Pathological gambling as unusual manifestation of CADASIL: a case report.

M. Plastino, D. Messina, D. Cristiano, G. Lombardo, D. Bosco

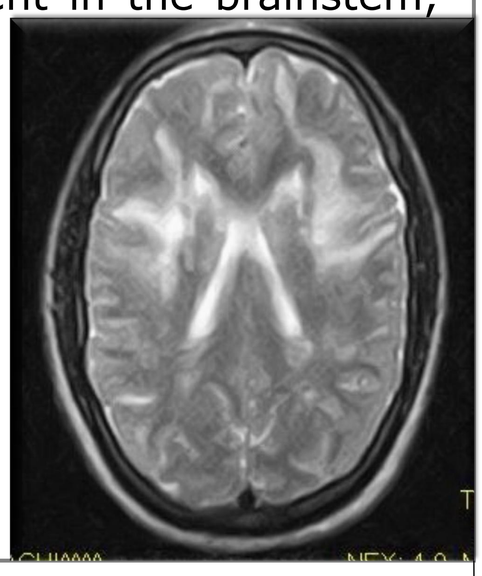
Department of Neuroscience, S. Giovanni di Dio Hospital, Crotone, Italy

Introduction

Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL) is an inherited small artery disease caused by mutation of the NOTCH-3 gene on chromosome 19. The disease is clinically characterized by a variable combination of migraine, recurrent transient ischemic attacks or lacunar strokes, psychiatric disturbances and cognitive impairmement. Psychiatric manifestations, mainly episodes of mood disturbances, are reported in 20-40% of patients [1]. Pathological gambling (PG) is a maladaptive and recurrent pattern of gambling behaviours that persists despite substantial negative consequences for the individual, his/her family and his/her work. In the fifth DSM edition (DSM-5) gambling disorder is classified under the category of "Addictions and Related Disorders" because of its similarities to substance use disorder (SUD) [2]. Here we report a case of PG in a patient with CADASIL.

Case presentation

A 39-year old married male, unemployed and genetically diagnosed with CADASIL, carrying a heterogeneous mutation (R90C in the exon 3) of the NOTCH-3 on chromosome 19p13.2-13.1, was hospitalized to our neurological outpatient clinic for an episode of transient confusion. He suffered from migraine attacks since the age of 20. He and his relatives denied depressive episodes or previous diagnosis of mood disorder. About six months before admission, he started to gamble every day, mainly football betting, slot machines and national lottery, cumulating about 40.000 euros in debts due to gambling. Magnetic Resonance Imaging (MRI) of the brain revealed a severe leukoencephalopathy with confluent and fairly symmetric T2 hyperintense foci in the deep and subcortical white matter of the cerebral hemispheres. Similar foci were present in the brainstem, thalami, basal ganglia and the subcortical white matter of temporal lobes (see Figure). To evaluate his comprehensive cognitive status, we performed the Mini Mental State Examination (MMSE), Alzheimer Disease Assessment Scale cognitive subscale (ADAScog) and the MMPI-II. In addition, the South Oaks Gambling Screen (SOGS) was administered to assess gambling severity. Depression was assessed by the Beck Depression Inventory (BDI). MMSE and ADAS-cog total scores were within normal range. The assignment of a high score to the SOGS scale (=10) revealed the presence of a serious problem of gambling disorder. Finally, BDI scores (=14) showed no clinical relevance of depression. No abnormalities were found in routine laboratiry tests, including VDRL, HIV antibodies.



Discussion

To our knowledge this is the first case of pathological gambling described in a patient with CADASIL. Psychiatric disorders in CADASIL are frequently reported to occur in the course of the disease. A recent review article has highlighted that personality and behavioural disorders were reported in seven (2%) cases of a previous 454-patient cohort, while mood disturbances were documented in 106 (24%) cases of a 451-patient series [1]. However, the majority of these studies did not use any defined criteria to assess psychiatric disorders and diagnoses were mainly based on history or review of clinical data. The location of ischemic lesions in the basal ganglia may play a key role in the occurrence of behavioural disturbances such as apathy or disinhibition [3]. In a previous study, Cognat et al. reported the case of a patient who showed sudden apathy and PG-like behaviour after bilateral ischemic lesion involving the dorsal portion of the caudate nucleus [4]. While the vast majority of pathological gamblers do not have neurological comorbidities, decision making impairment on "gambling tasks" has been reported in patients with prefrontal cortex lesions [5]. The exact mechanism underlying PG in our patient is unknown; we hypothesize that the accumulation of vascular sub-cortical lesions may have damaged in particular the striato-cortical circuits involved in reward mechanisms. We suggest to consider CADASIL hypothesis in PG patients with a family history of recurrent strokes and/or psychiatric disorders.

References

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