

Man-in-the-barrel syndrome: history and different etiologies

Síndrome do Homem do Barril: histórico e etiologias várias

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SUMMARY

The “Man-in-the-barrel syndrome” (MBS) makes reference to a picture of bilateral brachial muscular weakness, of proximal predominance, that incapacitates the patients in the execution of functional activities related with the elevation of the upper limbs. Since its description, there are several cases described of patients with this syndrome, whose etiology differs from the first one described. The most frequent causes of MBS are cerebral vascular disorders, the cardiac and extracardiac surgeries with demonstration of arterial hypotension, cerebral metastases, pons and spinal cord involvement. Bilateral brachial paralysis also can cause similar picture. Thus, the pathophysiologic substratum that becomes related it MBS, in some cases, is uncertain and deserves attention.

Keywords: Neuromuscular Diseases. Motor Neuron Disease. Anterior Horn.

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RESUMO

A síndrome do “homem do barril” (SHB) faz referência a um quadro de fraqueza muscular braquial bilateral, de predomínio proximal, que impossibilita os pacientes na execução de atividades funcionais relacionadas com a elevação dos membros superiores. Desde sua descrição, inúmeros casos de pacientes com essa síndrome foram relatados, cuja etiopatogenia difere daquela descrita inicialmente. As mais frequentes causas de SHB são distúrbios vasculares cerebrais, cirurgias cardíacas e extracardíaca com demonstração de hipotensão arterial, metástases cerebrais, e o envolvimento da ponte e medula espinhal. Paralisia braquial bilateral também ocasiona quadro similar. Assim, o substrato fisiopatológico que se relaciona a SHB, em alguns casos, é incerto e merece atenção.

Unitermos: Doenças Neuromusculares. Doença do Neurônio Motor. Corno Anterior.

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INTRODUCTION

Centrally originating brachial diplegia is defined by motor deficiency in the two upper limbs following damage to pyramidal cortico-spinal pathways. This clinical presentation is quite unusual and is distinct from more frequent paraplegia¹. The “man-in-the-barrel syndrome” (MBS) is characterized by bilateral upper extremity paresis with intact motor function in the lower extremities. The patient is unable to move his arm to any stimulus, appearing as if the upper limbs are confined within a barrel². It was originally attributed to cerebral infarctions in the watershed zones between the middle and anterior cerebral artery distributions³. However, other pathophysiologic processes localizing to the cerebral cortex, pons, anterior horn cells of cervical spinal cord and even the peripheral nerves may also produce this phenotype^{4,7}.

MAN-IN-THE-BARREL SYNDROME

Dide initially described bilateral paralysis of the upper limbs (brachial diplegia) in 1917⁸. It has been attributed to a variety of further clinical conditions, such as polyneuropathy, motoneuron disease, central pontine myelinolysis, ischemia of the anterior horns during septicemia, cervical epidural infection and, cervical spinal cord ischemia^{7,9-14}. In 1970, Bell introduced the term “cruciate paralysis” when he described the cases of three patients with paralysis of both arms caused by injury of the pyramidal decussation in the upper portion¹⁵. In the first patient the cause was a fracture of the odontoid process with a complete dislocation of the upper fragment, the second presented a hyperextension cervical spine injury, and the third there was the coexistence of a basilar impression and acute hydrocephalus following suboccipital craniectomy for removal of a meningioma of the posterior fossa.

Mohr, in 1969, used the term “distal field infarction” to describe borderzone infarcts caused by systemic hypoperfusion or carotid obstruction, where, in the latter, brachial paresis was the predominant clinical symptom¹⁶. Sage prospectively examined 34 comatose patients who had suffered systemic hypoperfusion of various causes and was the first to use the term MBS to describe the paresis of both arms that he found paresis in 11 of this patients¹⁷. This description emphasized the clinical aspect of the patient who, although is able to maintaining the mobility of lower limbs and face, seems to have the upper limbs stuck in a barrel¹. Since this

initial description, cases of MBS has been reported following cardiac surgery (with or out both of systemic hypoperfusion), following cardio-respiratory arrest, in association with bilateral hemorrhagic contusion of the inferior frontal lobes after closed head injury, brain metastasis, and infarction of the central spinal cord after unilateral vertebral artery dissection^{4,6,18-20}. The pathology of brachial diplegia is secondary to superficial anterior junctional infarcts, resulting in ischemic lesions in the border zone between the vascular territories of the middle and anterior cerebral arteries, which are responsible for lesions of the precentral gyrus, the motor region for the arm and shoulder. The brachial diplegia occurs when the damage is bilateral, as in cases of systemic hypofusion. The deficiency may be global or predominantly distal or proximal. The motricity of the face or of the lower limbs are not affected, and either deep or superficial sensitivity^{2,6,18-20}. Deep tendon reflexes are sharp because of damage to the pyramidal pathways¹⁸⁻²⁰.

The MBS is caused for severe systemic hypoperfusion, that can be defined as systolic blood pressure less than 50 mmHg for period of at least 5 min^{3,17,20,21}. Cases of MBS occurring in patients after anesthesia who a episode of severe systemic hypoperfusion in not clear have been described, but a moderate fall in systolic blood pressure of 55 and 90 mm Hg can be noted^{2,6,19}. In these cases, the rapidity fall of the blood pressure could be responsible for the ischemia.

MBS has also been reported secondary to brain involvement instead of watershed infarctions, like in metastatic disease and closed head injury²². MBS may also due to pontine lesions⁷. Two cases in which a central pontine myelinolysis was followed by brachial diplegia with involvement of the lower limbs was also reported^{7,11}. In spinal cord involvement there are cases reported of infarction of the spinal cord, upper spine injuries and cervical epidural infection^{23,24}. The motor neurons of the cervical cord may be involved as in amyotrophic lateral sclerosis (ALS) or in lower motor neurons disease (LMND)¹⁰. In these second disorders the arms are completely paralyzed with normality of the lower limbs. Sometimes there is involvement of both the brachial plexus as in rare cases of bilateral amyotrophic neuralgia²⁵.

The prognosis of these processes usually is negative in its majority, cause the gravity of the etiological process²⁵, like in ALS, with a mortality of up

to 90% in some series¹⁷. However, there are cases in which the recovery was total or there was a persistence of a mild distal deficiency, this fact suggest that the prognosis can be related, mainly, to the severity and the duration of the systemic hypoperfusion¹.

CONCLUSION

In many cases MBS is due to a serious cerebral hypoperfusion that secondarily originates bilateral ischemic infarcts in the bordering territories between the anterior and medium cerebral arteries, in the areas responsible for the brachial motility. There are other cases where this syndrome is due to cerebral metastases, hemorrhagic contusion by craniocerebral trauma, involvement of the pons, medulla and the cervical spinal cord as in ALS and LMND. Although the etiology of the MBS is varied, the prognosis of this syndrome is not benign in many cases.

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