Relato de Caso

Improving idiophatic orthostatic hypotension in a patient with multiple system atrophy (Shy-Drager syndrome) with cardiac pacemaker implantation. Case report.

Melhora da hipotensão ortostática em um paciente com atrofia de múltiplos sistemas (syndrome de Shy-Drager) com implantação de marca-passo cardíaco. Relato de caso.

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Resumo

Hipotensão ortostática é um sintoma freqüente em pacientes com atrofia de múltiplos sistemas e tem importante impacto na sua qualidade de vida.

Relatamos um paciente com hipotensão ortostática idiopática e atrofia de múltiplos sistemas (Síndrome de Shy-Drager), tratado com implantação de marca-passo resultando numa evidente melhora na qualidade de vida.

Palavras- chave: *atrofia de múltiplos sistemas, hipotensão ortostática, marca-passo, síndrome de Shy-Drager*

Abstract

Orthostatic hypotension is a frequent symptom in patients with multiple system atrophy and it has an important impact on their quality of life.

We report a case of idiopathic orthostatic hypotension, in a patient with multiple system atrophy (Shy-Drager syndrome), treated with pacemaker implantation resulting in a substantial improvement in the quality of his life.

Keywords: *multiple system atrophy, orthostatic hypotension, pacemaker, Shy-Drager syndrome*

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Introduction

Multiple system atrophy (MSA) is a neurodegenerative disorder classified according to its molecular pathogenesis as an á-synucleinopathy. The pathology is characterized by cell loss, gliosis, and glial cytoplasmic inclusions in several central nervous system structures, with a clinical combination of parkinsonian, autonomic, cerebellar, or pyramidal signs[1].

The estimated prevalence of MSA may vary from 1.9 to 4.9 cases per 100,000 people. It is uncommon before the sixth decade of life, with an expected survival of six to nine years, although there is a significant variation of disease progression[1].

As MSA has no specific treatment, improvement of quality of life is the main goal in these patients.

Dysautonomia is a characteristic symptom of MSA and it is considered a result of sympathetic disturbance from a neuronal preganglionic lesion[2]. Symptomatic orthostatic hypotension is described in 68% of patients[3]. A recent study of the quality of life in patients with MSA demonstrated that the severity of autonomic disturbance is a major factor for the variance in life satisfaction in this group[4].

There are two forms of idiopathic orthostatic hypotension. In one form there is probably a selective degeneration of neurons in the sympathetic ganglia with denervation of smooth muscle vasculature and adrenal glands without evidence of lesions in other parts of the nervous system[5]. In the second type (Shy-Drager syndrome)[6] there is a degeneration of preganglionic neurons in the lateral columns of gray matter in the spinal cord leaving postganglionic neurons isolated from spinal control. This form is associated with degeneration of other systems of neurons in the substantia nigra and locus coeruleus, or striatonigral degeneration and olivopontocerebellar degeneration. In the first two syndromes orthostatic hypotension is associated with parkinsonian manifestations, and in the third syndrome it is associated with cerebellar symptoms.

We report a case of multiple system atrophy, with severe dysautonomia and considerable improvement after cardiac pacemaker implantation.

Case description

The patient is a 64-year-old male, with no family risk factors, and a history of falling since 2001. He had a preceding occurrence of ischemic coronaropathy and myocardic revascularization in 1988, with a normal cardiac catheterization after the surgery.

The falls were always related to the transition to an orthostatic position and were preceded by becoming pale and minor confusion. Initially, it used to happen every 20 to 30 days with worsening of the frequency over time. By the year 2004, the patient presented with severe blood pressure drops every time he had to change from lying to a sitting or standing position. He had difficulty performing daily activities and self care and required continuous assistance. At this time, he started to have difficulty walking and writing, and his voice changed. Nevertheless, despite all the new symptoms, the major disturbance was still related to the falls.

At his evaluation in 2007, he had an arterial blood pressure of 130/90 in the dorsal position and 70/50 in the orthostatic position. He had dysarthric speech, with no aphasia. His gait was ataxic and unstable, with difficulty in controlling the left leg. His deep tendon reflexes were asymmetrical, more pronounced in the right leg, with a positive Babinski sign on this side. He had a plastic rigidity in all four limbs, more pronounced in the legs, and a dystonic posture of the left hand. Vertical eye movements were normal. There were no signs of dementia or history of hallucinations. The patient could be classified as "probable MSA" according to the consensus of MSA clinical criterias[7].

In the complementary investigation, 1.5 Tesla brain magnetic resonance imaging (MRI) was normal. A Tilt Table Test documented severe orthostatic hypotension (Figure).

Because of all the symptoms, the patient had a poor quality of life, mainly associated with the orthostatic hypotension. Levodopa and fludrocortisone were prescribed when the patient's symptoms became worse.

Consequently, having researched previous descriptions in literature, it was decided to implant a cardiac pacemaker to control the dysautonomic abnormalities. In August 2007, a bicameral pacemaker, with a rate drop response algorithm for vagal syndrome was implanted (Figure).

After the surgery, the patient had a dramatic improvement of the orthostatic hypotension. He spent four months without any falls and the first six months with few symptoms related to the dysautonomia. The patient and his relatives also reported that, in the first year after the pacemaker implantation, his voice and his writing improved and he had a better quality of sleep.

Since the beginning of 2008, his motor and cerebellar symptoms have become more pronounced. At his last

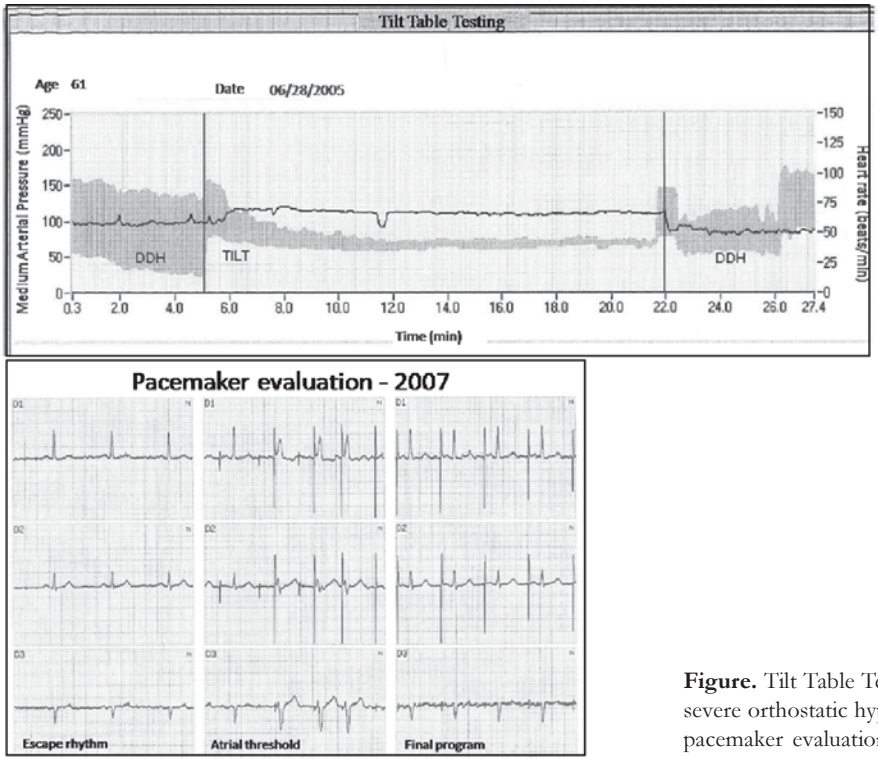

Figure. Tilt Table Test demonstrating severe orthostatic hypotension and pacemaker evaluation.

evaluation, in August 2009, the patient was almost completely restricted to a wheelchair. Nevertheless, he has a few events of brief dizziness when changed from one position to the other or when he is held in the standing position.

Discussion

Multiple system atrophy is a progressive neurologic disorder, with no specific treatment. Dysautonomia, especially orthostatic hypotension, is present in more than 50% of MSA cases and is responsible for the decrease in the quality of life of these patients.

In this context, particular attention should be taken of dysautonomic symptoms and neurologists should pursue the best treatment option in order to minimize patients' distress.

The treatment of orthostatic hypotension is important not only to improve quality of life in MSA patients, but also to prevent injuries related to the falls and to help the caregiver in advanced cases.

In orthostatic hypotension, although some patients respond to simple non-pharmacological or pharmacological treatments, others do not, especially patients with severe symptoms. A few cases of pacemaker implantation for the treatment of orthostatic hypotension have been published since the 1980s[8-10].

The patient described in this article had orthostatic hypotension as his first MSA symptom and it remained his major difficulty, despite all the other neurological systems involved, until the pacemaker implantation. At that time, he was dependent only because of the frequent falls, and his social life and daily activities were extremely impaired.

In spite of the fast progression of the disease after the procedure, he gained at least six months of meaningful independence. Moreover, even after he was restricted to the wheelchair, the fact that he no longer had blood pressure drops and prolonged confusion also helped the caregiver provide him better assistance.

It is also important to note that this patient lived for at least five years with minor neurological difficulties other than dysautonomia. If he had been evaluated earlier, he would probably have benefitted even more from the pacemaker implantation.

Conclusion

The satisfactory treatment of orthostatic hypotension in patients with MSA must be of major concern to neurologists. Pacemaker implantation should be considered as an option to improve the quality of life in these patients and it should not be postponed.

References

1. Wenning GK, Colosimo C, Geser F, Poewe W. Multiple system atrophy. *Lancet Neurol.* 2004, 3:93-103.
2. Goldstein DS, Robertson D, Esler M, Straus SE, Eisenhofer G. Dysautonomias: clinical disorders of the autonomic nervous system. *Ann Intern Med* 2002; 137(9):753-63.
3. Wenning GK, Ben-Shlomo Y, Magalhaes M, Daniel SE, Quinn NP. Clinical features and natural history of multiple system atrophy: an analysis of 100 cases. *Brain* 1994; 117:835–45.
4. Schrag A, Geser F, Stampfer-Kountchev M, et al. Health-related quality of life in multiple system atrophy. *Mov Disord* 2006; 21:809–815.
5. Bradbury S, Egglestone C. Postural hypotension: a report of three cases. *Am Heart J* 1925; 1:73-86.
6. Shy GM, Drager GA. A neurological syndrome associated with orthostatic hypotension: A clinical-pathologic study. *Arch Neurol* 1960; 2:511-527.
7. Gilman S, Low P, Quinn N, et al. Consensus statement on the diagnosis of multiple system atrophy. *J Neurol Sci* 1999; 163**:**94–98.
8. Moss AJ, Glaser W, Topol E. Atrial tachycardia in the treatment of a patient with primary orthostatic hypotension. *N Engl J Med* 1980; 302:1456-7.
9. Kristinsson A. Programmed atrial pacing for orthostatic hypotension. *Acta Med Scand* 1983; 214:79-83.
10. Cunha UGV, Machado ELG, Santana LA. Marcapasso atrial programável no tratamento da hipotensão ortostática neurogênica no idoso. *Arq Bras Cardiol* 1990; 55:47-49.