report demonstrates a possible usefulness of olanzapine as an antipsychotic mono-therapeutic strategy for Pisa syndrome, even if the syndrome is triggered by other atypical antipsychotics such as risperidone.

Katsuji Nishimura, M.D.
Department of Psychiatry, Tokyo Women's Medical University School of Medicine, Tokyo, Japan
Yuri Mikami, M.D.
Institute of Women's Health, Tokyo Women's Medical University School of Medicine, Tokyo, Japan
Seiichi Tsuchibuchi, M.D.
Soshu Mental Clinic, Kanagawa, Japan
Naoshi Horikawa, M.D.
Department of Psychiatry, Saitama Medical Center, Saitama Medical School, Saitama, Japan

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Prader-Willi Syndrome

SIR: Prader-Willi syndrome is a chromosomal disorder that was first described by Langdon Down[1] in 1887 and was reported by Prader et al.[2] in 1956. They reported a series of patients with mental retardation, obesity, and hypogonadism.

In 1981, Ledbetter et al.[3] identified deletions of genes in the proximal arm of the paternal chromosome 15. Loss of gene material at the maternal chromosome 15 results in Angleman Syndrome. This pattern of inheritance—when expression of a gene depends on whether it is inherited from the father or the mother—is called imprinting.[4]

Prader-Willi syndrome is the first human disease attributed to genomic imprinting.[4] In the United States, rate of prevalence of the syndrome has been reported to be 1 in 16,000.[1] Differences in prevalence between racial groups, sex, and age have not been reported.[1] Although the syndrome is best known for its food-related characteristics, including hyperphagia and obesity due to decreased perception of satiety secondary to hypothalamic abnormalities,[6,7] it also involves other non-food-related characteristics, such as cognitive dysfunction, academic weakness, and behavioral problems.[5,7,8]

Some reports have noted that behavioral problems increase in adolescence and adulthood years because of growing psychosocial stresses.[9] Often, these maladaptive behaviors pose multiple challenges to the clinicians and heighten the stress of the patient's family.

Case Report

A 26-year-old man was diagnosed with Prader-Willi syndrome. Despite his mild mental retardation and delayed developmental milestones, he finished 12th grade of special education.

His elderly parents were unable to provide the essential care for his condition and requested housing support in form of group home setting. The patient was unable to cope with the changes and responded with disruptive behavior that led to his first psychiatric admission.

On the day of admission, the patient became agitated with little provocation and attempted to break the fish tank in his house; when his mother interfered, he threatened her.

In the emergency room, he appeared with the following physical features: obesity (weight: 285 pounds, height: 5 ft), a round face with full checks, narrow nasal bridge, down-turned mouth with thin upper lip, small hands and feet, and undescended testicles.

His characteristic profile included compulsive eating, hoarding food, argumentativeness, stubbornness, irritability, aggressive behavior, skin picking, and sleep apnea.

His electrocardiogram, complete blood count, and blood chemistry were within normal levels.

On interview, the patient was uncooperative, loud, screaming, punching the walls, slamming the doors, and demanding to leave the hospital. He denied suicidal and homicidal ideation. No psychotic features were elicited. He was medicated and was admitted to the developmental delay unit in a different hospital.

Comment

Unfortunately, diagnosis of Prader-Willi syndrome is overlooked, despite the clinical criteria. Management of the syndrome requires a multidisciplinary approach; treatment implications involve medical and dietary management as well as psychiatric intervention.

Nahla A. Mahgoub, M.D.
Department of Psychiatry, Beth Israel Medical Center, New York, NY

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Association of Familial Ataxia and Restless Legs Syndrome

SIR: Restless legs syndrome (RLS), characterized by an intense urge to move the lower extremities especially during the night, is associated with a number of medical conditions, such as uremia, iron deficiency, and neuropathy. RLS with a prevalence of 5% to 15%, is frequently underdiagnosed[1–3] and can be confused with anxiety states and other psychological syndromes, hyperthyroidism and drug-induced akathisia. Previous reports have highlighted an association between RLS and familial spinocerebellar ataxias (SCA) in German and Dutch patients.[2–5] However, the long-term association of these two conditions is unknown. The prevalence of RLS is much rarer among Asians,[6] and to our knowledge, RLS has not been reported in SCA patients in Asia.

Here, we report an Asian patient with SCA who presented with restless legs symptoms that were initially attributed to stress and anxiety. We examined the relationship between RLS and SCA symptoms in this patient over a few years to highlight the need to recognize their coexistence early since RLS could be effectively treated.

Case Report

The patient of Asian origin presented with progressive unsteady gait and frequent falls. He had signs of cerebellar dysfunction, generalized hyperreflexia and lower limb spasticity, and was confirmed to have autosomal dominant SCA type 3 (SCA3), with an abnormal *CAG* trinucleotide repeat expansion. One year after the onset of his ataxia, he complained of an urge to move his lower extremities, particularly when at rest. This was preceded by an abnormal sensation in his toes and feet, which at times extended to his knees, described as "crawling," "creeping," and "itching." He was troubled and anxious because the urge and sensation were often intense and distressing, and he had to move his legs and get up to walk to obtain relief. He was more aware of his symptoms during quiet time in the evening or night and when not distracted by other activities. He frequently woke up more than 10 times nightly because of sensation associated with restless legs and complained of poor sleep quality. His symptoms were initially attributed to his anxiety and depression and treatment for these symptoms was not effective. Subsequently, he was diagnosed with RLS based on the International Restless Legs Syndrome Study Group criteria.[5] After L-dopa was prescribed, his symptoms improved significantly. Investigations did not reveal a secondary cause for his RLS. Full blood count, urea/creatinine/electrolytes, and serum ferritin levels were normal. A nerve conduction test did not reveal any significant peripheral neuropathy. He was followed up over a 3-year period. His RLS symptoms appeared to progress with deterioration of his ataxia symptoms but could be controlled with L-dopa. His RLS symptoms would reemerge with withdrawal of the medication.

Comment

RLS was most likely related to the patient's ataxia condition as the patient did not have any past or family history of RLS, his symptoms came only after the onset of his ataxia, and the clinical severity of both diseases appeared to move in tandem over a long period of time. Investigations did not reveal any evidence of anemia, renal failure, peripheral neuropathy, or arthritis, conditions which may predispose to RLS symptoms.

Functional imaging studies have provided evidence implicating dopaminergic system impairment in RLS. Similarly, striato-nigral involvement has been demonstrated in neuropathological specimens of SCA3. Hence, it is possible that SCA3 and RLS may share a common pathophysiological mechanism. RLS and sensorimotor axonal neuropathy have been associated with intermediate *CAG* repeat size (53 and 54 repeats) in an SCA3 Dutch family.[5] Our RLS patient had an abnormal *CAG* repeat size (>60 repeats) and no neuropathy. Whether there is a correlation between the size of the *CAG* repeat expansion and the risk of RLS symptoms is not known.

RLS symptoms can be confused with stress and anxiety in patients with familial SCA. The long-term follow-up of our patient supports the association of RLS and SCA reported in Caucasian patients. We