Pontine-Mesencephalic Cavernoma Presenting as the Millard Gubler Syndrome

Cavernoma Pontomesencefálico Cursando com Síndrome com Millard-Gubler

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ABSTRACT
Cavernous malformations (CMs) are relatively uncommon vascular malformations found in the central nervous system (CNS), with a prevalence of approximately 0.5% in the general population. The brainstem CMs represent only a small percentage of these lesions, which can vary between 4 to 35%. The Millard-Gubler syndrome (MGS) is one of the famous crossed brainstem syndromes, and although classic, its presentation is rare. We present the case of a patient with brainstem cavernoma who bled and presented with MGS. The patient underwent microsurgical treatment and presented good postoperative evolution.

Keywords: Cavernoma; Millard-Gubler syndrome; Cavernous malformation; Crossed brainstem syndrome

RESUMO
Malformações cavernosas (MCs) são malformações vasculares relativamente incomuns encontradas no sistema nervoso central (SNC), com prevalência de aproximadamente 0.5% na população geral. As MCs de tronco encefálico representam apenas uma pequena porcentagem dessas lesões, que podem variar entre 4 a 35%. A síndrome de Millard-Gubler (MGS) é uma das famosas síndromes do tronco encefálico cruzado e, embora clássica, sua apresentação é rara. Apresentamos o caso de um paciente com cavernoma do tronco cerebral que sangrou e apresentou MGS. O paciente foi submetido ao tratamento microcirúrgico e apresentou boa evolução pós-operatória.

Palavras-chave: Cavernoma; Síndrome de Millard-Gubler; Malformação cavernosa; Síndrome alterna do tronco encefálico

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Cavernous malformations (CMs) are relatively uncommon vascular malformations found in the central nervous system (CNS), with a prevalence of approximately 0.5% in the general population, making up between 5 to 10% of all vascular malformations within the brain\(^1\). The brainstem CMs represent only a small percentage, which can vary between 4 to 35%\(^1\). Regarding its histopathological features, CMs exhibit expanded, delicate, sinusoidal vascular channels that are enveloped by an endothelial lining\(^1\). These channels contain blood in various stages of thrombosis and organization, with lesions frequently encircled by hemosiderin and gliosis\(^1\). Thus, CMs in the brainstem are infrequent but hold particular significance due to their potential to induce severe and persistent neurological symptoms in such a noble location of the CNS. Minor alterations in these lesions can lead to conditions such as hemiplegia, respiratory impairment, and altered consciousness, contingent upon their specific location\(^9\).

We present the case of a patient with pontine-mesencephalic cavernoma associated with the Millard-Gubler syndrome (MGS), who was benefited from microsurgical treatment. MGS is one of the classical crossed brainstem syndromes defined by a unilateral lesion of the basal portion of the caudal pons involving fascicles of abducens and the facial cranial nerve, and the pyramidal tract fibers\(^2\). The hallmark of this syndrome is the contralateral paralysis affecting the limbs and ipsilateral involvement of the face, with ischemic stroke being the most common causes\(^3\).

A 21-year-old male presented with nausea, vomiting, and headache, accompanied by neurological symptoms including isochoric and photo reactive pupils, and left hemihypoesthesia. Magnetic resonance imaging (MRI) revealed a cavernoma with signs of subacute bleeding (Figure 1). Due to familial anxiety due to fear of the risks of surgery, the surgery was delayed until bruise absorption. He was on carbamazepine, tramadol, and clonazepam. A subsequent visit, one month later, reported recurrent symptoms — nausea, vomiting, and occipital headache — with new onset right facial paralysis and difficulty closing his right eye. Neurological examination revealed worsening of symptoms, including right miosis, left hemiparesis, and spastic gait. MRI indicated new apoplexy with enlargement of the lesion and obstructive hydrocephalus, prompting immediate surgical intervention. The patient was positioned in a semi-sitting position and a retrosigmoid craniotomy was performed. As the cavernoma was still in the acute bleeding phase, the surgical access route was easily identifiable. Complete microsurgical resection of the cavernoma was performed (Figure 2) and the patient evolved well after surgery. Eight months after surgery, the patient had fully recovered from hemiparesis and peripheral facial paralysis.

Patients with brainstem CMs may exhibit a variety of symptoms, commonly including headache, dizziness, nausea, vomiting, and, in rare cases, trigeminal neuralgia\(^1\). Following hemorrhage, symptoms may gradually diminish, but recurrences can occur, as observed in our case. Against these odds, MGS was noticed in the patient in its complete form, since it manifests as the sixth cranial and seventh nerve palsy on the same side, and severe hemiparesis on the opposite side (involving the pyramidal tract)\(^3\).

**Figure 1.** Pre-operative MRI. shows a large pontomesencephalic lesion. There are some areas of spontaneous hypersignal on T1 and areas of hypointense on SWI (by hematic material in a cavernomatous lesion), which measures about 5.0 × 4.0 × 4.0 cm (AP × LL × CC) and determining larger dimensions over the IV ventricle and reduced amplitude of the basal cisterns. There was increased ectasia of the supratentorial ventricular system. **A.** T1-weighted MRI in axial view. **B.** Sagittal view. **C.** T2-weighted in axial view.
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Treatment options encompass both monitoring and surgery, with consideration of hemorrhage risks and involvement of the family in the decision-making process. Factors such as age, presence of developmental venous anomaly, perilesional edema, lesion size, and location may influence the risk of new hemorrhage event. Approximately 8 months after microsurgery, the patient no longer exhibited neurological deficits, underscoring the importance of early intervention and a holistic approach in managing patients with brainstem CMs.

**REFERENCES**


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