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Scientific letter

Koro-like syndrome in Huntington's disease

Koro syndrome, also known as "genital retraction syndrome", is a rare an often-severe psychiatric disorder characterized by the development of acute anxiety and a fear of retraction of the penis, followed by an intense fear that the penis will recede into the body resulting in death.[1] This delusional disorder is considered to be culture-bounded, occurring in epidemic and sporadic forms mostly in South-East Asia, Japan and China,[2] but sporadic cases have also been reported in western regions in association with disorders usually involving the central nervous system. Thus, cases of Koro have been reported following stroke, in alcoholic hepatitis, following abrupt withdrawal of olanzapine, in a tumor of the corpus callosum, in bronchial carcinoma with metastatic spread to the brain, in epilepsy, and in mild cognitive impairment due to Alzheimer's disease.[3]

Huntington's disease (HD) is a monogenetic, autosomal-dominant, neurodegenerative disease caused by an abnormal CAG repeat expansion in the HTT gene.[4] Clinically, HD is characterized by the progressive development of motor, cognitive and neuropsychiatric manifestations. Subtle cognitive and behavioral changes usually precede the onset of motor symptoms[5] in concordance with the set of brain changes that involve extensive posterior-cortical and white matter territories and specially, the basal ganglia, years before motor-based diagnosis.[6]

Depressive mood, anxiety, irritability, suicidality, perseverative behavior and especially apathy, define the typical psychopathological profile of the HD.[7] However, HD may associate psychotic symptoms in up to 10% of cases, and up to 3% of people with HD exhibit a schizophrenia-like form of the disease.[8]

In this letter, we report a Koro-like syndrome acutely developed in a 54-year-old male affected by HD (CAG = 42). The patient presented a predominantly psychiatric and cognitive form of the disease associating loss of motivation, executive dysfunction, perseverative behavior and rumination, irritability and of somatic complaints, but there was no history of psychotic symptoms. Regarding motor symptoms, he presented oculomotor impersistence, decomposition of eye tracking and delayed latency. He also had some suppressible cephalic movements, mild generalized chorea predominantly in the trunk and mild dystonic posture of the right upper limb (UHDRS-TMS = 36). Pharmacological treatment consisted of venlafaxine 375 mg/day and olanzapine 5 mg twice/day.

During a follow-up visit the patient referred to be extremely concerned because of having the impression that his penis was getting smaller (see video in supplementary data). This concern forced him to repeatedly go to the toilet to check his penis. The patient explained that he has never seen his penis getting smaller, but that was unquestionably getting smaller. His companion, explained that the patient has the need to pull down his pants and constantly show them that his penis is getting smaller. During the visit the patient verbalized an immense anxiety and how worried he was about the possibility of dying due to this penis retraction (Fig. 1; supplementary data).

Although somatic complaints and some symptoms characterizing somatoform disorders are not uncommon in HD, Koro has never been reported in the context of this disease. The specific origin of

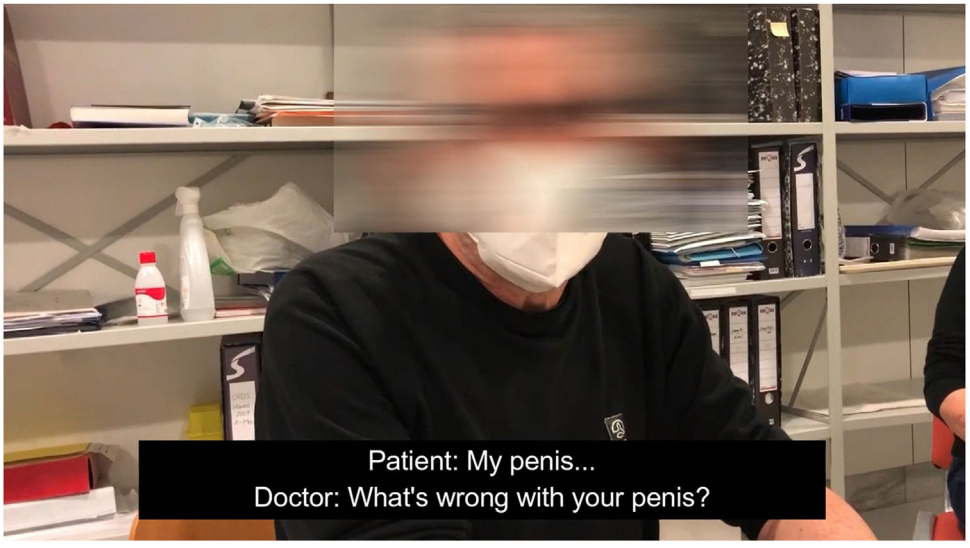

Fig. 1. Screenshot of the video included in supplementary data where part of the interview with the patient can be seen.

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these symptoms in HD is unknown, but it is reasonable to consider that progressive brain damage, cognitive impairment and the co-occurrence of other neuropsychiatric manifestations are central precipitating factors.

While the cases of Koro described in Asians have been consistently associated with anxiety and depression, the vast majority of Koro-like cases in non-Asian patients have been associated with primary and secondary psychotic disorders.[9] It has been proposed that Koro-like syndrome could share similar mechanisms than those precipitating some delusional misidentification syndromes.[9,10] Thus, the role of abnormalities in right fronto-temporo-parietal and limbic regions, especially those related to visual and spatial processing, to integration of external and internal signals, and to self-monitoring, are of particular relevance. Abnormalities in these processes can result in misidentification syndromes and in abnormal perceptions of the body, and possibly play a central role in Koro-like syndromes.[3]

Beyond the basal ganglia, HD associate changes in extensive temporo-parietal territories6 that contribute to sensorimotor integration disturbances. We consider that the confluence of aberrant proprioception of one's own body due to sensorimotor and proprioceptive integration deficits, with frontal-related cognitive dysfunction, perseveration and anosognosia, facilitated the construction of the delusion of penis retraction in our case.

To sum up, this is the first Koro-like syndrome ever reported in the context of HD, and it is a case that reminds us of the need to explore symptoms that go beyond the obvious or expected ones in the context of the general descriptions of the diseases we work with.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at https://doi.org/10.1016/j.rpsm.2023.01.006.

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