%), 40% were due to cerebrovascular disease, 8% to trauma, 7% to neoplasia, 3% to multiple sclerosis, 2% to degenerative disease and 2% to encephalitis. According to Jankovic and Pardo², infarction and hemorrhage of the brainstem were the main pathological cause of palatal myoclonus. It is admitted that palatal myoclonus is secondary to a disruption of the dentato-rubro-olivary pathway, known as the Guillain-Mollaret triangle^{3,4}. The unilateral palatal nystagmus arises from a lesion in the ipsilateral dentate nucleus or in the contralateral central tegmental tract and may be associated with a hypertrophic degeneration of the contralateral inferior olive^{1,5}. Furthermore, the only abnormal movement associated with lesion of the inferior olive is palatal nystagmus⁴.

The challenge of this case is to know whether the aneurysm is responsible direct or indirectly for the myoclonus or it is merely a radiological finding. It seems reasonable that they are related to each other once the cerebrovascular disease is the most frequent cause of the palatal nystagmus and the Guillain-Mollaret triangle is closely related with this involuntary movement. What should be the mechanism by which the aneurysm has induced the involuntary movement: ischemia or mass effect? Teasdall⁶, in 1958, described a similar case on which he demonstrated by angiography a posterior fossa arteriovenous aneurysm in which there was thrombosis of the right vertebral artery. Three months after the ictus, palatal nystagmus was substituted by right soft palate palsy. In that occasion, Teasdall has related the myoclonus with the vascular thrombosis.

The direct (ischemic areas on the CT scan, angina pectoris) and indirect signs of diffuse atherosclerosis in our patient, together with the presence of a giant aneurysm of the vertebro-basilar system, suggest brainstem ischemic lesions and not a compressive effect as the probable cause of segmental myoclonus.

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