Depression With Suicidal and Homicidal Ideation in Turner Syndrome

Neda Motamedi, MD[a,*]; Bibiana Susaimanickam, MD[a]; and Amir Riahinezhad[a]

Turner syndrome is a genetic syndrome that can present with a plethora of physical and psychiatric manifestations. It is caused by a genetic deformity in which the cells of the body only have 1 copy of the X chromosome (monosomy X) instead of the usual 2 X chromosomes in females. Turner syndrome is named after Henry H. Turner, MD, who, in 1938, was one of the first doctors to report on this disorder in the medical literature.[1] This condition occurs in about 1 in 2,500 newborn girls worldwide, but it is much more common among pregnancies that do not survive to term (miscarriages and stillbirths). Some of the common physical conditions are having short stature, losing ovarian function, and not undergoing puberty unless they receive hormonal therapy. Furthermore, a percentage of females with Turner syndrome have extra folds of skin on the neck, a low hairline at the back of the neck, puffiness of the hands and feet, and skeletal abnormalities in addition to their short stature.[2] In addition to physical obstacles, women with Turner syndrome also suffer from cognitive difficulties. Individuals with Turner syndrome exhibit relatively normal verbal IQ, but they possess weaknesses in visual-spatial and executive areas of their cognitive profile.[3]

This failure to sexually mature at the same time as their peers and lack of fertility due to ovarian complications can have deep psychological effects relating to one's self-image and femininity.[4] Women with Turner syndrome usually experience fewer romantic and sexual attachments and significantly lower occupational and academic achievement, even with similar verbal IQs.[4]

Several studies[3] suggest that girls with Turner syndrome have difficulty with processing emotion and social cognition. All of these factors put individuals with Turner syndrome at high risk for depression.[5] Adolescents and adults with Turner syndrome are at risk for depression, and adulthood appears to be the period of highest risk. Studies[5] in the last 12 years show consistently more severe depressive symptoms in individuals with Turner syndrome than in previous years.

A "Turner syndrome personality," characterized by excessive dependence, immaturity, depressiveness, passivity, distractibility, and docility, has been suggested by Nielsen and Thomsen.[6] No defined psychiatric condition has been traditionally related to Turner syndrome, and it is not mentioned in the *DSM-5*. *Kaplan & Sadock's Synopsis of Psychiatry*[7] only mentions Turner syndrome as a side note on intersex conditions and not in any psychiatric context.

Abnormalities in quality of life and cognitive measures have been observed in women with Turner syndrome, and a relationship between these phenomena and chromosomal constitution has been suggested. In contrast, few studies have systematically evaluated the presence of mood and behavioral syndromes in these women. The aim of this report is to explore the psychiatric manifestations and comorbidities in patients with Turner syndrome.

Case Report

The patient was a 28-year-old single, bisexual woman, who was unemployed and homeless but currently living with her brother and mother. She had a history of depression since 2015, and a medical history of seizure disorder, hypothyroidism, and Turner syndrome.

The patient was diagnosed with Turner syndrome at the age of 12 years at a community hospital to which she presented for evaluation of amenorrhea. There was no medical history found from age 12 to 21 years, and no additional information was provided by the patient or her family. In 2013, she presented to the BronxCare Endocrinology and Obstetrics and Gynecology Clinic and was found to have minimal complications thus far with no cardiovascular issues, sensorineural hearing impairment, hypothyroidism, or metabolic syndrome but definitely was at risk for osteopenia base on dual-energy x-ray scan. She reported moving to the Bronx for the change of care. She had attained menarche at age 21 years and was prescribed oral contraceptive pills and vitamin D but could not take the contraceptive pills due to lack of insurance, which was later reinstated. She was diagnosed with hypothyroidism the following year and started on medications. She had several visits with endocrinology and obstetrics and gynecology from December 2013 to January 2016. She had no follow-up after those visits and started to see endocrinology and obstetrics and gynecology after her admission to the inpatient psychiatry unit and outpatient referral was made to reconnect with care. She was diagnosed with seizure disorder in 2017 and started on an antiepileptic agent in outpatient neurology and had intermittent follow-up with neurology as well.

[a]BronxCare Health System, Icahn School of Medicine at Mount Sinai, Bronx, New York

*Corresponding author: Neda Motamedi, MD, BronxCare Health System, 1276 Fulton Ave, Bronx, NY 10456 (n.motamedi74@yahoo.com).

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The patient had 4 psychiatric emergency department admissions from February 2016 to May 2017, 1 inpatient admission in the past 5 years to a Brooklyn hospital for 15 days because of depression, and 3 admissions to the BronxCare Hospital in 2019 within 2 months in the context of depression and suicidal thoughts.

The patient attempted suicide in summer 2018 and October 2019 with medication overdose. She has no history of violence. She smokes cigarettes and marijuana every day and drinks alcohol twice daily.

She endorsed being sexually abused when she was a babysitter by the brother of the family in 2013 and by a stranger in 2014. She did not report any of the incidents to the police. Since 2014, the patient had been living between the shelter system and her mother's home but was currently living in an apartment with her brother and mother. She dropped out of high school in the 11th grade due to anxiety. She is bisexual and has not been sexually active since November 2018. Her last relationship with a boyfriend was in 2014. She broke up with her girlfriend in 2019. Her last job was in 2016. Psychological testing done during her psychiatric inpatient admission showed that her IQ as measured by the Wechsler Adult Intelligence Scale[8] was within the borderline range of functioning, consistent with her test of premorbid functioning. Her verbal comprehension skills, perceptual reasoning skills, and processing speed were borderline. Memory was a particular area of weakness, and it was in an extremely low range.

First Admission: The patient was brought in by her brother, as her depression was worsening in the context of noncompliance with her medications. She stated that she was depressed due to multiple stressors such as homelessness and a history of being molested and raped. She also reported having intermittent thoughts of suicide without a plan. She was then treated with antidepressants with improvement. She was scheduled for outpatient follow-up, and a care coordinator was assigned upon discharge.

Second Admission: The patient presented with worsening of depression and suicidal ideation. She reported that since her discharge 2 weeks ago she had been feeling depressed and suicidal with a plan to jump out of the window. She also had been noncompliant with medications and had been smoking marijuana after discharge from the hospital. She felt more suicidal when she had thoughts of being lonely.

She also endorsed that she heard voices. Her activities of daily living were poor, and her speech was slow and soft and was incoherent at times. She was treated with an antidepressant, and an antipsychotic was added to her treatment with improvement in symptoms. Psychological testing was also done during this admission.

The patient was scheduled for outpatient follow-up and treatment. In addition, referrals were made for a day program and a mobile crisis team that could check in on the patient.

Third Admission: The patient was brought to the hospital by emergency medical staff activated by her therapist due to suicidal ideations and overdose on levetiracetam and an antidepressant. She reported paranoid thoughts such as people were watching her and knew everything about her life, which made her uncomfortable, sad, and depressed and pushed her to want to kill herself. She reported that 2 weeks ago, she heard voices telling her that she is hopeless and to kill herself. She also complained of flashbacks of when she was raped. She was having suicidal ideation of jumping from the window or jumping in front of a train if she were to be discharged. During this admission, she also spoke about gender dysphoria, feeling she is more masculine and being trapped in a female body. She reported feeling worthless and that she was a burden to her family. She also revealed that her ex-girlfriend is now in a romantic relationship with her sister, and this was causing conflict and anger. She endorsed homicidal ideation against her ex-girlfriend, but she did not have any specific plan. She was treated with 2 antidepressants and a long-acting injectable antipsychotic, which improved her symptoms. She was discharged to partial hospitalization after a high-risk assessment and safety plan was done.

Discussion

Women with Turner syndrome can manifest several psychiatric diagnoses, which can affect their social functioning and employment. The presented patient was diagnosed with recurrent and severe major depressive disorder with psychotic features, anxiety disorder, substance use disorder (cannabis and nicotine), borderline range of functioning IQ, borderline verbal comprehension skills, borderline perceptual reasoning skills, and weakness in processing speed memory. These diagnoses correlate with the finding that in women and girls with Turner syndrome, an increased prevalence rate of attention-deficit/hyperactivity disorder and autism has been reported.[9,10] Several studies also show more self-reported shyness, anxiety, and depression symptoms than in controls[11] and impaired social competence, although women with Turner syndrome generally seem to cope well with life.[12] Individuals with a wide diversity of disorder of sex development (DSD) conditions can experience similar mental health issues. Emotional difficulties are expected in all conditions because having a chronic condition can be stressful in general. More unexpectedly, neurodevelopmental difficulties also occurred in the nonchromosomal DSD conditions, a finding that is important for clinicians in DSD care.[13] Clinicians should also be aware of the relevance of shame, self-esteem, body image, and satisfaction with care for mental health in people with DSD conditions. Individuals with different DSD conditions have shared that they easily have feelings of shame, and developing a healthy self-esteem and positive body image can be challenging. Although DSD conditions cannot be cured, building resilience and coping with their previous issues will substantially improve the mental health of persons with DSD.[13]

Gender dysphoria can have adverse effects, especially romantically. Our patient's romantic life was further complicated due to her sexuality, gender dysphoria, and

complicated relationship dynamics. Her ex-girlfriend's relationship with her sister meant that it was unlikely that she could get emotional support from her family due to the nature of the conflict. All these psychosocial factors provide further insight needed to help patients with DSD.

Patients with Turner syndrome need multidisciplinary management and psychosocial care. This care should be an integrated part of management with these patients. Several countries have already implemented a program with centralized adult Turner syndrome clinics. These countries include France, Denmark, the Netherlands, Sweden, parts of England, and possibly other countries. This integrated management should ensure the availability of high-quality multidisciplinary care for all women with Turner syndrome to treat and detect all the conditions associated with Turner syndrome, which typically appear at odd times during the lifetime of these women.[14] Turner syndrome patients need care, especially during the transition from childhood to adult. When a child with Turner syndrome has finished treatment in the pediatric department, usually consisting of growth hormone treatment for promoting linear growth and induction of puberty to ensure appropriate development of secondary sex characteristics and neurocognitive development in line with peers, transitioning to an adult caretaker will take place. This sounds like an easy task; however; in relation to Turner syndrome, this transition has proven to be troublesome, and several studies[15–17] have documented that many patients "disappear" in an apparent gap between pediatric and adult care.

The multiple psychiatric comorbidities, as in our case, in addition to poor social support, dysfunctional family dynamics, poor social functioning with low self-esteem, and lack of coping mechanisms make treatment of such patients challenging and lead to rehospitalization and poor response to treatment.

Continuity of care, access to services, and multidisciplinary management play an important role in meeting the needs of a patient with Turner syndrome, especially those with intellectual disabilities. In this case, there were lapses in care during all stages of the patient's life, including childhood and transition to adulthood. Her family was from a lower socioeconomic status, and they were not aware of her issues. She had impaired continuity of care during adolescence because of lack of family involvement and insurance issues. She did not receive enough medical and mental health care because her mother was not aware of her needs and was unable to support her financially and emotionally.

She had no psychological testing done in school to detect her borderline IQ. Hence, she was not connected to the Office for People With Developmental Disabilities and could not obtain appropriate services and adequate education to develop skills to prepare her for the future. Unfortunately, she dropped out of high school, most likely because of her mental illnesses, intellectual disability, and borderline IQ. Thus, she was unable to find a job and be independent in the community. She was unemployed for many years and was not aware of the possibility to apply for social security income/social security disability to be supported financially, as no one educated her enough to understand her disability situation. She was homeless and also utilized substances to cope with her issues.

After every discharge from inpatient psychiatry, she received more social services and outpatient support in the form of a care coordinator, a mobile crisis team and day program, and partial hospitalization to keep her connected to medical and mental health care and to help her comply with treatment. She was also reconnected with outpatient medical services. Currently, she is medically and psychiatrically stable in the community and following up in the outpatient departments.

Conclusion

Suicide is the second most common cause of death in adults. Patients with Turner syndrome, who are more vulnerable due to their cognitive and physical limitations, pose increasing challenges for clinicians to identify these psychiatric issues early and to manage them. These patients are at risk, which makes multidisciplinary management and psychosocial care extremely important to manage their well-being.

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