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Case Report / Olgu Sunumu

# Case of Klinefelter's Syndrome in an 18 Year Old Male Presented with Antisocial Personality Disorder

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**ABSTRACT**

Case of Klinefelter's syndrome in an 18 year old male presented with antisocial personality disorder

Klinefelter's syndrome (KS) is the most common chromosomal aberration among men and is associated with multiple psychiatric comorbidities. Individuals with KS have an increased prevalence of psychiatric disturbances, ranging from attention deficit and hyperactivity disorder (ADHD) in childhood to schizophrenia and severe affective disorders during adulthood. We want to present a case report of KS who presented with antisocial personality disorder and related disorders which are ADHD, conduct disorder and alcoholism. The common feature of all these disorders may be associated with X-linked chromosomes.

**Key words:** Antisocial personality disorder, Klinefelter's syndrome

**ÖZET**

Antisosyal kişilik bozukluğu ile karşımıza çıkan 18 yaşında Klinefelter's sendromu tanısı alan bir erkek olgu

Klinefelter sendromu (KS) erkeklerde en sık görülen kromozomal bozukluktur ve birçok psikiyatrik ek tanı ile ilişkilidir. KS olanlarda çocukluk çağında görülen dikkat eksikliği hiperaktivite bozukluğundan (DEHB) şizofreniye kadar birçok psikiyatrik bozukluğun yaygınlığı artmıştır. Bu yazıda antisosyal kişilik bozukluğu olan ilişkili çocukluk çağı bozukluklarından DEHB ve davranış bozukluğu ve de alkolizm ektanısı olan bir olguyu sunmak istedik. Tüm bu bozuklukların X'e bağlı kromozomla ilişkili olabileceğini belirttik.

**Anahtar kelimeler:** Antisosyal kişilik bozukluğu, Klinefelter sendromu

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## INTRODUCTION

Klinefelter's syndrome (KS, 47, XXY) is the most common chromosomal aberration among men and the incidence of the syndrome is 2.0 per 1,000 live-born males. The physical and cognitive phenotype associated with XXY is highly variable. Individuals with XXY may have hypogonadism, fertility problems, tall stature, gynecomastia, language based learning disabilities, and disorders of executive function (1).

Previous research has raised concerns that individuals with KS have an increased prevalence of psychiatric disturbances, ranging from attention deficit hyperactivity disorder (ADHD) in childhood to schizophrenia and severe affective disorders during adulthood (2). 47, XXY aneuploidy is found in about 0.8-1% of men hospitalized for schizophrenia, representing a four-fold to five-fold excess over the incidence at birth of KS (3). A survey of hospital admissions and discharge diagnoses among individuals with XXY in Denmark (n=832) and a randomly selected age-matched control group (n=4033) found that individuals with XXY had an increased relative risk of being hospitalized for psychiatric disorder, particularly for psychoses (4). A psychiatric screening of 31 adults with XXY showed an increased prevalence of psychosis (6.5%) and depression (19.4%) (5).

Bruining et al. (2) found that language disorder 65% as the most prevalent disorder followed by ADHD (63%) and autism spectrum disorder (27%) in this

syndrome. Behavioral impairment was most evident among cases classified as autism spectrum disorder and psychotic disorder 12%. Although there are much reports regarding to KS associated with psychiatric disorders in literature, there are limited reports regarding to personality traits (6). We want to present a case report of KS who presented with antisocial personality disorder.

## CASE

A 17 year-old boy was admitted to the Emergency Department because of aggressive behavior, substance use and self mutilation. Upon presentation to the emergency room, he described his mood as "out of temper" with sleep disturbances. He reported having difficulty in maintaining interpersonal relationships. He denied significantly depressed mood. He also stated that he got nervous from time to time but denied any symptoms of generalized anxiety disorder, post-traumatic stress disorder, obsessive-compulsive disorder or panic disorder. He reported that he felt paranoid while in a stressful situation but denied any auditory or visual disturbances or paranoia during that encounter.

He started drinking alcohol when he was 8 years old, and he started to use cannabis when he was 11 years old. He had impulsive behaviors since childhood. He reported that he had multiple self-inflicted injuries. He added that he had legal problems due to fights at school. His past psychiatric history was significant for a diagnosis of ADHD and conduct disorder. He was started on methylphenidate 20mg two times a day for ADHD when he was 8 years old but he could not tolerate and did not use it. His grandfather has schizophrenia. He had significant dysphoric mood in mental status examination. Physical examination indicated that he is a tall (200cm) and overweight (110kg) boy with long arms and legs. His blood pressure and his heart rate were normal. There were no withdrawal symptoms. We thought of Klinefelter's syndrome due to his physical appearance. Cytogenetic studies showed a 47, XXY karyotype. All other investigations, including blood count, renal function, electroencephalogram, cerebral MRI and serum testosterone levels, were within normal limits. His scores on Wechsler Scale were 77 on performance, 86 on verbal and 83 on full scale.

## DISCUSSION

In this case, we presented a KS who presented with antisocial personality disorder and related disorders which are ADHD, conduct disorder and alcoholism. The common feature of all these disorders is that they may be associated with X-linked chromosomes.

Many KS men are not significantly affected. On the other hand, for some the presence of an extra X chromosome is associated with cognitive, psychosocial, motor and language deficits. The presence of an extra X chromosome in KS is associated with the abnormal development of both grey and white matter in the frontal and temporal lobes. Because individuals with KS show cognitive deficits even before puberty, at a time when testosterone levels are near normal (7), it is unlikely that the profile results from androgenic insufficiency affecting neural development. However, there are anecdotal studies that testosterone supplementation leads to better grey matter preservation in the superior temporal gyrus (8).

KS is associated with multiple psychiatric comorbidities. McDanal et al. (9) had demonstrated that presentation of mixed emotional and behavioral clinical picture is similar to and difficult to distinguish from that of other child psychiatric disorders. Caroff (10) observed that individuals with KS often had a passive-aggressive constitution. He also reported that KS is also associated with schizophrenia, neurologic syndromes, mental retardation, personality disorders, paraphilia, criminality and alcoholism. Recent studies on the genetics of alcoholism have suggested an association between antisocial behaviors and the MAO-A gene. Saito et al. (11) found that the MAO-A promoter polymorphism was present in 3% of type 2 alcoholics. They suggest that MAO-A is X linked, the heterozygotes are probable cases of KS suggesting that X-chromosome aneuploidy may increase the risk for developing type 2 alcoholism (11). Previous studies consistently demonstrate that for men, and probably

for women, a history of conduct disorder in childhood and adulthood predisposes one to develop an alcohol use problem (12). Previous reports also have shown that alcoholism with antisocial personality disorder is characterized by an early onset of alcohol-related problems and increased severity of dependence (13). Brunner et al. (14) reported a Dutch family with a complete MAO-A deficiency due to a point mutation in exon 8 of the MAO-A gene, and the male family members displayed abnormal aggressive behavior and borderline mental retardation.

In conclusion, KS may be presented with a personality disorder as much as various other psychiatric disorders. Healthcare professionals can play an important role to prevent and manage worsening of the psychiatric symptoms.

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