Comment

What makes this case noteworthy, apart from the rarity of the syndrome and survival to adulthood, is the evolution of the psychosis, which is not a feature of Patau syndrome. The patient started to develop auditory hallucinations at the age of 15, for which she had multiple admissions and received different antipsychotic medications.

She started to decompensate after her antipsychotic medication was changed because of questionable seizures. She presented in the emergency room with agitated behavior. She was observed conversing with God, the devil, and her deceased siblings. She was delusional; she believed that people were taking her strength. The patient was medicated and admitted to the psychiatric floor. She was put on a regimen of valproic acid, 500 mg twice daily, aripiprazole, 20 mg daily, and olanzapine, 20 mg daily.

She was discharged to an assisted living facility after she stabilized. Since then, she has not had any hospitalization.

Aijaz Nanjiani, M.D.
Ashgar Hossain, M.D.
Bergen Regional Medical Center, Paramus, NJ
Nahla Mahgoub, M.D.
Beth Israel Medical Center, New York, NY

References

1. Patau K, Smith DW, Therman E, et al: Multiple congenital anomaly caused by an extra autosome. Lancet 1960; 1:790
2. Baty BJ, Brent L, Cary JC, et al: Natural history of trisomy 18 and trisomy 13, I: growth, physical assessment, medical histories, survival, and recurrence risk. Am J Med Genet 1994; 49:175–187
3. Baty BJ, Brent L, Cary JC, et al: Natural history of trisomy 18 and trisomy 13, II: psychomotor development. Am J Med Genet 1994; 49:189–194
4. Delatycki M, Garder R: Three cases of trisomy 13 mosaicism and a review of the literature. Clin Genet 1997; June:403–407

Pisa Syndrome Resolved After Switching to Olanzapine

SIR: Pisa syndrome, or pleurothotonus, is a rare dystonic reaction commonly associated with prolonged antipsychotic medication. The treatment of Pisa syndrome at present is empirical, reflecting a poor understanding of its underlying pathophysiology.[1] The first-line treatment for Pisa syndrome remains a reduction in dose or discontinuation of antipsychotics, and the second-line treatment is an anticholinergic medication.[1] Here, we present a case of Pisa syndrome that developed during treatment with risperidone. Although both therapies were ineffective, the problem was resolved successfully after the medication was switched to olanzapine.

Case Report

"Mr. A," a 29-year-old man with a 3-year history of DSM-IV schizophrenia, had no history of head trauma or other neurological problems, and had no family history of dystonia or other movement disorders. In October 1999, he was admitted to the psychiatric unit for 9 months because of a severe psychotic exacerbation resulting in self-injuries of the tongue with scissors. A regimen of bromperidol, a maximum of 27 mg/day, and biperiden, 3 mg/day, was implemented with consequently good results. In the outpatient clinic, bromperidol was switched to risperidone at 4 mg/day, and then biperiden was discontinued.

In January 2001, 4 months after the initiation of risperidone, Mr. A was observed walking with a tilt toward the left. Physical examination showed tonic flexion of the trunk toward the left along with a slight backward axial rotation, classically referred to as Pisa syndrome. No evidence of other extrapyramidal symptoms was found. Secondary dystonias resulting from metabolic disorder, organic disorder, or infection were ruled out. The regimen of trihexyphenidyl was attempted for 4 weeks but no benefit was seen. A regimen of risperidone, 4 mg/day, was then reduced, step by step and cautiously, to 1 mg/day in 2 months. No improvement in Pisa syndrome was observed after 2-month observation with this dose. Because of the risk of psychotic relapse, risperidone, 1 mg/day, was successively switched to olanzapine, 5 mg/day, despite its discontinuation in August of 2001. Pisa syndrome gradually improved, and the symptoms disappeared within 2 months. Two years after olanzapine treatment, the patient was found to be in remission from the psychosis with no signs of Pisa syndrome.

Comment

The substitution to atypical antipsychotics, particularly clozapine, may provide alternatives for the treatment of patients with tardive dystonia.[2] Olanzapine, which has pharmacological similarities to clozapine, has been also suggested to be effective in the treatment of tardive dystonia.[3] For the treatment of Pisa syndrome, the usefulness of atypical antipsychotics has been mentioned only in a few case reports on clozapine[4] and amisulpride.[5] In this case, it remains unclear whether the clinical resolution of Pisa syndrome represents an antidystonic effect of olanzapine or simply a spontaneous remission of Pisa syndrome after the withdrawal of the offending drug, risperidone. Similar claims have been made for clozapine in the treatment of tardive dystonia.[4]

Although the pathophysiological mechanism is still unclear, this case

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

References

1. Suzuki T, Matsuzaka H: Drug-induced Pisa syndrome (pleurothotonus): epidemiology and management. CNS Drugs 2002; 16:165–174
2. Tarsy D, Baldessarini RJ, Tarazi FI: Effects of newer antipsychotics on extrapyramidal function. CNS Drugs 2002; 16:23–45
3. Lucetti C, Bellini G, Nuti A, et al: Treatment of patients with tardive dystonia with olanzapine. Clin Neuropharmacol 2002; 25:71–74
4. Diez-Martin JM: The Pisa syndrome as another acute dystonia subtype. J Drug Dev Clin Pract 1995; 7:59–61
5. Ziegenbein M, Schomerus G, Kropp S: Ziprasidone-induced Pisa syndrome after clozapine treatment. J Neuropsychiatry Clin Neurosci 2003; 15:458–459

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

References

1. Down JL: Mental Affections of Childhood and Youth. London, Churchill, 1887