Chronic Koro-like Syndrome (KLS) in recurrent depressive disorder as a variant of Cotard’s delusion in an Italian male patient. A case report and historical review

Un caso di Koro-Like Syndrome (KLS) cronica, come variante di un delirio di Cotard, in un paziente italiano affetto da depressione monopolare cronica. Presentazione del caso clinico con revisione storica della letteratura

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SUMMARY. Cotard’s syndrome is a delusional syndrome, first described in the 1880ies by Cotard, characterized by a nihilistic delusion about the self and/or the world. In same other cases there is an intense nihilistic belief that the patient’s entire body or parts of it are disintegrated or dead. The syndrome is often associated with severe depression, but are also described neurological cases. Koro was described a little later from Asia and consisted in the belief that one’s own genitalia are shrinking or disappearing and death will ensue thereafter, but there are many cultural variants and the syndrome may present in an incomplete form. We report on a KLS sharing more features with annihilation delusions, such as Cotard’s syndrome. In KLS, the délire de négation may be limited to localized systems or organs. We believe that some complete and incomplete forms of Koro, when embedded in a depressive core, may represent a variant of Cotard’s delusion. In fact, our patient did not reach a complete denial of his entire body, but rather focused on sexual identity. We analysed the psychosexual issues of our case according to Kretschmer’s 1918 view of a “bipolar setting” between sthenic and asthenic characters of a patient suffering from sensitive delusions of (self-) reference. This view may allow us to relate the personological character to the genetic comprehensibility of the delusion.

KEY WORDS: Koro-Like syndromes, Cotard’s delusion, recurrent depressive disorder, Kretschmer.

RIASSUNTO. La sindrome di Cotard consiste in un disturbo delirante descritto per la prima volta nel 1880 da Jules Cotard ed è caratterizzata da un delirio di negazione riguardante sia sé che il mondo circostante. Successivamente cominciarono a essere descritti altri casi in cui pazienti presentavano il convincimento delirante che l’intero corpo, o parti di esso, fossero morte o assenti. Detta sindrome è spesso associata a casi gravi di depressione maggiore, anche se sono stati descritti casi dovuti a malattie neurologiche. Il Koro, descritto in alcuni Paesi asiatici, consiste nel convincimento delirante che i propri genitali si retraggano nell’addome o scompaiano, con un inevitabile morte che sopraggiglierà di lì a poco. Vengono però descritte molte varianti nelle diverse culture asiatiche rispetto a questo quadro principale, e la sindrome può anche presentarsi in forma incompleta. In questo lavoro presentiamo un insolito caso di KLS in un paziente italiano, e che presenta molte caratteristiche comuni al delirio nihilistico di Cotard. Riteniamo che sia le forme complete sia quelle incomplete di Koro possano rappresentare una variante del delirio di Cotard. Nel caso da noi descritto, il paziente non presentava una negazione globale del proprio corpo, ma una sensazione focalizzata sul tema della perdita dell’identità sessuale. In questo lavoro abbiamo inoltre analizzato l’aspetto psicosessuale nel quadro di riferimento del delirio sensitivo di rapporto proposto da Kretschmer nel 1918, e relativo alla costituzione “bipolare” tra caratteristiche personologiche sottostanti “steniche” e “asteniche” nella genesi di un delirio di negazione riferito a sé. Questa cornice teorica ci ha permesso di elaborare ipotesi sulle caratteristiche di personalità di base sottostanti a questo delirio.

PAROLE CHIAVE: Koro-Like Syndromes, delirio di Cotard, disturbo depressivo ricorrente, Kretschmer.

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INTRODUCTION

Cotard’s syndrome is a delusional disorder introduced in the early 1880ies by the French neuro-psychiatrist Jules Cotard (Issoudun, Berry, 1st June 1840-Paris, 19th August 1889), who had worked with Charcot at the Salpêtrière in Paris. Cotard first presented a case of a “lypemanic” (with delusional depression) 43-year-old suicidal woman (with delusional inside organs) did not exist and who stated she needed no food, as she was immortal, but prompted others to burn her alive, as she believed fire was the only way she could give an end to her life (1). He coined the term “délire de négations” two years after (2), mistranslated into “nihilistic delusions” (3). In fact, the translation does not render the fact that Cotard’s term was encompassing also other, non-delusional aspects of the syndrome, which comprised also anxiety and depression (4). The attribution of the syndrome to the man who had first described it systematically was made by Emmanuel Jean-Baptiste Joseph Régis (1855-1918) four years after Cotard’s premature death (5).

Koro is a psychiatric illness characterized by acute anxiety accompanied by the belief that one’s own penis (or vulva or nipples and breasts, in women) is shrinking and will dissolve in one’s own abdomen, and by the fear that upon completion of this process, death will result (6). This syndrome has been variously viewed as obsessive-compulsive illness, anxiety neurosis, psychotic depersonalization and hysteria. It is a culture-bound syndrome most frequently occurring in South China, Malaysia and Indonesia, especially among people of Chinese origin. However, there are some rare case reports from the Western hemisphere.

In the DSM-IV-TR (7), koro is mentioned only in Appendix I (letter, not Latin number), in the context of culture-bound syndromes, as «a term, probably of Malaysian origin, that refers to an episode of sudden and intense anxiety that the penis (or, in females, the vulva and nipples) will recede into the body and possibly cause death. The syndrome is reported in south and east Asia, where it is known by a variety of local terms, such as shuk yang, shook yong, and suo yang (Chinese); jinjinia bemar (Assam); or rok-joo (Thailand). It is occasionally found in the West. Koro at times occurs in localized epidemic form in East Asian areas. This diagnosis is included in the Chinese Classification of Mental Disorders, Second Edition (CCMD-2)».

The complete Third Edition of the Chinese Classification of Mental Disorders (CCMD-3) (8) became available three years after the DSM-IV-TR, in both Chinese and English; koro is defined as «excessive fear in men of the penis shrinking or drawing back in the body. This has been associated with cultures placing a heavy emphasis on balance, or on fertility and reproduction». It may immediately be noted that the disorder is strictly confined to men, and it is broadened to encompass the concepts of ancient Chinese philosophy.

The ICD-10 (9) in its Mental and Behavioural Diseases, Diagnostic Criteria for Research manual, defines koro as «Acute panic or anxiety reaction involving fear of genital retraction. In severe cases, men become convinced that the penis will suddenly withdraw into the abdomen; women sense that their breast, labia, or vulva will retract. Victims expect the consequences to be fatal. Studies cite factors such as illness, exposure to cold, or excess coitus as precursors, but interpersonal conflict and sociocultural demands reportedly exert greater influence on the condition. Onset is rapid, intense, and unexpected. Responses vary, but include grasping of the genitals by the victim or a family member, application of splints or devices to prevent retraction, herbal remedies, massage, or fellatio».

It also suggests encoding it as “F48.8 Other specified neurotic disorders” or “F45.34 Somatoform autonomic dysfunction of the genitourinary system (may be used if autonomic anxiety symptoms are present)” and differentiating it from Indian “dhat” and “prameha” (10,11) or Taiwanese “shen-k’uei” (10) (acute anxiety and somatic complaints such as fatigue and muscle pain, related to a whitish discharge with urine, interpreted as loss of semen-like material in both men and women and attributed to excess coitus, urinary dysfunction, imbalance among body humours or diet) and Egyptian or other Arab “rabt (al azhár)” (impotence or other sexual dysfunction due to sorcery or spell) (12).

The first medical reports on koro, a reportedly Malaysian word having to do with the tortoise’s head, that may be retracted in its shell, were published in the mid-1890ies, while the term “lasa Koro”, referring to dangerous penile shrinkage in Indonesian natives, had already been introduced in Western literature by Benjamin Frederik Matthes in his 1874 Buginese-Dutch dictionary (quoted in Edwards) (14). However, the same syndrome was long known in China as suo yang (Mandarin), suk yong in Yue (Cantonese) or shook yiang in Wu (Shangainsese) and was found in Emperor’s Huang Di Classic of Internal Medicine (15). Upton (16) supports that the text was presumably not written by the Emperor himself, although he is celebrated as the father of Chinese medicine, because during these times (the Yellow Emperor is believed to have existed between 2697 and 2597 BC.) it was cus-

* A language spoken by more than three million people, mainly in the southern part of Sulawesi, an Indonesian island.
In the Western World, the first account of penile retraction was on the Lancet (21); the author described the case of a man who saw a couple of years before, and was prompted to report the case because a similar case, though more severe, had been published in a Russian journal in the same year by Ivanov. In these two cases, shrinkage had actually occurred and was intensely anxiogenic. The first report of the culture-bound syndrome was in 1895 by Blonk (22), a military surgeon in Indonesia. The paper was in Dutch, the language that dominated the early literature on koro. The condition was viewed as an anxiety neurosis with hysterical features (23), or psychodynamically framed as castration fear (24). Although the element of delusional belief, often shared, that the penis will somehow disappear may lack from Western World cases, it is difficult to differentiate qualitatively the culture-bound syndrome from isolated Western cases.

Several authors attempted telling complete from incomplete forms of koro (25). In the incomplete forms, the belief that the penis will disappear into the abdomen or cause death is not present; however, the male patient is persuaded that he is impotent and that he lost his virility. Koro-like Syndromes (KLS) in Western countries are not distinct clinical entities, but represent a concomitant syndrome that requires treatment of the underlying illness.

We present a case of atypical KLS in which the patient presented with the delusion that his body would switch from male to female.

**CASE HISTORY**

C.M., a 58 year-old single male was admitted to our Psychiatric Hospital due to worsening of pre-existing symptoms of depression. On admission he was feeling de-
pressed with loss of interest and drive, low self-esteem and
guilt, feelings of inadequacy and of existential failure. The
patient also complained of symptoms in which “every-
thing” was over, and any solution would have been impos-
sible to resolve his case, because he had lost not just pos-
sessions, but also the social status and power, as well as any
ability to “be” in the world.

During a thorough psychiatric interview, he gave infor-
mation about his illness and showed marked anxiety and
fear about his penis, which he thought was gradually
shrinking.

A few weeks prior to his admission, the patient felt that
his penis was retracting gradually. He felt that as a result of
this, he would become impotent, with no sperm, and that
he would lose his gender. His belief that his body would
change from male to female was supported with delusion-
al features.

The patient was 18 when he first manifested concerns
over his penis, which he believed will disappear, and that
this would be followed by loss of his virility. He described
himself as being much skinny at that time, with a poorly
developed beard and no muscles. He had very low mascul-
ine self-esteem at that time. The symptoms waxed and
waned for about 30 years. He lately started to grow a
beard just to obtain a masculine self-image. During admis-
sion he believed that doctors would cut-off his penis to
help him change his body from male to female. He eager-
lly sought assurance about his virility and often touched
his abdomen, as he believed that some parts of the ab-
domen had lost function and also that he had serious
physical illness.

He was the third of four children. He lived with his sis-
ter, who had been hospitalized for schizophrenia for few
months. There were no other cases of psychiatric illness in
his family.

His childhood was normal. He has always been shy, sen-
sitive, polite, and had some difficulty making friends. As a
child, he felt somewhat inadequate. At age 23 he aban-
donned his studies of architecture, after an academic failure.
For the past 15 years, he had worked as a designer. Since
the age of 25 he had been keeping an assiduous correspon-
dence with many women from all around Europe and occa-
sionally met them, but had no romantic relationships un-
til the age of 30. Although he had some romantic involve-
ment with women with whom he kept correspondence, at
age 35 he stopped such occasional encounters because he
believed that these women could perceive his loss of viril-
ity and masculinity.

The patient used minimal quantities of alcohol and did
ever abuse any substance. When he was 35 went to work
in Libya, where he stayed for about 13 years. He was not
specifically acquainted with the cultural beliefs of the Far
East. He was first diagnosed as affected by major depres-
sive disorder, recurrent type, at age 54. Since then, he was
admitted to our hospital five times, about once a year. Dur-
ing these admissions, he had the terrifying perception that
his penis was shrinking. This perception tended to subside
during the symptom-free interval of his recurrent depres-
sive disorder. In many of these admissions the patient felt
extremely inadequate and unable for routine activity, as he
felt overwhelmed by even the easiest tasks. During his last
admission, the delusion regarding his male-to-female body
transformation presented for the first time. Tested on the
Dissociative Experience Scale (DES) (26), he was found to
be free from depersonalization and dissociation. On the
short-form of the MMPI-2 he scored 94 on the D (Depres-
sion) scale (T-score) (normal T scores are considered those
below 60). He also scored higher than normal on the Sc
(Schizophrenia) (T-score=67) and Si (Social Introversion)
(T-score=64) scales. These results, combined with those of
the Structured Interview for the DSM axis II for personal-
ity disorders (SCID-II), which had been completed four
year ago, confirmed the patient’s premorbid personality,
characterized by shyness, unassertiveness and some fea-
tures of paranoia. Routine blood and urologic investiga-
tions were normal.

During his last admission, he was prescribed a drug
combination consisting of an antidepressant (venlafaxine,
150 mg/day), an antipsychotic (olanzapine, 20 mg/day) and
an anxiolytic (lorazepam, 7.5 mg/day). The patient was
treated with this combination throughout his 30-day hospi-
tal stay. He was discharged with this treatment combina-
tion. At the one-month follow-up he was free from delu-
sional ideation and depression. His Brief Psychiatric Rat-
ing Scale (BPRS) scores had dropped from 73 to 39. He
provided written, informed consent for the publication of
his case.

DISCUSSION

In our case, severe depressive illness led to disrup-
tion of perceptual continuity, determining a distortion
in body image perception. Dissociative mechanisms,
such as those leading to depersonalization, are com-
patible with distorted body image perception, that may
lead to the perception of parts of the body as dysmor-
phic and dysfunctional. Depersonalization has been
advocated to explain the pathogenesis of Cotard’s syn-
drome (27), a mechanism similar to the one claimed to
account for koro (28), however, we found no deperson-
lization in our case. If depersonalization is involved in
this case, it should be partial. According to Berrios et
al. (29), at least some delusions might be based on de-
personalization experiences. Thus depersonalization
might serve as a general “experiential substratum”
which (modulated by different cognitive frames) will
crystallize out into different delusional phenomena. In
particular, it has been suggested that some forms of ni-
hilistic or hypochondriacal delusions could have their
origin in somatic depersonalization experiences, differ-
ently conceptualized by the psychotic patient.

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While in KLS body-image disturbances may be limited to localized systems or organs, in Cotard’s syndrome the disturbance usually extends to the whole body. In our case, fears over male-to-female body transformation does not mean discontinuity of one’s own identity. Abnormality of body image may be the result of abnormal sensations (30). Our patient felt that he was impotent, with no sperm, and that if his penis retracted completely, he would change his gender and he would be “nothing”. In fact, our case did not reach a complete denial of one’s own body, but rather a selective change related to sexual identity; in this patient, the fear to be transformed into a woman could represent not an absolute discontinuity with respect to self-identity, but an inevitable corollary of the fact that on the face of a reduced perception of one’s male identity there is an unavoidable transformation into the opposite sex. The delusional thinking linked to such psychopathological condition is in fact holothymically comprehensible, as described by Kretschmer (31) in 1918.

According to Kretschmer (31), the presence of an underlying mood disorder provides an acceptable explanation behind the body image disturbance. He argued that psychosexual conflicts led to sensitive delusions of (self-)reference. In his clinical cases of sensitive delusion of (self-)reference, the accent is on critical episodes caused by ethical-sexual conflicts. Human experience developing in the space between the self and the external world, according to this author, tends towards a “bipolar” pattern, swinging between two poles, represented by superiority feelings and high self-ideal (sthenic polarity) and those characterized by inferiority, discouragement and shame (asthenic polarity). In particular, Kretschmer’s interest focuses on the psychological development of character, in which a sthenic disposition is subjected to the stimulant influence of a strong asthenic disposition that forms the opposite pole, or vice versa. In the first case we obtain expansive natures, whose most typical psychological manifestation is paranoia, in the second we obtain sensitive natures. Most often, perceived inefficacy and vulnerability are encountered at the background of people with strongly sthenic sensitivity. The same way paranoid people display a wide range of sthenic qualities, albeit with a hidden core of vulnerability, the sensitive shows a mainly asthenic constitution, which is however subject to a sthenic counter-reaction (31).

The internal experience of these patients is isolated; they jealously abstain from exteriorizing their feelings, which remain in a permanent tension state. They demonstrate much introspective power and insight, scrupulous morality and true altruism. We may hypothesize that the sensitive character, who usually lives in a conscious way the conflict between grandiosity and shame, due to particularly traumatic experiences which threaten self-esteem, is forced to externalize this conflict between self-ideal and shameful inadequacy, thereby identifying his internal persecutory instances in another, external person. The feeling of self-cohesion is preserved by the angry-sthenic turnover of the breakdown caused by shame in a persecutory feeling. These subjects have an extreme sensibility, their spiritual life is isolated, their feelings remain tense, and they have a scrupulous attention to other people. In our case, these personal characteristics are well represented, mainly in their asthenic expression (feelings of inadequacy, shame) only partially counterbalanced by sthenic drives, as shown by a higher than normal score on the Pa (Paranoia) scale of the MMPI, by excessive scrupulousness, and by inadequate feelings when facing everyday tasks.

Hence, our case bears some similarities with one case described in the mid-nineties by Wolff and McKenzie (32). These authors observed misidentification syndromes of both the Fregoli and the Capgras subtypes in a patient with shared psychotic disorder and koro during periods of depression. It could be possible to trace an analogy between our patient’s “misidentification” of his own body and the misidentification of other people by the patient described by Wolff and McKenzie (32). Their patient could be viewed also according to Kretschmer’s conceptualization, with sthenic instances promoting the paranoid, outward-directed misidentification, and asthenic ones being related with inward-directed misidentification, like koro-like syndrome, which should be viewed like a particular form of Cotard’s delusion. In this sense we can find resemblances between the thought of Kretschmer and the modern theories of attributional style and its relationship with delusion content.

Attributional biases are cognitive biases which affect attribution, the way we determine who or what was responsible for an event or action. Such biases typically rely on actor/observer differences, and attribution theory provides a framework for understanding the causal explanations that individuals give for their own behaviour and the behaviour of others. Normal subjects consistently demonstrate a self-serving attributional bias in explaining the causes of events; that is, they tend to take credit for success (internal attribution of positive events: the “self-enhancing” bias) and to deny responsibility for failure (external attribution of negative events: the “self-protective” bias). Such biases may serve to enhance self-esteem. Patients with persecutory delusions (with diagnoses of paranoid schizophrenia or delusional disorder), show an exaggeration of this self-serving attributional bias (33).
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In Cotard patients are thought to have a damage to neural pathways underpinning the emotional component of face recognition (34) and (35). The ensuing discordance between experiences of the way someone “looks” and the way they “feel” is thought to underpin the impostor delusion of these patients. Young et al. (36), have suggested that disruptions to the affective component of visual recognition may occur in Cotard cases as well as in Capgras cases. However, whereas Capgras patients interpret the resultant experiences in accordance with a paranoid, projective attributional style, Cotard patients interpret them in accordance with a depressive, introjective attributional style (37), like Kretschmer’s theoretical framework.

Our patient had a chronic course, supporting that sporadic KLS could be not a self-limited condition; our case is similar at this respect with the two cases from the East Indies, who were recently described by Kar (38).

It should be stated that our case does not bear all these features that render koro a culture-bound syndrome. He was poorly acquainted with cultural backgrounds of the Far East and had only a multi-year experience with Arabic culture. Although to date no koro case has been reported from Libya, some cases were described in Asian Arab patients (39,40) and patients from seven West African countries (41). However, it is unlikely that our patient could have been influenced by culture, since he had already developed his fear of penis retraction by the age of 18, long time before he visited a foreign country. Furthermore, our patient extended his uncertainties about his masculinity also to his beard, and this places him nearer to Cotard’s delusion rather than to classical, culture-bound koro. Psychiatrists should be more sensitive to their patients’ significance of their symptoms and to other coexisting symptoms to distinguish classical koro (in which, for instance, the patients may use physical maneuvers to prevent penile retraction, and many times believe they will die as consequence of Koro), from possible cases of KLS with higher affinity for Cotard’s delusion and depression, and treat them accordingly. In this regard it is important to point out the differential diagnosis of non-Asian Koro like cases with major depression (42), schizophrenia (43), bipolar disorder, as well as neurological disorders with psychiatric comorbidity, as epilepsy, brain tumors and Parkinson disease (4). It is interesting at this respect, that whereas koro is usually treated with antipsychotics, two cases of formae frustae of koro, one in a Caucasian (44) and one in a Japanese patient (45) were successfully treated with selective serotonin re-uptake inhibitors.

CONCLUSIONS

Our case is a KLS sharing more features with annihilation delusions, such as Cotard’s syndrome, rather than the culture-bound syndrome which we intend as koro. This is an only partial annihilation delusion, which shows polarity, as shown by the fear of transformation into the opposite gender. Such polarity recalls Kretschmer’s framing of sthenic and asthenic characters, which best suit our case. The intense emotional turmoil could affect consciousness by producing altered perceptual constancy, which in turn would result in delusional perception of body or one part of it. In our case, perceived sexual inadequacy characterizes the entire existence of the patient and becomes delusional upon worsening of depressive symptoms.

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