Camptocormia

Camptocormia is a rare, involuntary movement disorder, presenting as truncal flexion while standing or walking, and is mainly observed as a feature of Parkinson's disease (PD) and primary dystonia.

Lateral trunk flexion is often seen in patients with Parkinson disease, sometimes coming on as a subacute phenomenon associated with medication adjustments, and in others with gradual onset that seems related to a neurodegenerative process related to the evolution of the disease.Either acute or subacute presentations seem to be pure abnormalities in the coronal plane and are usually reversible. However, a chronic form occurs often in a combined fashion with anteroposterior flexion (camptocormia), improves only partially, remains stable, or even worsens over time.The acute/subacute phenotype is the condition originally named as Pisa syndrome (PS).The pathophysiology of PS remains poorly understood, and a cholinergic-dopaminergic imbalance has been suggested as being involved in the cause of this disorder. The role of other neurotransmitters and how they become dysfunctional in PS remains to be elucidated.Specific treatments, other than discontinuing the medications responsible for the disorder, whenever possible, are undeveloped because of the unknown etiology [1)].

Epidemiology

A total of 275 consecutive outpatients were systematically screened for camptocormia with a clinical evaluation. Patients who screened positive for camptocormia were subsequently reassessed by formal goniometric analysis. The demographic and clinical features of the patients with and without camptocormia were then compared.

A 6.9% (19/275, 95% CI, 4.2 to 10.6) prevalence of camptocormia was found. Camptocormia was found in patients with more severe PD, as clinically assessed by the Hoehn-Yahr (HY) staging and the motor Unified Parkinson Disease Rating Scale (UPDRS) part III, longer l-dopa treatment duration and greater l-dopa daily dose and presence of DSM-IV dementia. Camptocormia was reported to develop after the clinical onset of PD. No correlation was found between the degree of trunk flexion and age, duration of PD, UPDRS motor score, HY staging, and l-dopa treatment duration and dose. As a risk factor, the study identified previous vertebral surgery.

Camptocormia, a relatively common sign in PD seems to be related to the clinical severity of PD [2)].

Etiology

There is ongoing controversy concerning its mechanisms and no consensus regarding the underlying etiology. This report demonstrates a case in which a dopaminergic agonist (DA) was implicated in the onset of camptocormia episodes in a non-PD patient who developed camptocormia after the start of DA treatment. Over a course of 8 years, the patient experienced intermittent camptocormia, which resulted in multiple falls. After cessation of the DA, the patient showed decreased camptocormia symptoms. This case report suggests that clinicians should consider the possibility of DA-induced camptocormia in patients with PD and non-PD patients receiving DA treatments, and serves to caution clinicians regarding the administration of DAs [3)].

Camptocormia (CC), and head drop syndrome (HD) in PD are predominantly myopathic. Aberrant protein aggregation may link PD and CC [4].

Clinical features

Camptocormia causes significant spinal and abdominal pain, impairment of balance, and social stigma.

Treatment

Although Parkinson's disease (PD) is the most frequent etiologic factor, this postural disorder responds poorly to levodopa or other medications.

Surgical options include transient, external spinal stimulation; DBS targeting the subthalamic nuclei; and spinal deformity surgery. Benefit from DBS stimulation was inconsistent. Spine surgery corrected spinal imbalance but was associated with a high complication rate [5].

The success of spinal surgery in patients with Parkinson disease depends on an interdisciplinary approach, including both surgeons and movement disorder specialists, to select appropriate surgical patients and manage postoperative movement in order to decrease mechanical failures. Achieving appropriate correction of sagittal alignment with strong biomechanical instrumentation and bone fusion is the key determinant of satisfactory results [6].

In a systematic review, Chieng et al., aimed to profile the various reported interventions for camptocormia in Parkinson's disease (PD) and give an overview of the benefits of deep brain stimulation (DBS). Currently, there is no consensus in the literature regarding this. PD manifests in several ways and camptocormia is one of the commonly encountered problems for both spine and functional neurosurgeons. DBS was introduced in 2002 in the USA, and since then its efficacy and applications have tremendously increased.

They reviewed the PubMed and Medical Subject Headings database using the phrases “Parkinson's disease” or “Parkinson” in combination with “spinal deformity” or “camptocormia” or “bent spine syndrome” and “deep brain stimulation”. The review was limited to English language literature and they excluded camptocormia of non-PD origin. The search yielded 361 articles with 131 patients in the pooled data. The majority (59%) of patients were women and the age range was 48-76 years. While half the patients on levodopa (n=42) saw no improvement of their camptocormia, 71% of the lidocaine group (n=27) and 68% of the DBS group (n=32) showed significant improvement. For mean flexion angle, the spinal surgery and DBS group demonstrated profound improvement in the bending angle, 89.9% and 78.2%, respectively. However, major complications following spinal surgery were noted. Although the results are from a small group of patients, DBS has achieved sustained improvement in camptocormia with low postoperative morbidity, and appears to be a promising treatment option. A larger, long term study is necessary to establish comprehensive outcome data [7].

Seven patients (4 with dystonia, 3 with PD; mean age 60.3 years at surgery, range 39-73 years) with

camptocormia were included in the study. Five patients underwent bilateral GPi DBS and two patients underwent bilateral STN DBS guided by CT-stereotactic surgery and microelectrode recording. Pre- and postoperative motor assessment included the BFM in the dystonia patients and the UPDRS in the PD patients. Severity of camptocormia was assessed by the BFM subscore for the trunk at the last available follow-up at a mean of 17.3 months (range 9-36 months). There were no surgical complications. In the four patients with dystonia there was a mean improvement of 53% in the BFM motor score (range 41-79%) and of 63% (range 50-67%) in the BFM subscore for the trunk at the last available follow-up (mean 14.3 months, range 9-18 months). In the three patients with camptocormia in PD who underwent bilateral STN DBS (2 patients) or pallidal DBS (1 patient), the PD symptoms improved markedly (mean improvement in the UPDRS motor subscore stimulation on/medication off 55%, range 49-61%), but there was no or only mild improvement of camptocormia in the two patients who underwent STN DBS, and only moderate improvement in the patient with GPi DBS at the last available follow-up (mean 21 months, range 12-36 months). GPi DBS is an effective treatment for camptocormia in dystonia. The response of camptocormia to chronic STN or GPi DBS in PD is more heterogenous. The latter may be due to a variety of causes and needs further clarification [8)].

Postural abnormality in patients with PD could be ameliorated by STN DBS, and therefore surgery should be considered before irreversible spinal deformity develops [9)].

Outcome

The CSA of paraspinal muscles and erector spinae width can be good predictive markers for improving camptocormia in patients with PD after deep brain stimulation. [10)].

Although the results are from a small group of patients, DBS has achieved sustained improvement in camptocormia with low postoperative morbidity, and appears to be a promising treatment option. A larger, long term study is necessary to establish comprehensive outcome data [11)].

A relationship may exist between improvement of camptocormia and severity of paraspinal muscle degeneration. [12)].

Case series

evaluated 17 patients with camptocormia whose preoperative off-medication thoracolumbar angle exceeded 45°. We used photographs to measure their thoracolumbar angle preoperatively, 3 months after surgery, and at the last follow-up (mean 36.5 months postoperatively) in status on-medication and off-medication. The patient age, duration of PD and camptocormia, daily medications, Unified Parkinson's Disease Rating Scale (UPDRS) subscores and the Schwab-England activity of daily living scale (S-E) were also recorded. Univariate analysis was performed to identify factors predictive of the postoperative improvement of camptocormia.

STN stimulation significantly improved the UPDRS subscores and S-E, and resulted in a reduction of daily medications 3 months post-treatment. The preoperative thoracolumbar angle (mean±SD) in

status off-medication (84.0±29.5°) was significantly ameliorated 3 months postoperatively (49.8±29.3°) and at the last follow-up (54.8±28.3°). There was no correlation between the postoperative camptocormia improvement rate and preoperative parameters other than the duration and severity of camptocormia and the levodopa responsiveness of the thoracolumbar angle. Symptom duration negatively affected levodopa responsiveness.

STN stimulation improves PD-associated camptocormia in parallel with preoperative levodopa responsiveness. Long symptom duration interferes with levodopa responsiveness [13].

Reese et al., report on a retrospective clinical assessment of three patients with primary dystonic camptocormia treated with GPi-DBS.

All three patients showed marked response to bilateral GPi-DBS within days to weeks after surgery which was preserved in the long-term (38-45 months after implantation: mean improvement 82% as rated on the Burke Fahn Marsden Dystonia Rating Scale, 89% in the subitem “trunk”). Two patients developed mild stimulation induced speech problems (stuttering or dysarthria) which resolved with reprogramming or were acceptable in return for the control of dystonic symptoms.

The diagnosis and treatment of camptocormia will continue to require expert knowledge in movement and neuromuscular disorders, but DBS may expand treatment options in this difficult patient population [14].

Case reports

Paucar et al published a variant ataxia-telangiectasia with prominent camptocormia [15].

A 38-year-old man with anterior truncal bending that developed when he was 36 years old. Prior to the onset of the symptom, he had been taking antipsychotic drugs for schizophrenia. There were no features of PD; the symptom severely interfered with his walking and daily life. He was given anticholinergics, clonazepam, and botulinum toxin injections, which did not result in much success. Because of the patient's unwillingness to undergo implantation of a hardware device, he underwent staged bilateral pallidotomy with complete resolution for a diagnosis of tardive dystonic camptocormia. The Burke-Fahn-Marsden dystonia rating scale subscore for the trunk before and after bilateral pallidotomy was 3 and 0, respectively. No perioperative adverse events were observed. Effects have persisted for 18 months. Bilateral pallidotomy can be a treatment option for medically refractory dystonic camptocormia without the need for device implantation [16].

Ekmekci et al., present the case of a 51-year-old woman who has been followed with PD for the last 10 years, and also under the therapy for PD. An unappreciated correlation low back pain with camptocormia developed. She underwent deep brain stimulation (DBS) in the subthalamic nucleus bilaterally and improved her bending [17].

A 42-year-old male, presented with dystonia of 5-year duration, that initially started with cervical dystonia and later progressed to severe disturbance of posture causing involuntary truncal flexion induced by standing or sitting. The camptocormia was completely relieved when sitting on a chair or lying down on the bed. Routine blood testing was normal. The workup for secondary dystonia including slit lamp examination for Kayser-Fleischer ring and serum copper studies did not reveal any abnormality. Magnetic resonance imaging of the brain was unremarkable. The electromyogram of the lumbar and thoracic paraspinal muscles was also normal.

The patient was initially treated with multiple drugs and Botulinum A toxin which were ineffective. He underwent bilateral globus pallidus interna (GPi) DBS Over a period of 2 weeks; there was a mild reduction in the dystonia of the trunk and neck. The maximum improvement in dystonia, approximately 30% over baseline, was noted at 2 weeks postsurgery and over a further long-term follow-up, the improvement was 50% as determined by the sub-item (trunk) assessment of the Burke-Fahn-Marsden (BFM) dystonia score. Cervical dystonia improved by >90% in sub-item (neck) assessment of BFM scale.

In this report, we have shown the efficacy of GPi DBS in the treatment of drug refractory dystonia associated camptocormia. Although only reported for PD associated camptocormia, evaluation for truncal extensor myopathy is mandatory in these cases also to achieve a good outcome [18)].

A 63-year-old female with a long history of severe Parkinson's disease (PD) and subsequent onset of debilitating camptocormia who underwent successful bilateral subthalamic nucleus deep brain stimulation surgery (STN DBS). The literature and previous reports are reviewed. The patient history and details of the surgical procedure are reported including the implantable pulse generator (IPG) settings and response to stimulation. The results of her PD symptom and camptocormia improvements are discussed. Five year postoperatively, the patient has enjoyed good results for both her PD symptoms as well as significant and sustained improvement in her thoracolumbar flexion deformity. She remains on minimal medications and no longer requires any assistive devices for ambulation. Comparison and contrast of the current world literature on DBS for camptocormia is reviewed. We discuss the current targets used for DBS in the setting of camptocormia in the cases reported, both PD associated and not associated. The optimal target has yet to be defined and further work on appropriate patient selection is needed [19)]

A 57-year-old woman with Parkinson disease developed severe camptocormia, which did not improve with trials of antiparkinsonian and muscle relaxant medications. The patient was successfully treated with bilateral globus pallidus interna deep brain stimulation surgery under general anesthesia. High-frequency neuromodulation afforded relief of camptocormia and improvement in Parkinson disease symptoms.

Camptocormia in Parkinson disease may represent a form of dystonia and can be treated effectively with chronic pallidal neuromodulation [20)].

Sakas et al., reported on 2 young patients who developed drug-resistant idiopathic dystonic

camptocormia (bent spine) and were treated successfully by deep brain stimulation (DBS) of the globus pallidus internus (GPi). The first patient, a 26-year-old woman, suffered for 3 years from such severe camptocormia that she became unable to walk and was confined to bed or a wheelchair. The second patient, a 21-year-old man, suffered for 6 months from less severe camptocormia; he was able to walk but only for short distances with a very bent spine, the arms in a parallel position to the legs, and the hands almost approaching the floor to potentially support him in case of a forward fall. Within a few days following DBS, both patients experienced marked clinical improvement. At most recent follow-up (44 months in one case and 42 in the other), the patients' ability to walk upright remained normal. Similar findings have only been reported recently in a few cases of camptocormia secondary to Parkinson disease or tardive dyskinesia. On the basis of the experience of these 2 idiopathic cases and the previously reported cases of secondary camptocormia with a favorable response to GPi DBS, the authors postulate that specific patterns of oscillatory activity in the GPi are vital for the maintenance of erect posture and the adoption of bipedal walking by humans [21)].

Although a mild stooped posture is a hallmark of parkinsonism, extreme trunk forward flexion is not common. This phenomenon was described in different etiological entities and called camptocormia. Other similar presentations called Pisa syndrome and antecollis were described mainly in extrapyramidal disorders. Authors present two cases of probable multiple system atrophy (MSA) with predominant parkinsonism and Pisa syndrome (or camptocormia). Both of them were previously misdiagnosed as idiopathic Parkinson's disease (PD) and one was reported 1 year earlier. The typical clinical presentation fulfilling the diagnostic criteria for multiple system atrophy, rapid progression with lack of responsiveness to L-DOPA and apomorphine and typical MRI putaminal pathology observed in both cases allowed us to make a diagnosis. Accuracy of clinical diagnosis in multiple system atrophy is still very poor. Therefore, unusual or rare clinical presentations may support the final diagnosis. The camptocormia, Pisa syndrome and antecollis may represent the continuum of the same motor phenomenon and most of the authors refer them to unusual form of axial dystonia. According to many clinical presentations on different forms of camptocormia/Pisa syndrome authors conclude that not etiology, but the localization of specific lesion, probably within putamen is responsible for that form of dystonia. In cases of parkinsonism and severe forward flexion of trunk multiple system atrophy, diagnosis should be considered [22)]

Yamada et al. reported a PD patient in whom chronic bilateral subthalamic nucleus stimulation produced a striking alleviation of camptocormia [23)].

A man with early non-fluctuating Parkinson's disease developed disabling camptocormia. The patient was treated with posterior thoracolumbar fixation, which subsequently had to be augmented with anterior interbody fusion. Although the patient ultimately achieved excellent sagittal correction, his postoperative course was complicated and prolonged. Therefore, although this case demonstrates that spinal fixation surgery can be successful, it should probably only be offered after subthalamic nucleus deep brain stimulation has been unsuccessful, or for well motivated patients who express a strong wish for this major reconstructive surgery [24)].

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