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Pisa Syndromes in Patients with Intellectual Disability

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Case Report: Pisa Syndrome in Patients with Intellectual Disability

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Abstract

The aim of this report is to describe Pisa syndrome in two adults with intellectual disability. This case report is significant as, to our knowledge, Pisa syndrome has not previously been reported in an individual with intellectual disability (learning disability or mental retardation). Pisa syndrome is a reversible condition characterised by sustained flexion of the head and trunk and usually caused by psychotropic medication. It may be challenging to identify this condition in individuals with intellectual disability who may have coexisting postural and mobility problems. Both patients developed sustained involuntary flexion of the trunk and head tilt (Pisa syndrome) due to psychotropic medications. Both patients were prescribed risperidone, and their condition improved as risperidone was tapered off and olanzapine was commenced. Extra vigilance is required to identify Pisa syndrome as a reversible side effect of psychotropic medication in people with intellectual disability who may have pre-existing mobility and postural abnormalities.

Key words: Pisa Syndrome, Intellectual Disability, Mental Health, Psychotropic Medication, Reversible Side Effects

Introduction

Pisa Syndrome (pleurothotonus) was first described by Ekbom in 1972 as a side effect of neuroleptic treatment, and defined as sustained involuntary flexion of the trunk and head to one side and slight rotation of the body so that the person appears like the “leaning Tower of Pisa” (Ekbom, 1972). It is differentiated from tardive dyskinesia on the basis of its responsiveness to the withdrawal of antipsychotic medication (Suzuki, 2002). Pisa syndrome has been mostly reported in association with prolonged use of antipsychotic medication; however, there are case reports of its occurrence in association with tricyclic or antiemetic medication, cholinesterase inhibitors, without any medication (idiopathic Pisa syndrome), and in those with neurodegenerative disorders such as Parkinson disease and Alzheimer dementia (Uemura 2008). Little is known about the pharmacological mechanism underlying this syndrome. Dopaminergic dysfunction has been implicated. A role for serotonergic and noradrenergic mechanisms has also been suggested (Suzuki, 1997).

A literature search failed to find any case reports of Pisa syndrome in people with intellectual disability (learning disability or mental retardation). Both of our patients have intellectual disability and developed Pisa syndrome, which responded to withdrawal of risperidone. Written consent was obtained from the next of kin for each patient as both lacked the capacity to consent for themselves. Ethical approval was obtained from the local ethical committee.

Case 1

The first patient is a 31-year-old man with severe intellectual disability of unknown aetiology and bipolar affective disorder. He also has upper thoracic kyphosis with deviation to the right side, which is not prominent and does not affect his movements and balance. He developed a marked, sustained, right tilt in his body and head with some upward tilt in his neck. He lost weight of 10 kgs over a period of 4 to 5 months, although his appetite did not change. There was no recent change in his medications before the onset of change in his posture. He was prescribed the following long-term psychotropic medications for bipolar affective disorder and management of his behaviour: risperidone, 6 mg twice a day; carbamazepine, 200 mg three times a day; diazepam, 10 mg three times a day; and procyclidine, 5 mg once a day.

The patient was assessed initially by his general practitioner and then a consultant physician to investigate the change in his posture and weight loss. The results of all assessments revealed no abnormalities that could explain the weight loss. After continuous tilt in his body and head for 4 months, improvement was noted for the first time when the dose of risperidone was reduced. The change in his posture disappeared when risperidone was discontinued. A retrospective diagnosis of Pisa syndrome was made in this case, as other causes for the tilt in his body had been ruled out, and his posture responded to reduction and discontinuation of the antipsychotic medication. When manic episodes recurred, he was prescribed olanzapine, and his condition stabilized on a dose of 10 mg twice a day. The carbamazepine was changed to a slow-release formulation for ease of administration, and the dose was adjusted to 600 mg twice a day. His weight also started to normalize slowly.

Case 2

The second patient is a 46-year-old woman with severe intellectual disabilities, living in one of the residential centres for people with learning disability in the West of Ireland. Her behaviour problems include pushing and hitting fellow service users and staff. She has had a depressive episode. Her prescribed medications included risperidone, 2.5 mg twice a day; chlorpromazine, 50 mg twice a day and 25 mg at lunchtime; sertraline, 100 mg once a day; and flurazepam, 30 mg at bedtime. She developed a sustained left tilt in her body and was holding her left hand in her right hand. She lost 8 kgs in 3 months. Her weight loss was associated with poor appetite, but the rapidity of the weight loss could not be explained merely by poor appetite.

On physical examination, no abnormality was found that could explain her weight loss. There was no loss of power in her limbs, and no other neurological deficit could be found. The results of all blood tests and computed tomography revealed no abnormality. The tilt in her body remained for almost 3 months until the dose of risperidone was reduced. With reduction in the risperidone dose, the lean in her body slowly improved. Risperidone was discontinued, and the patient was prescribed olanzapine for her behaviour. The lean in her body resolved, her appetite improved, and she started to gain weight. As in the first case, the diagnosis of Pisa syndrome was made retrospectively as all other possible causes for the tilt in her body had been ruled out, and improvement occurred with the reduced dose of risperidone.

Discussion

Pisa syndrome has been reported as common with organic brain syndromes and has been reported with neurodegenerative disorders such as Parkinson disease and dementia (Suzuki, 2002: 165, Harada, 2006: 771). No report, to our knowledge, describes Pisa Syndrome in people with intellectual disability.

We could not explain the weight loss in both cases, although the weight loss in one case was associated with poor appetite. In that patient, a depressive episode could not be ruled out, but the rapid weight loss could not be explained by only a depressive episode. In both cases, the weight loss improved, as did the symptoms of Pisa syndrome, when the dose of risperidone was reduced and then discontinued. The weight loss could be a side effect of risperidone, but both patients had been prescribed risperidone for a long time, and the weight loss coincided with the change in body posture. Although there is no reported association of weight loss with Pisa syndrome, both of our cases had coexisting weight loss that could not be explained otherwise. To establish an association of Pisa syndrome with weight loss, more evidence is needed. Both of our patients were prescribed olanzapine after discontinuation of risperidone, but there have been case reports where olanzapine was the cause of Pisa syndrome (Arora, 2006). Although risperidone was the cause of Pisa syndrome in our case, in one case report, it was used to treat a patient who developed Pisa syndrome (Harada, 2006: 771). From the reported cases, there is no evidence that one psychotropic is more likely than another to cause Pisa syndrome, and according to these reports, the psychotropic medication should be changed to an appropriate alternative when Pisa syndrome occurs.

Conclusion

Extra caution should be taken with patients who have intellectual disabilities when administering psychotropic medications where lack of communication and coexisting physical and movement disorders may mask the diagnosis of Pisa syndrome, a reversible condition.

Declaration of Interest: None

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