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CLINICAL CASE

Immediate recovery of neurological function in response to deep brain stimulation of the *globus pallidus internus* in a patient with idiopathic camptocormia

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Camptocormia is a major disabling abnormality characterized by severe forward flexion of the thoracolumbar spine. We report here on the effectiveness of deep brain stimulation (DBS) for the management of a case of untreatable idiopathic camptocormia. The patient, a 51-year-old male, with an 11-year-long history of radicular pain. Camptocormia symptomatology initiated 4 years ago. Preoperative muscle electrodiagnostic testing was within normal limits. Myopathy was ruled out. In the standing position myokymic discharges were recorded. Under local anesthesia and stereotactic control, electrodes for DBS were placed bilaterally in the globus pallidus internus. Patient's symptoms disappeared immediately following DBS. This response cannot be attributed to the surgical procedure itself. When stimulators were turned ''off'' accidentally, the patient returned immediately to his pre-surgery condition. Erect posture and walking were restored when stimulators were back ''on''.

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PALABRAS CLAVE
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Síndrome de la columna vertebral doblada

Recuperación inmediata de la función neurológica en respuesta a la estimulación cerebral profunda del globo pálido interno en un paciente con camptocormia idiopática

Resumen La Camptocormia es un trastorno incapacitante caracterizado por flexión anterior severa del tronco. Aquí presentamos un caso de camptocormia idiopática intratable donde la estimulación cerebral profunda (ECP) fue altamente eficaz. El paciente, un hombre de 51 años de edad, con historia de dolor radicular de 11 años y sintomatología de camptocormia de 4 años de evolución. Se descartó la presencia de miopatías. Las pruebas musculares preoperatorias mediante electrodiagnóstico resultaron normales, aunque en la posición de pie se registraron descargas mioquímicas. Para proveer la ECP se colocaron electrodos en el globo pálido interno bilateralmente, bajo anestesia local y control estereotáctico. Los síntomas de camptocormia desaparecieron inmediatamente después de la ECP. Esta respuesta no es atribuible a la intervención quirúrgica en sí, pues cuando el estimulador se apagó accidentalmente, el paciente regresó a su condición pretratamiento y la postura erecta se restauró al restablecer la estimulación.

Introduction

Sir Benjamin Collins Brodie (1783–1862) described in 1818 a disease in which the low back pain and abnormal curvature of the spine were caused by a vertebral destructive process, perhaps he never named this entity.[1–3]

The term camptocormia was first used by Souques and Rosanoff-Saloff (cyphose hystérique, 1915) to describe bent spine in military personnel (''bent back of soldiers'' or ''war hysteria'') who had been sent to the front lines in the First World War.[1,2] Originally considered as a psychogenic disorder, the expression has been adapted to describe severely flexed postures observed in Parkinson disease (Unified Parkinson's Disease Rating Scale and the Hoehn and Yahr scale) and other etiologies (drug induced, muscular disease, paraneoplastic and etcetera); is a heterogeneous disorder, even described as idiopathic in some cases.[3–6]

Camptocormia is more of a syndrome rather than a sign; it can be attributed to the progressive weakness of the antigravity muscles linked to trunk extension,[5] as well as weakness of the gluteus maximus.[4]

Camptocormia, also known as bent spine syndrome (BSS), is a major disabling acquired abnormality characterized by severe forward flexion of the thoracolumbar spine (angle over 45°) with total correction to the supine position. This abnormal posture is involuntary and is only evident when the patient is standing or walking. At severe status, normal activities are difficult or impossible to perform.[1–7]

We report here on the neurological recovery of a patient diagnosed with idiopathic camptocormia after functional neurosurgery treatment.

Clinical case

A 51-year-old man, business consultant, with a history of spine events for radicular pain without myelopathy: a lumbar disk surgery (L5-S1) eleven years ago, and two years ago a cervical C4-C5 and C5-C6 discectomy and anterior fusion.

Four years ago he initiated his camptocormia clinical presentation with the sensation of anterior traction of his body, resulting in uncontrollable trunk flexion that was progressive, and became permanent, severely impairing his daily activities and the quality of his life (Fig. 1).

Eight months before being referred to us he was diagnosed with idiopathic camptocormia, and was started unsuccessfully treatment on levodopa. Later, on two occasions, he received *botulinum* toxin type A injections into the *rectus abdominus*, it caused discrete clinical benefit for 3 months after the first dose; the second dose produced no effect. In our hands, electromyography (EMG) and imaging studies (plain X-rays and MRI) did not reveal any abnormality associated with camptocormia.

During the physical exploration, he was found in good general condition (vital signs: blood pressure 128/85, 72 BPR, 18 BPM, 36.6 °C; height 1.81 m, weight 86 kg), suffering weakness for the truncal extension and pronounced back flexion with an approximate angle of 120° (chess over knees); compensatory hyperextension of the neck and half-bent knees (210°). Also, the position of the arms hanging and swinging like an ape walking.

Preoperatively, motor and sensory nerve conduction velocity were within normal limits in upper and lower extremities. Median, ulnar, peroneal, and tibial nerves were studied. With the patient lying in the supine position, F waves and H reflexes were also found to be within normal limits.

Simultaneous EMG four-channel recordings of abdominal muscles (right-left, upper-lower) were obtained preoperatively in the operating room. Insertional activity was normal when the patient was lying on his back. Spontaneous EMG recordings did not show fibrillation nor fasciculation. During voluntary contraction, motor unit action potentials (MUAPs) showed normal parameters, and recruitment and interference patterns were within normal limits. When the patient was in the sitting position, a spontaneous mild contraction of the lower and upper *rectus abdominus* was felt through palpation. EMG recordings showed muscle activity