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BRIEF COMMUNICATION

Homicide and Klinefelter syndrome: a complex interaction

Stéphane Richard-Devantoy,[1] Fabrice Jollant,[1] Anne-Isabelle Bouyer-Richard,[2] Jean-Paul Lhuillier,[3] Philip Gorwood[4]

[1]*McGill Group for Suicide Studies Montréal, Department of Psychiatry & Douglas Mental Health University Institute, McGill University, Québec, Canada.* [2]*Institut Philippe-Pinel de Montréal, Montréal, Québec, Canada.* [3]*Hospital of Psychiatry, Sainte-Gemmes-sur-Loire, France.* [4]*INSERM U894, Paris-Descartes University, CMME, Sainte-Anne Hospital, Paris, France.*

Introduction: Several studies have shown an association between homicide and sexual chromosomal abnormalities, but data are still lacking regarding Klinefelter syndrome.
Methods: We retrospectively reviewed two cases of homicide perpetrators who were both diagnosed with Klinefelter syndrome on the basis of a karyotype analysis. A neurocognitive assessment was also performed (MMSE, Frontal Assessment Battery, brain CT, and electroencephalogram).
Results: Numerous intermediate risk factors of homicide were shared by our two cases, including dispositional (male gender, young age, low socioeconomic status), historical (prior arrest record and past conviction for any offense), contextual (unemployment), and clinical (alcohol abuse).
Conclusion: It is important that clinicians go beyond obvious risk factors, such as chromosomal abnormalities, to pinpoint other meaningful risk factors and potentially facilitate preventive approaches.

Keywords: Homicide; Klinefelter syndrome; risk factors; chromosomal abnormalities

Introduction

Homicide is defined as fatal injuries inflicted by another person with intent to injure or kill.[1] Its prevalence is high, with an overall rate ranging from 1.0 per 100,000 population in established market economies to 44.8 per 100,000 in sub-Saharan Africa.[2] Epidemiological studies[3,4] have shown a strong association between homicide, major mental disorders,[5] and chromosomal abnormalities.[6] However, the role of Klinefelter syndrome in homicide is inconclusive.

Klinefelter syndrome is caused by one or more supernumerary X chromosomes in men, and represents the most frequent chromosomal aneuploidy, with a prevalence of 1 in 700 men.[7,8] Klinefelter syndrome also represents the leading cause of male hypogonadism and is characterized by cognitive and psychosexual dysfunctions and abnormalities in physical maturation.[7] Many cases of Klinefelter syndrome remain undiagnosed because of substantial variation in clinical presentation. Only 10% of subjects with Klinefelter syndrome are diagnosed prenatally, and another 25% diagnosed during childhood or adulthood, leaving 65% undiagnosed. Therefore, this syndrome may be relatively common yet often undiagnosed among psychiatric patients.[7-9]

Correspondence: Stéphane Richard-Devantoy, McGill University, Department of Psychiatry & Douglas Mental Health University Institute, McGill Group for Suicide Studies Montréal (Québec), Canada, Address: Douglas Institute, FBC building, 3rd floor, 6875 boulevard Lasalle, Montréal, Québec, H3W 2N1, Canada.
E-mail: richarddevantoy@orange.fr
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This syndrome has also been associated with violent behavior,[10-12] but the involved risk factors for homicide in these patients remain unknown. In the present study, we report two cases to shed light on the relationship between Klinefelter syndrome and violence. We reviewed two cases of homicide perpetrators (2 out of 276 perpetrators retrospectively examined), who had been imprisoned for homicide and were subsequently diagnosed with Klinefelter syndrome. Although this syndrome seems to be associated with an increased risk of aggressive behavior, we hypothesize that a large set of vulnerability risk factors found in individuals without Klinefelter syndrome are also common in men with Klinefelter syndrome and might have a significant impact on the risk of homicide.

Case report

Two men, aged 25 and 26 years respectively, were imprisoned for homicide in Western France (Table 1). They were examined by a forensic psychiatrist as part of a comprehensive psychiatric evaluation, which includes standardized psychological tests and additional physiological tests (such as blood tests, electroencephalography, and, in some cases, a computed tomography [CT] scan of the brain). Psychiatric reports were requested from the trial courts. In both cases, the victim was an adult stranger to the perpetrator. In the first case, the victim was killed with a firearm, whereas in the second case, the victim was killed with a blunt object. In both cases, the homicide occurred at night, in a private residence, and during a physical argument. In addition, both perpetrators had consumed alcohol, but no other

Table 1 Clinical and criminological characteristics of two homicide perpetrators with Klinefelter syndrome

	Perpetrator 1	Perpetrator 2
Clinical data		
Gender	Male	Male
Age (years)	27	26
Family situation	Unmarried, childless	Unmarried, childless
Professional situation	Unemployed	Unemployed
Previous convictions for any offense	Theft	Theft
Previous convictions for any violence	No	No
History of psychiatric conditions	Alcohol abuse, conduct disorder, suicide attempts	Alcohol abuse, conduct disorder
MMSE (max. score = 30)	28	29
FAB (max. score = 18)	12	13
Intelligence quotient (WAIS)	85	93
Circumstances of the homicide		
Place	Outdoors	Outdoors
Time	Evening	Evening
Type of weapon	Firearm	Blunt object
Mental state at the time of the crime	No delusions, hallucinations, or depression	No delusions, hallucinations, or depression
Alcohol intake	Yes	Yes
Relationship with victim	Stranger	Stranger

FAB = Frontal Assessment Battery; MMSE = Mini Mental State Examination; WAIS = Wechsler Adult Intelligence Scale.

substance, at the time of the offense. After the homicide, the perpetrators either called for assistance or ran away. In regards to sociodemographic characteristics, both perpetrators were married, childless, and unemployed. Additionally, they both had psychiatric antecedents, such as a history of conduct disorder in childhood, suicide attempts (first perpetrator), and a history of alcohol abuse (second perpetrator). They both had at least one conviction for a past non-violent crime. Physical examination of both individuals revealed hypogonadism. The Mini Mental State Examination (MMSE) scores were 28 of 30 and 29 of 30 respectively, and the Frontal Assessment Battery (FAB) scores, 12 of 18 and 13 of 18, respectively (normal score = 18). The neurological examination was normal. The intelligence quotient (IQ) was assessed with the Wechsler scale and scored at 85 and 93 (normal ranges score between 70 and 130), respectively. Brain CT showed no ischemic or hemorrhagic lesions or any signs of dementia. The electroencephalogram was normal in both cases. The karyotype revealed a 47,XXY genotype in both cases; neither perpetrator had been previously diagnosed with Klinefelter syndrome.

Discussion

There has been much debate as to whether men with the 47,XXY genotype are more likely to engage in criminal and violent behavior than their 46,XY counterparts.[13-15] We would like to emphasize the complex interaction of homicide factors in men diagnosed with Klinefelter syndrome, and the importance of the early diagnosis of this syndrome.

The two individuals with undiagnosed Klinefelter syndrome described above had committed homicide. However, they also showed several other risk factors for this violent act, including dispositional (male gender, young age, low socioeconomic status), historical (prior arrest record and past conviction for any offense), contextual (unemployment), and clinical (alcohol abuse) factors.

The factors that lead to homicide in Klinefelter syndrome are complex, heterogeneous, and interactive. Klinefelter syndrome has been widely associated with mild cognitive impairment and language impairments. The frequency of psychiatric disorders in patients with Klinefelter syndrome is also suggested to be higher than in the general population.[16,17] Personality features reported in men with the XXY karyotype include passivity, poor concentration, emotional immaturity, shyness, and hypersensitivity.[7] The association between Klinefelter syndrome and violence could be linked to a lower IQ,[18] with selective reduction in verbal IQ scores.[19] In addition, low IQ and psychiatric symptoms and disorders are more clearly associated with homicide.[5] Regarding the normal but below-average IQ of the two patients reported herein, it is interesting to note that a deficit in inhibitory executive functions has been detected in XXY males as compared with XY male and female (XX) controls,[20] potentially reflecting an impairment in the prefrontal cortex.[21]

A recent study suggests that the overall risk of homicide is moderately increased in men with Klinefelter syndrome; however, it was similar to that of controls after adjusting for socioeconomic parameters.[22] Therefore, the discovery of rare chromosomal abnormalities in cases of homicide should not be construed as an exclusive explanatory or predictive factor for the criminal act. Homicide has also been related to several non-psychiatric factors, including the personal history of the perpetrator (i.e., history of violence, juvenile detention, physical abuse, and parental arrest record),[5,23] dispositional (i.e., male gender, younger age, and low socioeconomic status)[5,23] and contextual factors (i.e., recent divorce, unemployment, and victimization).[5,23] It is important that forensic psychiatrists examining XXY patients be aware of these issues, as the association between XXY genotype and homicide may be raised in court.

Nevertheless, future research should take into consideration the prevalence of Klinefelter syndrome among homicide perpetrators and the need to study the risk of homicide in this population vs. the general psychiatric population. Finally, prospective studies should be conducted to ascertain whether Klinefelter syndrome constitutes an independent risk factor for homicide.

In summary, to explain the criminal act, we propose the hypothesis that numerous comorbid risk factors might be much more clearly involved in homicide than Klinefelter syndrome per se, even though a multiplicative effect is possible, i.e., many of these risk factors may be maximized when a person has a 47,XXY karyotype. As "one cannot see the forest for the trees," it is also important that clinicians diagnose Klinefelter syndrome earlier in its course.

Disclosure

The authors report no conflicts of interest.

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