

Differential diagnosis among parkinsonian and dystonic camptocormia

Martínez Héctor R,¹ Cantú-Martínez Leonel,²

Martínez-Ramírez Daniel,² González Humberto C,² Venegas-Moniet Pamela²

ABSTRACT

Background: Camptocormia is characterized by a marked thoracolumbar spine flexion that disappears in resting position. A clinical analysis among a patient with parkinsonian camptocormia and a case with dystonic camptocormia is described. **Methods:** Among our cohort of 140 patients with Parkinson disease we found one patient with camptocormia. The diagnosis was performed when the patient exhibited a flexion ($> 45^\circ$) of thoracolumbar spine increasing during walking and disappearing in recumbent position. This patient was compared to a patient showing a severe lateral flexion of the spine that was diagnosed as dystonic camptocormia. Both cases had complete neurological examination, UPDRS, and head, cervical and thoracic magnetic resonance imaging. Clinical manifestations and response to treatment are described. **Results:** A 46 years-old male patient started with right lateral flexion of his trunk, head drop and laterocollis 3 years before evaluation, no extrapyramidal signs were observed. He received thioridazine for 2 years at the age of 40. MRI showed a normal brain, cervical lordosis and thoracic kyphosis. A moderate improvement with botulinum toxin was observed. A 72 years-old Parkinsonian male developed forward flexion ($> 45^\circ$) in the last year that increased during walking. Head and spine MRI were normal. He showed improvement on his extrapyramidal signs and posture after levodopa treatment. **Discussion:** Patient with dystonic camptocormia showed a lateral flexion of the trunk associated to dystonic posture of the neck, head drop, absence of extrapyramidal signs and moderate response to botulinum toxin injection. On the other hand, the patient with Parkinsonian camptocormia showed severe forward flexion of the trunk, extrapyramidal signs, absence of head drop and improvement in his posture with levodopa treatment. Since the pathogenesis of camptocormia is unknown and the etiologies are multiple, there is no accepted form of treatment. **Conclusion:** Differential diagnosis of camptocormia subtype may be useful for an accurate classification and for an appropriate treatment.

Key words: Camptocormia, dystonia, botulin toxin, parkinsonism, bent spine, Pisa syndrome.

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Diagnóstico diferencial entre camptocormia parkinsoniana y camptocormia distónica

RESUMEN

Introducción: La camptocormia se caracteriza por flexión marcada de la columna toracolumbar que desaparece en reposo. Se describe análisis clínico en un paciente con camptocormia parkinsoniana y otro con camptocormia distónica. **Métodos:** En nuestra cohorte de 140 pacientes con enfermedad de Parkinson sólo uno mostró camptocormia. El diagnóstico se efectuó en presencia de flexión $> 45^\circ$ de la columna toracolumbar que aumenta al caminar y desaparece en posición supina. Se compararon los datos clínicos con la severa flexión lateral de la columna observada en un paciente con distonía. En ambos casos se efectuó examen neurológico, UPDRS, resonancia magnética de cerebro, columna cervical y torácica. Se describen hallazgos clínicos y tratamiento. **Resultados:** Masculino de 46 años inició con flexión lateral derecha del tronco y cabeza con laterocollis de tres años de evolución y sin signos extrapiramidales. Recibió tioridazina por dos años a la edad de 40 años. La IRM cerebro fue normal y la columna reveló lordosis cervical y cifosis torácica. Posterior a la aplicación de toxina botulínica se observó mejoría de su postura. Masculino de 72 años de edad con enfermedad de Parkinson, desarrolló flexión anterior del tronco ($> 45^\circ$) en el último año que aumentaba al caminar. La resonancia resultó normal. Se observó mejoría de su postura y signos extrapiramidales con levodopa. **Discusión:** El paciente con camptocormia distónica muestra flexión lateral del tronco asociada a postura distónica de cabeza y cuello, ausencia de signos extrapiramidales y moderada respuesta a la toxina botulínica. Por otra parte, el paciente con camptocormia parkinsoniana mostró severa flexión anterior del tronco, signos extrapiramidales, ausencia de posturas anormales de cabeza y cuello y mejoría de la postura con levodopa. La etiología de la camptocormia es variada, su patogénesis es desconocida y no existe tratamiento aceptado. **Conclusión:** El diagnóstico diferencial del subtipo de camptocormia puede ser útil para una clasificación adecuada y tratamiento apropiado.

Palabras clave: Camptocormia, distonía, toxina botulínica, parkinsonismo, espina curva, síndrome de Pisa.

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1. MD, FACP. Servicio de Neurología, Hospital Universitario UANL.

2. MD, Servicio de Neurología, Hospital Universitario UANL.

Correspondence:

Héctor R. Martínez, MD, FACP.

Servicio de Neurología, Hospital Universitario UANL. Ave Madero y Gonzalitos S/N. Col. Mitras Centro, Monterrey, N.L. México CP 64460.

INTRODUCTION

A stooped posture referred as camptocormia or bent spine is characterized by severe forward flexion of the thoracolumbar spine. This disorder is a motor phenomenon of heterogeneous etiology that was first described by Brodie in 1818.¹ Souques in 1915 proposed the term camptocormia.² A number of papers have recently appeared describing camptocormia as a clinical entity.³⁻¹¹ These include a proposal for an etiologic classification⁹ and controversies by acronyms used in cases with bent spine syndromes such as Pisa syndrome, head drop (head ptosis) and camptocormia.¹² Pisa syndrome (or Pleurothotonus) first described by Ekbom¹³ in 1972, is a clinically similar form of severe axial dystonia characterized by tonic flexion of the trunk to one side or forward and laterally oriented. It was described as a motor phenomenon appearing several days after starting neuroleptic treatment.¹³⁻¹⁵ Head drop, is characterized by marked anterior curvature or angulation of the cervical spine that has been described in association with neuromuscular and extrapyramidal disorders. Camptocormia, head drop and Pisa syndrome are descriptive terms of many different organic and functional disorders of spinal posture.⁹⁻¹⁵ However, these clinical conditions might be present in the same patient and probably implies variants or subtypes of camptocormia. Azher and Jankovic reported their own experience and literature review about camptocormia, head drop and bent spine syndrome proposing an etiologic classification of camptocormia.⁹ The aim of the present report is to describe the clinical differences observed among a patient with camptocormia in Parkinson disease and a case of camptocormia as a manifestation of axial dystonia and their response to treatment.



Figure 1. Distonic camptocormia.

CASES REPORT

Patient 1

A 46 years-old male patient with history of epilepsy in his childhood and thioridazine treatment during 2 years at the age of 40 by a Psychiatrist due to psychotic depression and agitation with normalization of his psychiatric disorder after 2 years follow up. He started with right lateral flexion of his trunk 3 years before evaluation. The neurological exam revealed right lateral flexion of his trunk, head drop and laterocolis. The flexion of the trunk and head drop increased during walking and disappeared in recumbent position, laterocolis remains unchanged either on walking or lying on bed (Figure 1). He neither showed pyramidal or extrapyramidal signs nor sensitive or cranial nerves abnormalities. He denied back pain, difficulty arising from a seated position, slowness of movements or tremor. MRI of head and spine showed normal brain and spinal cord but cervical lordosis and thoracic kyphosis were observed. The dystonic posture of the head and trunk progressed in subsequent evaluations despite of treatment with biperiden, piracetam, baclofen and valproate. Prior to our evaluation he received clonazepam and levodopa without improvement. His lateral flexion of the trunk and head drop showed moderate improvement after botulinum toxin injection.

Patient 2

A 72 years-old male presented with rigidity, bradykinesia and forward flexion of his trunk. He started 4 years before evaluation with rigidity in right upper and lower limbs and rest tremor in his right hand. He was treated with trihexiphenidile with



Figure 2. Parkinsonian camptocormia.

improvement on his tremor. He had occasional visual hallucinations that disappeared spontaneously. During the last year he referred forward flexion of his trunk that increased during walking. On examination he exhibited hypomimia, rest tremor in his right hand, bradykinesia, rigidity in all limbs greater in the right side and stooped posture with forward flexion of his trunk ($> 45^\circ$). When he was sitting in a chair or lying on bed he maintained erect posture, however during walking his stooped posture increased (Figure 2). MRI of the head and spine showed a normal brain and spinal cord. He was treated with levodopa-carbidopa-entacapone (100-25-200 mg) four times a day with marked improvement of his Parkinson signs and in his posture.

DISCUSSION

Camptocormia is becoming an increasingly recognized clinical entity that has been associated to parkinsonian and dystonic disorders.³⁻¹¹ Parkinson disease was recently described as the most frequent etiology of camptocormia.⁹ Although in our cohort of 140 Parkinson patients only one satisfied the criteria for camptocormia. This disorder has a broad spectrum of musculoskeletal and neurologic etiologies. Besides Parkinson disease, camptocormia has also been described in focal and axial myopathy,^{6,16} stroke,¹⁷ amyotrophic lateral sclerosis,^{18,19} multiple system atrophy,^{10,11,20} primary dystonia,^{9,21,22} drug induced^{14,15,23} and Tourette syndrome.⁹ Spine deformities are considered among the musculoskeletal disorder.²⁴

Recently Azher and Jankovic proposed an etiologic classification of camptocormia.⁹ They performed an analysis of 16 cases diagnosed in a 24 years period and a thorough review of the literature of camptocormia, head drop, and bent spine syndrome. Camptocormia associated to Parkinson disease and as a manifestation of axial dystonia without parkinsonism are the most common neurologic etiologies reported in recent literature.⁴⁻⁹ Psychogenic etiology was considered in early reported cases.^{2,25,26} Until quite recently, it was described as a conversion reaction in male military recruits and soldiers during World War I and II.^{2,9}

In our series, one case was diagnosed as dystonic camptocormia since he showed severe lateral flexion of his trunk, dystonic posture of the neck, head drop and absence of extrapyramidal signs. Moreover he received thiroidazine before the age of 45 years and no response was observed after treatment with levodopa or some other drugs. However, after botulin toxin injection a moderate improvement was observed. This finding is in agreement with a previous report describing notable improvement of camptocormia in 4 out of 9 patients after botulin

toxin injection.⁹ All these data allow us to establish the diagnosis dystonic camptocormia. Patients described as Pisa syndrome are clinically similar to our case, therefore we consider those cases correspond to dystonic camptocormia.

Our patient diagnosed with parkinsonian camptocormia was older than the dystonic case. Clinically, showed severe forward flexion of the trunk, presence of extrapyramidal signs, absence of head drop and clinical improvement after levodopa treatment. In most cases of parkinsonian camptocormia described in the literature and presented in clinical pictures,^{3,6,8,9} the patients are observed with forward (no lateral) flexion of the trunk as it is seen in our case (Figure 2). The clinical response observed with levodopa in our case is in disagreement with a minimal or no clinical improvement observed in 12 camptocormia cases after levodopa treatment.⁹ The improvement observed in our case is explained by the absence of previous levodopa treatment and the spectacular response that is frequently observed in Parkinson disease patients naive to treatment at the beginning of levodopa.

The pathogenesis of camptocormia in patients with Parkinson disease is not known, but it may represent one end of the spectrum of the related abnormal postures in this disorder.^{27,28} These abnormal postures may range from striatal hand or foot deformity (on one end of the spectrum) to scoliosis and camptocormia (on the other end of the spectrum).⁹ Camptocormia may be the most disabling feature of Parkinson disease and other parkinsonian disorders.

A number of papers have recently appeared describing camptocormia as a clinical entity.³⁻¹¹ A controversy was raised not only in regards to idiopathic camptocormia as a distinct clinical entity but also by the acronyms used in cases with bent spines "camptocormia" and "head ptosis".¹² Camptocormia, head drop and Pisa syndrome are descriptive terms of many different organic and functional disorders of spinal posture. However, these clinical conditions might be present in the same patient and probably implies variants or subtypes of camptocormia.

The abnormal posture of the neck is described to be typically present in patients with multiple system atrophy (flexion of the neck) and in progressive supranuclear palsy (neck extension).^{9-11,20} However, we consider that the presence of head drop and the abnormal posture of the neck observed in our patient is a clinical feature that may be useful in the differential diagnosis among dystonic camptocormia from parkinsonian camptocormia. The age of presentation, absence of extrapyramidal

Table 1
Clinical features in camptocormia

Feature	Parkinsonian	Dystonic
Extrapyramidal signs	Present	Absent
Forward flexion of the trunk	+	-
Lateral flexion of the trunk	-	+
Head drop	-	+
Laterocolis	-	+
MRI brain or spinal cord lesions*	±	±
Response to levodopa	+	-
Response to botulin toxin	-	+

* Both patients did not show parenchymal injuries. In the literature abnormalities in camptocormia secondary to BVE or multiple system atrophy are described.

signs and the clinical improvement after botulin toxin injection are some other clinical features that may be considered in the differential diagnosis of the camptocormia (Table 1). In conclusion, since the pathogenesis of camptocormia is unknown and the etiologies are multiple, there is not an accepted form of treatment. Differential diagnosis of camptocormia subtype may be useful for an accurate classification and for an appropriate treatment.

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