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CASE REPORT

Medicine Science International Medical Journal

Medicine Science 2018;7(2):446-7

Psychiatric aspects of 47, XYY (Jacobs) syndrome: A case report

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Received 30 November 2017; Accepted 29 January 2018
Available online 17.05.2018 with doi: 10.5455/medscience.2017.07.8802

Abstract
In this case, psychiatric aspects of 47, XYY syndrome is discussed in a male adolescent patient diagnosed with Attention Deficit Hyperactivity Disorder (ADHD), conduct disorder (CD), mild intellectual disability, and Tourette syndrome. A 13-year-old boy was diagnosed with ADHD, CD, mild intellectual disability, Tourette syndrome, and 47, XYY syndrome. As in our case, the presence of the XYY syndrome with Tourette syndrome, ADHD, CD and mild intellectual disability is the first demonstrative case. As it is known that XYY syndrome can often lead to neurodevelopmental disorders, genetic counseling can be provided when more than one neurodevelopmental disorders are detected as comorbid.

Keywords: 47, XYY Syndrome, neurodevelopmental disorders, tourette syndrome, psychiatry

Introduction

47, XYY syndrome, also known as Jacobs syndrome, is a syndrome caused by the insertion of a male Y chromosome to 46, XY. It occurs in 0.1 % of the male population . This syndrome has some physical traits and behavioral features. As a physical property, male tallness rarely is seen. Behavioral features include developmental delays, speech-language disorders, cognitive impairments, social-emotional difficulties. Despite the fact that in history 47, XYY syndrome was associated with aggression and violent crime, this claim is now disproved. The association of this syndrome with behavioral problems and developmental retardation is quite diverse. Although this syndrome is a known disease for a long time, the relationship between 47, XYY and neurodevelopmental disorders has been recently detected [1]. It has shown a relationship between XYY syndrome and neurodevelopmental psychiatric disorders such as attention deficit hyperactivity disorder (ADHD), autism spectrum disorder (ASD), intellectual disabilities, specific learning disorders, and tic disorders. Also, conduct disorder (CD) and temper tantrums can be associated with 47, XYY [2,3]. In this case report, the psychiatric aspects of 47, XYY syndrome is discussed in a male adolescent patient diagnosed with ADHD, CD, mild intellectual disability and Tourette syndrome.

Case report

A 13-year-old boy admitted to our outpatient clinic with his mother. The patient said that he often winked his eyes and shrugged his shoulder. He started clearing his throat. The movements and voices were more severe, when he was worried. Before winking the eyes, he felt as if itching of the eye. Sometimes he could stop the tics voluntarily for 1-2 hours. He did not make any movements or voices when he was busy with his work. The tics occasionally happened, passed, and started again. From his mother's knowledge, he was a sixth-grader, but he was trying to read and write by combining the letters and do basic mathematic operations with finger counting. The patient immediately forgot what he learned, still did not have bladder control. The child had difficulty paying attention, was overactive, quarreled with his friends, swore a lot, experienced anger attacks. Also, he had repetitive movements like arm movements and eye movements, voiced improper sounds. Movements and sounds had non-rhythmic characteristics. According to the information received from his teacher, it was learned that he lagged behind his peers in learning, could not sit at his desk for longer than five minutes, mostly was overactive, told a lot, broke the order of class and made strange movements and sounds. The patient was on a special education program for four years. In his psychiatric evaluation, he was fully conscious and cooperated. His appearance was younger than his age. The mood state was euthymic, but his affection was inappropriate. During the interview, he seemed inattentive, preoccupied and fidget, had meaningless smiles. Judgment and abstract thinking were retarded compared to their peers. It was revealed that motor and vocal tics started about 6 months ago and were seen simultaneously, his developmental milestones completed later than his peers. Stanford-Binet Intelligence scale score was 61 as compatible with mild Intellectual Disability. Turgay ADHD scales filled by parents and teacher. The results were consistent with ADHD and CD. Total Tic Severity Score of Yale Global Tic Severity Scale

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(YGTSS) and Total Yale Global Tic Severity Scale Score were 21 and 51, respectively. Blood tests were normal, and there was an asymmetry between the bilateral ventricles in the supratentorial sections of brain MR image. After the clinical interview and applied psychometric measures, according to Diagnostic and Statistical Manual of Mental Disorders (DSM-5), the patient was diagnosed with ADHD, CD, mild intellectual disability, and transient tic disorder. While atomoxetine was preferred for ADHD treatment, risperidone was started for tic disorder and CD. The drugs were gradually increased to effective dosage. He was treated with atomoxetine 40mg/day, risperidone 0,75mg/day. It was determined, with the current treatment of the patient, that the tics improved, the attention period increased, and the mobility decreased. Because of his intellectual disability, genetic tests were requested. The fragment analysis for Fragile X was negatively detected, whereas the chromosome analysis revealed 47, XYY. In addition to the existing diseases, the patient was diagnosed with 47, XYY syndrome. The patient's treatment was regulated and medical follow-up continued. In the follow-ups, seven months later, tic attack started again, risperidone dosage was increased to 1 mg/day. The patient benefited from the treatment. Tourette's syndrome was diagnosed because the tic disorder continued longer than 1 year.

Discussion

Neurodevelopmental disorders are defined as the deteriorations of the growth and development of the central nervous system. There are some impairments in brain functions, mostly learning, memory, emotion, self-control skills. Neurodevelopmental diseases arise from the complex effects of genetic and environmental factors. In a study conducted in the United States, neurodevelopmental disease was detected in 15% of children [4]. Neurodevelopmental disabilities include ADHD, ASD, learning disabilities, intellectual disability, tic disorders and sensory impairments. Neurodevelopmental disorders are often accompanied by another neurodevelopmental disorder or psychiatric disorders. ADHD is the most common neurodevelopmental disorder [5].

The prevalence of ADHD is 5,3% of children in the world [6]. ADHD is frequently comorbid with diseases such as oppositional defiant disorder (ODD), conduct disorders (CB), intellectual disability, special learning disorders, tic disorders, mood disorders, adjustment disorders and substance use disorders [7]. The coexistence of ADHD, Tic disorder and intellectual disability in our patient confirms that neurodevelopmental disorders often occur together.

47, XYY syndrome is a genetic syndrome characterized by an additional copy of a male Y chromosome, usually not hereditary. Prominent behavioral characteristics, without physical changes, are seen in this syndrome. These behavioral characteristics, known as neurodevelopmental diseases include intellectual disabilities, ADHD, ASD, speech-language disorders, specific learning disorders, and tic disorders. These neurodevelopmental disorders also frequently co-occur in this syndrome [8]. In our case, ADHD, CD, Tourette syndrome and intellectual disability coexisted as neurodevelopmental disorders. In addition to these disorders, the detection of XYY karyotype in our case indicates the probable relationship of 47, XYY genetic syndrome and neurodevelopmental disorders.

In the literature, there was a case report that showed the association of ADHD, CD and mild mental retardation with XYY syndrome [2]. Solely, Tourette syndrome, with the 47, XYY syndrome, was reported in an another previous case [9]. However, as in our case, the presence of the XYY syndrome with Tourette syndrome, ADHD, CD and mild intellectual disability is the first demonstrative case.

Conduct disorder is associated with ADHD at around 50%. CD symptoms present in our patient may be related to this association [7]. Patients with ADHD and CD diagnoses may be prone to crime [10]. Although it is stated that the tendency to commit crime of 47-XYY patients are not accepted any longer [1]; committing crime tendency shown in some of the 47-XYY patients may be due to the accompanying symptoms of ADHD and CB.

Conclusion

The association of these diseases may be due to their neurodevelopmental origins. As it is known that XYY syndrome can often lead to neurodevelopmental disorders, genetic counseling can be provided when more than one neurodevelopmental disorders are detected as comorbid. The patients with neurodevelopmental disorders, it should be kept in mind, might have the XYY syndrome.

Competing interests

The authors declare that they have no competing interest.

Financial Disclosure

The financial support for this study was provided by the investigators themselves.

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