Sir,

Tourette syndrome (TS) is an inherited neurological disorder with onset below 18 years of age characterized by motor and vocal tics, occurring in 0.6–3% of schoolchildren.[1] The structural basis of TS is incompletely understood. The TS occurring in association with Neurofibromatosis type-1 (NF-1) is a rare feature.[2] However, such a case might provide useful insights into the structural basis of clinical features in TS.

A 12-year-old boy presented with multiple motor tics since the age of 5, involving the neck, trunk, upper limbs, and eyelids. The symptoms included eye blinking, arm thrusting, kicking, shoulder shrugging, and jumping. Repeated throat clearing was noted for 18 months. There was no history of seizure, behavioral change, or poor scholastic performance. There were no features to suggest an associated obsessive-compulsive disorder (OCD) or attention deficit hyperactivity disorder (ADHD). There was no similar family history. On examination, multiple café-au-lait macules and axillary freckling were present. Neurological examination was unremarkable.

Magnetic resonance imaging (MRI) of the brain showed T2W hyperintense lesions in bilateral basal ganglia [Figure 1] and thalami [Figure 2]. Electroencephalography was normal. The erythrocyte sedimentation rate was 19 mm/h. Serum ceruloplasmin was 96 mg% (normal > 30 mg%). Antinuclear antibody was negative. Serum complement was 80% and antistreptolysin O titer was normal.

The TS was diagnosed on the basis of established clinical criteria. The child showed significant improvement with risperidone 1 mg, taken twice daily. However, MRI abnormalities persisted even after 6 months.

On the basis of the etiology, TS is classified into two categories: idiopathic and symptomatic. Symptomatic TS can occur after head injury, encephalitis, stroke, rheumatic fever, and drug intake (methylphenidate, neuroleptics, opioids, antiepileptics).

The neuroanatomical localization of tics has remained a challenging feature. Given the wide clinical heterogeneity in TS and frequent association with OCD or ADHD, several neuroanatomical localizations have been evoked, including the cortex (frontal or temporal lobe), limbic system, basal ganglia, and brain stem. The most favored are the striato-thalamo-circuit and striatal compartments.[3] The MRI studies in TS have shown significant reduction in the volumes of caudate and lentiform nuclei. Our patient too had involvement of bilateral caudate and lentiform nuclei [Figure 1]. Therefore, one can speculate that any neurological disease with a significant involvement of caudate or lentiform nuclei or both could present with secondary TS. This argument was strengthened by a recent case of varicella zoster encephalitis with basal ganglia imaging abnormalities, where the patient developed a chronic tic disorder associated with ADHD.[4] In our patient, the basal ganglia abnormalities were related to NF-1, a feature that has been well described.[5] Hyperintense lesions in the basal ganglia in patients with NF-1 may regress over time in 40% of patients, and this might suggest that these lesions are due to de-myelination.[5]

In conclusion, TS can result from a variety of causes, NF-1 being an uncommon cause. Our case and data from previous reports point towards the pathological involvement of caudate and lentiform nuclei in the genesis of tics in TS. Tics in secondary TS too respond well to risperidone.

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Punding in Parkinson’s disease related to high-dose levodopa therapy

Sir,

A 69-year-old lady a case of idiopathic Parkinson’s disease (PD) was brought with one-month history of repeated and excessive arranging and counting of dresses. The patient spent a lot of time searching for clothes and dresses and meticulously arranging them according to their sizes. She would repeatedly count them. She would get up from sleep and repeat this act for several hours. On questioning, the patient agreed that her act of this was irrational and unproductive, however, expressed an inability to control her from doing so.

She was on levodopa for 13 years and the dose had been gradually increased to 1000 mg per day. It was discovered that she was taking an additional dose of 500 mg per day on her own. Other medications included bromocriptine 5 mg, selegeline 10 mg, and trihexyphenidyl 6 mg daily. The duration of levodopa effect had gradually decreased from initial six hours to one-two hours at the time of presentation. “Peak-dose” dyskinesias were also noted.

Clinical examination was unremarkable except for hyperreflexia. She had mild autonomic dysfunction but no dementia. Computerized tomography of the brain, electroencephalography, routine hemogram and biochemistry were normal. A possible diagnosis of punding and dyskinesias related to high-dose levodopa was made. Levodopa dose was reduced to 750 mg daily and amantadine 100 mg thrice daily was added. She had a good response and punding subsided within three days.

“Punding” is a stereotypical motor behavior in which there is an intense fascination with repetitive handling and examining of mechanical objects, such as picking at oneself or taking apart watches and radios or sorting and arranging of common objects, such as lining up pebbles, rocks, or other small objects. Punding differs from compulsions in that performance of these activities is not distressing to patients and it is only if the act is interrupted that any compulsive urge becomes apparent.

Punding is well known to occur in association with addiction to central stimulant drugs such as amphetamine or cocaine.

Punding on levodopa therapy was initially reported by Friedman about 10 years ago. However, there is not much data available on this topic yet. The previously reported cases were women in the age group 65-72 years, similar to our case. The duration of disease was 10-20 years and they were receiving levodopa at a dose of 500-1900 mg per day, which is similar to our case. Symptoms subsided after decreasing the dose of levodopa in all. Therefore, punding is thought to be related to excessive dopaminergic stimulation. The brain region most likely involved in mediating these effects involves mesolimbic dopaminergic projections and the nucleus accumbens.

Regional cerebral blood flow studies using positron emission tomography have shown that dopaminergic effects on orbitofrontal cortex via dopamine D3 receptors may in part cause punding and other complex dopamine-induced stereotypies.

Punding in PD is not a specific problem related to levodopa, as cases with punding have been described in association with quetiapine (an atypical antipsychotic) used for treating psychosis in PD.

In conclusion, one should be aware of the possibility of punding in patients with PD on long-term dopaminergic therapy, especially levodopa. As punding is not distressing to the patient, the history may not be volunteered. Failure to recognize this early may be a source of discomfort to the patient and carergivers, whereas early diagnosis can result in prompt relief of symptoms.

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References