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Commentary

Understanding of skeletal deformities in Parkinson's disease

Abnormal posture and skeletal deformities of limbs, neck and trunk are common in patients with parkinsonism including Parkinson's disease (PD), multiple system atrophy (MSA), and progressive supranuclear palsy (PSP). These deformities include striatal deformities, camptocormia, Pisa syndrome, antecollis and scoliosis. They interfere with activities of daily living, resulting in postural instability, gait problems, and increased falls[1]. Though some of these deformities are caused by ongoing parkinsonian symptoms or are associated with nigrostriatal dopamine deficiency, especially in scoliosis[2], the exact mechanism is poorly understood. The true incidence or prevalence of these deformities in unknown, especially in Asia including India. The study by Pandey and Kumar[3] in this issue provides some information on striatal and postural deformities in Indian patients with PD. Though this study has a few limitations with a small number of participating patients, it provides important data on Asian patients, especially Indian patients.

There are some interesting results in this study[3] compared to a previous study[1]. First, 48.6 per cent of 70 patients with PD were reported to have either striatal or postural deformities. This rate was higher than the results of 33.5 per cent of 164 PD patients in America in a previous study[1]. Striatal foot was the most common deformity in the present study[3] as well as in other studies[1,2]. Second, the female to male ratio (2.04:1) in this study[3] was higher than that reported (1:1) in a previous study[1]. Third, although the difference was not significant in the mean age of onset, but PD patients with deformity were older than those without deformity (53.5 vs. 49.5 yr)[3]. This finding was different from that (54.7 vs. 62.5 yr) of a previous study[1]. Finally, patients with scoliosis had a contralateral deformity in relation to initial PD symptoms.

Usually, striatal limb deformities can be seen in patients with advanced PD, although these might also be seen in the early stage of PD and other parkinsonian disorders[4-6]. Striatal limb is characterized by flexion of the metacarpophalangeal (MCP) joints, flexion of the distal interphalangeal (IP) joints, ulnar hand deviation in the hand, and great toe extension, flexion of the remaining toes and equinovarus foot positioning in the foot[7]. Foot deformities are more frequent in PD patients as part of the disease or as "wearing off" dystonia associated with levodopa therapy than hand deformities. According to a previous study[8], the percentage of striatal foot and hand deformities is up to 10 per cent in untreated patients with advanced PD. Foot deformities can develop in 20-40 per cent of PD patients receiving sustained levodopa treatment[9]. Furthermore, as the earliest signs of striatal hand, MCP flexion and IP extension can be seen, due to contraction of lumbricals and interossei, respectively[10].

Abnormal postures of trunk such as camptocormia, antecollis, Pisa syndrome and scoliosis are common in PD patients. Majority of the patients with camptocormia have a combination of rigidity and dystonia[11]. The main pathophysiological mechanism might be due to increased susceptibility of the paraspinal muscles to injury in the setting of kyphotic postural changes and age-dependent loss of tissue elasticity in PD patients[12]. In the study of Pandey and Kumar[3], camptocormia and antecollis were seen in 20 and 7.14 per cent PD patients, respectively. These were associated with disease severity and longer duration of levodopa therapy. These rates were higher than those reported in previous studies[1,13]. Pisa syndrome also known as pleurothotonus is defined as marked lateral flexion of the trunk and head to one side with axial rotation

of the torso[2]. Because the underlying pathology may be related to cholinergic excess, this syndrome often occurs either as a result of decreased breakdown of acetylcholine or as a result of decreased dopaminergic inhibition of acetylcholine secondary to dopaminergic antagonism or dopaminergic depletion[14]. This syndrome is not usually associated with age, disease duration or the severity of the disease. It was found in about seven per cent of PD patients in the present study[3]. Scoliosis is defined as a lateral curvature of the spine with vertebral rotation that leads to asymmetric deformity of the trunk. It occurs more frequently in PD patients than in the general population. Previous clinical and animal studies have reported that the direction of scoliosis is congruous with the laterality of major signs and symptoms of PD[15,16]. However, such findings remain controversial. These findings may suggest that scoliosis is closely associated with nigrostriatal dopamine deficiency. Baik *et al*[17] found 33 per cent of PD patients with scoliosis and the majority of them were women. They also found that patients with scoliosis were significantly older than those without scoliosis. However, there was no significant association between laterality and scoliosis. In addition, the occurrence rate of scoliosis was not different between *de novo* and levodopa-treated patients[17]. Although genetic, hormonal, biomechanical and neuromuscular factors have been proposed as possible causes of scoliosis in PD, the exact pathophysiology of PD is not yet fully understood[18]. In accordance with frequent dystonia in women with PD[8], scoliosis is known to occur more frequently in women than in men with PD[1,15,17] including the present study[3].

Though PD-related dystonia and skeletal deformities are known to have some risk factors, such as young age, female gender and long disease duration, the relationship of age, gender, family history or duration of L-dopa treatment with skeletal deformities in PD is not fully understood. Some skeletal deformities can be improved by botulinum toxin injection, especially in focal deformities such as striatal finger or toes[18]. However, for large muscle group involving deformities such as camptocormia and scoliosis, botulinum toxin injections could be less effective than focal deformities.

Because skeletal deformities can be seen in elderly persons easily, this can be underestimated in PD patients, especially at early stage. Sometimes, one of these deformities such as antecollis in early time can help differentiate the diagnosis between PD and MSA. Otherwise, to prevent permanent contractures, early diagnosis of skeletal deformities, from striatal hand to scoliosis, is important. Hence, if more study results on skeletal deformities in patients with PD are added in future, these will help us understand the clinical features, natural course and pathogenesis of these deformities. Based on these appropriate guidelines, early diagnosis and more accurate treatment will be achieved.

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