

Review Article

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Postural & striatal deformities in Parkinson's disease: Are these rare?

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Parkinson's disease (PD) is the most common neurodegenerative disease and is characterized by tremor, rigidity and akinesia. Diagnosis is clinical in the majority of the patients. Patients with PD may have stooped posture but some of them develop different types of postural and striatal deformities. Usually these deformities are more common in atypical parkinsonian disorders such as progressive supranuclear palsy and multisystem atrophy. But in many studies it has been highlighted that these may also be present in approximately one third of PD patients leading to severe disability. These include antecollis or dropped head, camptocormia, Pisa syndrome, scoliosis, striatal hands and striatal toes. The pathogenesis of these deformities is a complex combination of central and peripheral influences such as rigidity, dystonia and degenerative skeletal changes. Duration of parkinsonism symptoms is an important risk factor and in majority of the patients these deformities are seen in advanced stage of the disease. The patients with such symptoms may initially respond to dopaminergic medications but if not intervened they may become fixed and difficult to treat. Pain and restriction of movement are most common clinical manifestations and these may mimic symptoms of musculoskeletal disorders like rheumatoid arthritis. Early diagnosis is important as the patients may respond to adjustment in dopaminergic medications. Recent advances such as deep brain stimulation (DBS) and ultrasound guided botulinum toxin injection are helpful in management of these deformities in patients with PD.

Key words Deformities - Parkinson's disease - postural - striatal

Introduction

James Parkinson in his essay on shaking palsy described the propensity to bend forward as an important feature of Parkinson's disease (PD)¹. In one of his case description he highlighted that as the disease progressed to its last stage the trunk permanently bowed. Later Charcot described the extension of great toe and flexion of other toes as an important feature of PD which is now known as striatal toe². Since then although there are many published studies³⁻⁶ on postural and striatal deformities in PD but these

are still considered to be rare and if present, raises a possibility of atypical Parkinsonian disorders such as progressive supranuclear palsy (PSP), multiple system atrophy (MSA) and drug induced parkinsonism (DIP). The classical deformity described in a PD patient is stooped posturing characterized by flexion of hips and knees. Patients with atypical parkinsonism have severe type of postural deformities which may also be present at the onset but PD patients usually have these deformities in advanced stages many years after the initial diagnosis. These deformities cause significant pain, discomfort and walking difficulty leading to

severe disability and poor quality of life⁴. On many occasions these deformities are wrongly diagnosed as skeletal and joint disorders leading to wrong diagnosis and management. The aim of this review is to highlight that postural and striatal deformities are not rare in PD patients and these need to be identified and treated at the earliest possible time.

Types of deformities

Parkinson's disease patients may have different types of postural deformities (Table I and Fig. 1) in sagittal and coronal planes³. Deformities in sagittal plane include antecollis, dropped head, and camptocormia. Pisa syndrome and scoliosis are coronal plane deformities.

Retrocollis is rarely seen in PD patients and usually considered as a red flag sign for the diagnosis. Striatal deformities may involve hand (striatal hand) or toes (striatal toes)⁴. These may either be present alone or in combination with postural deformities⁵. In a study which included 164 PD patients; 55 (33.5%) had some type of deformities⁶ (Table II). This study also included 19 patients with MSA and 19 PSP patients where 13 (68.4%) and five (26.3%) patients, respectively had any of the deformities. This study highlighted that approximately one third of the PD patients had some type of limb deformities which was a significant number and contrary to the belief that these are only common in MSA and PSP patients.

Antecollis and dropped head

In a PD patient there may be forward flexion of neck and head which is known as antecollis. When this forward flexion is severe and chin of the patient is bent down upon sternum it is called dropped head.

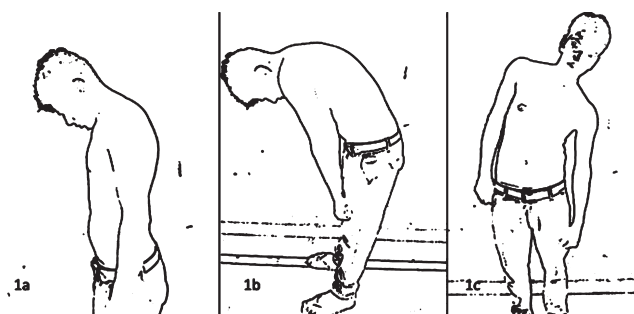


Fig. 1. Line diagrams showing antecollis (a), camptocormia (b) and Pisa syndrome (c).

Commonest symptom because of antecollis and dropped head is neck pain and occasional swallowing difficulties. It is important to do detailed neurological examination to rule out neuromuscular weakness like polymyositis and myasthenia gravis. Suspicion should be stronger if there is extreme degree of neck extensor weakness. Many PD patients show fluctuation in symptoms with dopaminergic medications. It is important to rule out role of DOPA agonists which may lead to neck drop in some patients. Antecollis and neck drop have been reported in 5-9 per cent of patients in different studies^{12,13}. In a study which included 124 cases of dropped head syndrome, 21 patients had parkinsonism symptoms which included 10 patients of Parkinson's disease⁷. Dystonia was also present in 12 patients, four had myopathy and five patients had dystonia and myopathy⁷.

Pisa syndrome

Pisa syndrome (PS) is defined as lateral flexion of the trunk which may also have rotation of the spine

Table I. Definition of deformities usually seen in Parkinson's disease patients

Deformities	Types	Definition
Postural deformities	Antecollis	Neck and head flexion (>45°)
	Dropped head	Severe neck and head flexion and chin is bent down upon sternum
	Camptocormia	Forward flexion at thoracolumbar spine manifesting during standing and walking and completely subside in recumbent position
	Pisa syndrome	Lateral flexion of the trunk which may also have rotation of the spine which completely subside in supine position
	Scoliosis	Lateral flexion of the spine does not subside by passive movement and has radiological evidence of rotation of vertebra
Striatal deformities	Striatal hand	Flexion at metacarpophalangeal, proximal interphalangeal and distal interphalangeal joints
	Striatal foot	Extension of great toe with flexion of other toes

Source: Refs 2-4

Table II. Prevalence of postural and striatal deformities in published recent series

Study	Patient population	Deformities	Comment
Ashour & Jankovic ⁶ 2006	PD(164) Some individuals had more than one deformities	Deformities (55) included striatal limb (21), camptocormia (20), antecollis (9), scoliosis (14)	Striatal deformities were on the side of initial symptom in 100% of patients
Savica <i>et al</i> ⁷ 2012	PD (10) MSA(10) Drug induced (1)	All dropped head (124) patients who had parkinsonism	Dystonia alone (12) myopathy (4) dystonia+myopathy (5)
Tassorelli <i>et al</i> ⁹ 2012	PD (300)	Scoliosis (55) included Pisa syndrome (20)	Scoliosis (>15°) on wall goniometer
Tinnazi <i>et al</i> ¹¹ 2013	PD (13)	Pisa syndrome (13)	Pisa syndrome minimum (10°) lateral flexion
Doherty <i>et al</i> ⁸ 2013	PD (15)	Pisa syndrome (15)	Mobile scoliosis (3) and structural scoliosis 12
Seki <i>et al</i> ¹⁰ 2011	PD (531)	Camptocormia (22)	Flexion of thoracolumbar spine (>45°). Higher frequency of autonomic symptoms

PD, Parkinson's disease; MSA, multiple system atrophy. Figures in parenthesis indicate number of patients

which completely subsides in supine positioning. This syndrome is seen commonly in patients on antipsychotic therapy and has also been described in MSA and Alzheimer's disease. It is not clear why some PD patients develop this syndrome. Diagnostic criteria and definition lack consensus leading to a high variability in reported prevalence ranging from 2 to 90 per cent¹⁴. A small group of patients may show response in symptoms of PS following levodopa treatment³. In a study from Italy which included 1400 parkinsonism patients, PS was found in 2 per cent cases¹⁵. The criteria for diagnosis of PS was lateral flexion ($\geq 15^\circ$ measured on wall goniometer) which increased during walking and disappeared in the recumbent position. In a study 15 PD patients fulfilled the diagnostic criteria for PS but all had radiological evidence of scoliosis⁸. Clinically, three had mobile scoliosis and 12 had structural scoliosis which persisted in the supine position (Table II). In another study, it was highlighted that the majority of patients with PS, trunk bending was contralateral to the side of symptom onset and paravertebral thoracic muscles and abdominal oblique muscles had abnormal tonic hyperactivity on the side of the deviation⁹.

Scoliosis

Scoliosis is defined as lateral flexion of the spine which does not subside by passive movement and has radiological evidence of rotation of vertebra. The available studies have found much variation

in prevalence of scoliosis ranging from 9 to 91 per cent^{6,16}. In a study scoliosis was seen in only 9 per cent of PD patients assessed by clinical examination⁶. In another study, 19 patients among 21 consecutive patients of parkinsonism (17 PD, 4 postencephalitic parkinsonism) detected by clinical examination had scoliosis¹⁶. But in a different study where scoliosis was measured by Cobb's method considering $\geq 10^\circ$ as significant lateral flexion, scoliosis was found in 33 per cent PD patients¹⁷. It was also documented that the prevalence of scoliosis among healthy elderly patients varied from 6 to 30 per cent, but this figure ranged from 43 to 90 per cent in PD patients¹⁷. Scoliosis is usually contralateral to the initial side of symptoms. This happens due to gradual deviation of spine away from the initial affected side due to cholinergically mediated tonic influences on posture and locomotion by caudate nucleus.

Camptocormia

The earliest description of camptocormia was given by Brodie in 1818¹⁸. Different criteria have been used for the diagnosis of camptocormia but most of the authors have considered flexion (>45°) at thoracolumbar joint while walking or standing and subsiding in lying down position^{3,6}. The commonest symptoms of camptocormia is back pain which aggravates on walking or standing position. Prevalence of camptocormia among PD patients have ranged

from 3-17.6 per cent^{6,19-21}. In a study of camptocormia patients, 68.8 per cent had a diagnosis of PD²². It has been seen that camptocormia is positively associated with disease severity and duration with average duration being 7-8 yr. In a recent study of 531 PD patients from Japan 22 had camptocormia which was associated with disease severity, higher L-DOPA usage and autonomic symptoms such as constipation and urinary frequency (Table II)¹⁰. PD patients with camptocormia should be investigated with radiography and associated degenerative spondyloarthropathy should be ruled out if there is fixed deformity.

Striatal hand and striatal toe (Figs 2 and 3)

Striatal hand deformities include flexion at metacarpophalangeal, proximal interphalangeal and distal interphalangeal joints. Striatal foot deformities are characterized by extension of great toe and plantar flexion of other toes. There may be associated claw like or talipes equinovarus like deformities. Initially these deformities may be non-fixed but later on these become fixed. Striatal deformities might be present in up to 10 per cent of untreated cases of advanced PD and these are more common in early onset or younger PD patients (PARKIN mutation)²³. These deformities may be responsive to levodopa and may be a part of



Fig. 2. Striatal deformity of left hand characterized by flexion at metacarpophalangeal joint in a Parkinson's disease patient (71 yr old male).



Fig. 3. Striatal deformity of left foot characterized by big toe lateral flexion in a Parkinson's disease patient (66 yr old female).

wearing off dystonia. Misdiagnosis of these deformities as rheumatoid or some other types of arthritis is very common.

Pathogenesis

Pathogenesis of postural and striatal deformities in Parkinson's disease is a combination of ongoing parkinsonian symptoms (rigidity and slowness), dystonia, musculoskeletal changes, loss of postural reflexes and influence of dopaminergic medications (Fig. 4). Duration of disease, amount of rigidity, female sex (possibly due to role of oestrogen) and family history of PD have been considered as important risk factors⁶. It has been found that PD patients on DOPA agonist develop contracture earlier²⁴. It is not clear whether treatment with levodopa has any role in the pathogenesis or not. Dystonic complication may be due to an imbalance between decrease in dopaminergic level and increase in GABAergic level²⁵. Animal models have helped to understand the pathogenesis. In monkeys where parkinsonism was induced by MPTP (1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine) foot dystonia was reported similar to PD patients²⁶. In a mouse model abnormal posturing was induced by 6-OHDA (hydroxydopamine) injection in the sensorimotor striatum²⁷.

Treatment

Every PD patient presenting with postural and striatal deformities should be evaluated regarding duration of symptoms, current dopaminergic medications and extent of disability. Important modalities of treatment are modification in

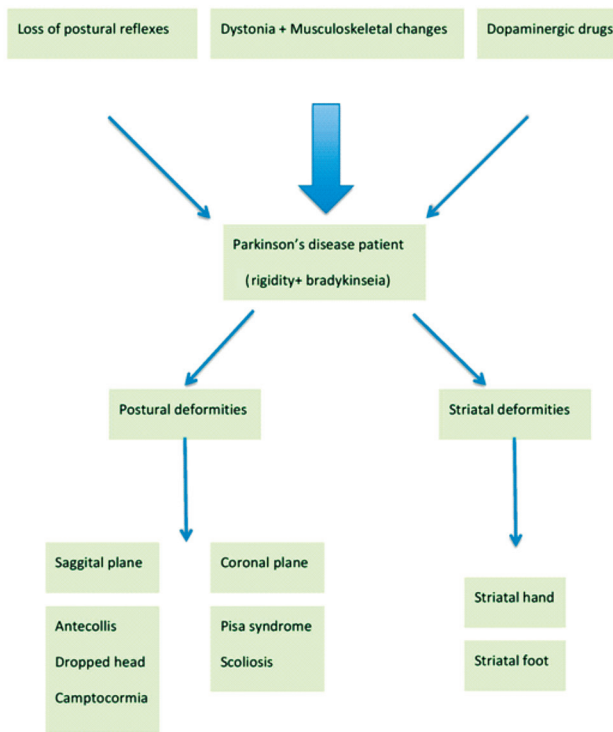


Fig. 4. Pathogenesis and spectrum of postural and striatal deformities in Parkinson's disease.

dopaminergic drugs, anticholinergic agents, botulinum toxin, neurosurgery, orthopaedic surgery, non-surgical corrections and physical-occupational therapy.

Antiparkinsonian drugs: Postural and striatal deformities are harbinger of advanced PD, but sometimes these can occur in early stage also. DOPA agonists should be avoided and adjustment in other dopaminergic medications may be helpful^{6,7}. Camptocormia is considered unresponsive to levodopa. Only a few reports of antecollis improving by levodopa are available, but results are not consistent^{3,6}. Anticholinergics can be used to control dystonic component leading to postural deformity (Pisa syndrome, scoliosis, and striatal deformity). Beclofen and benzodiazepines have also been found to be useful for striatal hand and foot dystonia⁴.

Botulinum toxin: Botulinum toxin acts by producing chemodenervation and local paralysis in the injected muscle due to presynaptic neuromuscular junction defect. This is helpful in alleviating dystonia which is an important component of postural and striatal deformity. Dosages and location of botulinum toxin injection is better guided by use of electromyography

and ultrasound technique. Paraspinal, rectus abdominis and iliopsoas muscles are usually injected for camptocormia. In Pisa syndrome paraspinal muscles and in antecollis neck muscles are site of injections. For striatal hand deformity injections in finger flexors and lumbricals have been found to be quite effective⁴. In a study on 14 patients of dystonic clenched fist which included seven PD patients who were treated with botulinum toxin injection, all had significant improvement in pain and muscle relaxation leading to better quality in life²⁸.

Neurosurgery: Bilateral deep brain stimulation (DBS) has been used for management of camptocormia, antecollis and Pisa syndrome. Initial cases included PD patients with camptocormia who had globus pallidus internus (GPi) and subthalamic nucleus (STN) stimulation. Postural abnormality is improved due to improvement in dystonia and rigidity of paraspinal muscles²⁹. In another study seven patients (3 dystonia, 3 PD and 1 dystonia and myopathy) having camptocormia underwent DBS surgery³⁰. Two PD patients had bilateral STN DBS and one had GPiDBS and all three had 55 per cent improvement in their off/on unified Parkinson's disease rating scale (UPDRS) in a mean follow up of 21 months (range, 12-36 months). Zona incerta and pedunculopontine nucleus are being explored as an alternative target for postural deformities. Combination of PPN and standard target such as STN may be another option³¹.

Orthopaedic surgery and non-surgical intervention: Different types of surgical corrections have been tried in striatal and postural deformities in Parkinson's disease. Spinal corrective surgery is helpful in camptocormia and Pisa syndrome. In a study two patients who had PD with severe camptocormia showed significant benefit with surgery³². On the contrary, in a case series of PD patients undergoing spine surgeries 86 per cent had a need for revision surgeries following the first procedure³³. Spinal braces and neck collar may be helpful for camptocormia and antecollis. Tendon transfer and Z-lengthning procedure may be helpful in striatal deformities not responding to dopaminergic medications⁴. Rehabilitation is another integral part of management of PD patients with postural and striatal deformities. Lating postures applied on shoulders and passive relaxation is helpful in reducing scoliosis³⁴.

Ongoing research

Most of the ongoing research on postural and striatal deformities in PD are related to pathogenesis

and treatment. Newer development in neuroimaging has helped in better understanding of pathogenesis of paraspinal and non-paraspinal muscles³⁵. Treatment related research is primarily related to deep brain stimulation surgery and repetitive trans-spinal magnetic stimulation (r-TSMS) which have been found to be effective in treatment of camptocormia in PD patients¹¹. Another ongoing investigation in the management of camptocormia is lidocaine which can be injected ultrasound guided in the abdominal external oblique muscles³⁶.

Conclusion

Postural and striatal deformities are common in PD patients but these are under-reported. These deformities lead to increased incidence of unsteadiness and falls in these patients. Younger age of onset and duration of symptoms are most important risk factors. Early diagnosis and treatment is important as majority may respond to adjustment in dopaminergic medications. Other patients will require botulinum toxin injection, deep brain stimulation, orthopaedic surgical interventions and rehabilitative therapies.

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