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Wernicke-Korsakoff syndrome in a young adult

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Abstract

Introduction: Wernicke-Korsakoff Syndrome (WKS) presents a high underdiagnosis, and great clinical variability, mainly in brief alcohol consumption histories, being important to consider environmental and genetic factors that contribute to its early presentation. Clinical case: 35-year-old patient with SWK of alcoholic etiology assessed by neuropsychological study, magnetic resonance imaging studies and brain SPECT. He presented cognitive findings compatible with SWK. The MRI was normal and the SPECT was compatible with the SWK.

Conclusions: SWK can occur in young patients, and environmental and genetic factors may influence its early development.

Keywords: alcoholism, amnestic syndrome, Wernicke-korsakoff

Introduction

Wernicke-Korsakoff syndrome (WKS), a consequence of severe thiamine deficiency, is characterized by an acute phase usually presenting as a confusional syndrome associated with oculomotor and cerebellar signs, and a chronic phase in which diencephalic amnesia is prominent.[1–3] However, cases linked to malnutrition, hyperemesis gravidarum, anorexia nervosa and genetic factors, among others, have been widely described.[1,2,4]

Patients with WKS may present with a typical clinical triad of delirium, oculomotor disorders and ataxia.[2,3] However, the complete triad is only present in 33% of cases, so underdiagnosis of this pathology is often high, especially in young adults with relatively short histories of alcohol use.[4]

In the chronic stage, various cognitive disorders are evident, in addition to the characteristic diencephalic amnesic syndrome.[5] The damage caused by alcoholic neurotoxicity involves extensive frontal and limbic circuits, generating deficits in executive and frontal functions. Behavioral disturbances are frequent, mainly due to irritability, apathy and emotional lability.[1,2] Fabulation is characteristic, but not necessary, possibly linked to the prefrontal extent of the lesion.[5] Alcoholic subjects present vitamin deficiencies linked to multiple factors and similar levels of consumption or malnutrition do not necessarily imply the development of encephalopathy.[6] Environmental factors that contribute to the expression of encephalopathy should be considered, as well as the possible increase in individual susceptibility in relation to acquired or congenital alterations.[6] Conditions such as substance abuse or neurodevelopmental conditions are linked to a reduction in cognitive reserve, and may act as precipitating factors or as obstacles to patients' recovery.

Clinical case

We describe a 35-year-old right-handed man with 7 years of formal schooling. He has no chronic diseases or cardiovascular risk factors, encephalocranial trauma or cerebrovascular accidents. He has a 10-year history of substance abuse (cocaine base paste and marijuana), problematic alcohol consumption and smoking. Premature birth, with 7 months of gestation and low birth weight (2100 kg), with no requirements for assistance in CTI. No delays in developmental maturational patterns, although he has a history of school failure.

He presented at the emergency room with a confusional syndrome associated with cerebellar syndrome with ataxia. Paraclinical evaluation showed negative serology for HIV, VDRL, HCV, negative drugs in urine and CSF for virus and negative bacteriological cultures. The neurological physical examination showed a bilateral VI pair, with diplopia in all gaze positions, vertical nystagmus and ataxic gait that he could not walk without support. He presented with fabulations. From the initial imaging evaluation, a cranial MRI was obtained with no pathological findings in relation to the clinical data. He was admitted to the neurology ward for a month, with a diagnosis of Wernicke's encephalopathy, receiving pharmacological treatment with thiamine, diazepam and tiapridal. In the evolution he presents anterograde amnesia and difficulty in walking. He maintains a moderate degree of dependence for basic activities of daily living, mood with depressive elements, lack of initiative and apathy.

The neuropsychological evaluation showed a dys-executive syndrome with increased central processing time and reduced cognitive flexibility, a severe amnesic syndrome for both audio-verbal and visuospatial material and a decrease in conceptual functions (analogies 8/24). Brain perfusion SPECT with ^{99m} Tc-ECD showed moderate bilateral prefrontal, anterior temporal and right mesial moderate hypoperfusion, and in both thalami, following the typical pattern described for WKS (Figure 1). In the evolution he persists with loss of autonomy for basic and instrumental activities of daily living, unable to ambulate, requiring the use of a wheelchair, with inappropriate behaviors and subsequent institutionalization in a care center, configuring a dementia.

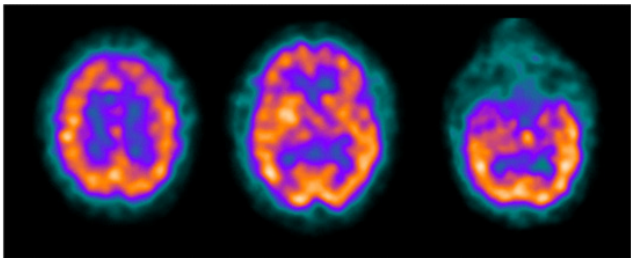

Figure 1 Brain perfusion SPECT with ^{99m} Tc-ECD showing moderate bilateral prefrontal, anterior temporal and right mesial hypoperfusion and in both thalami. Increased perfusion in the right striatum and hypoperfusion of the left striatum. The findings are compatible with Wernicke-Korsakoff encephalopathy.

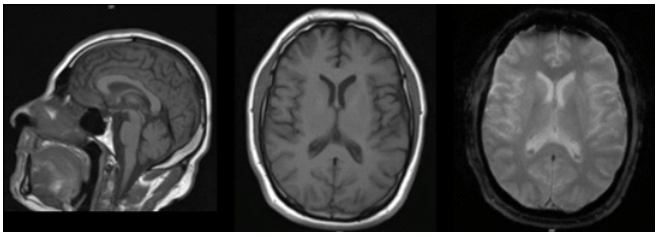

Figure 2 Magnetic Resonance Imaging without parenchymal lesions.

Discussion

Clinical variability and possible individual susceptibility to develop or not the syndrome could explain the underdiagnosis in young patients. In this case, which results from a toxic ingestion, with a few years of evolution, we consider three facts that should be highlighted. Firstly, the patient presents a series of factors that may have an impact on encephalic vulnerability: consumption of various drugs and a history of school failure without clear social causality, which may constitute an indicator (albeit indirect) of neurodevelopmental difficulties. The patient also shows a very low performance in the analogies test, indicating a decreased capacity for abstraction, which is also suggestive of neurodevelopmental difficulties. Secondly, the classic spontaneous evolution after thiamine replacement, with reduction of neurological signs such as ataxia, is not observed in this case either. This leads us to think that there are intercurrent factors that condition a low cognitive reserve, leading to a slower and more limited evolution in terms of clinical improvement, both from the cognitive and motor point of view.

Thirdly, the absence of structural lesions in MRI is highlighted, while a characteristic pattern is obtained in the functional imaging study (SPECT). This reaffirms the relevance of cerebral perfusion techniques in this type of diagnosis, in which the clinical manifestation and its variability is the result of several factors and early treatment makes the difference in terms of functional recovery of the patient. In relation to imaging techniques, cranial magnetic resonance imaging (MRI) is considered a method of great diagnostic value. With a specificity of 93% it is among the recommended methods both in cases of alcoholic and non-alcoholic patients.[8] In the acute stage it shows hyperintense symmetric changes in T2 and FLAIR sequences,[8] typically in periventricular regions - especially the third ventricle, thalamus, mammillary bodies and periaqueductal region.[8,9] Cortical atrophy can also be observed in orbitofrontal and mesotemporal regions.[9] However, the sensitivity of the technique is low, so that, despite being a useful tool, the absence of alterations does not exclude the diagnosis. In this sense, nuclear medicine techniques have proven to be very useful in the diagnosis of these pathologies. Currently, cerebral perfusion studies are the studies of choice in patients with WKS because they show greater sensitivity compared to structural techniques.[6]

Conclusion

Alcoholic WKS is infrequently observed in young adults. We present the case of WKS in a 35-year-old patient, who manifests with the complete initial clinical triad (delirium, oculomotor disorders and ataxia) and with typical cognitive and behavioral disorders compatible with the syndrome. Considering that most of the subjects with problematic alcohol ingestion do not develop WK encephalopathy, we propose that there are intercurrent factors at the cerebral level that favor the evolution of WKS or that do not provide the brain with sufficient resistance to the decline in its functioning when exposed to neurological alterations. Factors such as the use of various toxic substances, as well as educational history and possible neurodevelopmental alterations, may condition brain vulnerability in relation to the concept of cognitive reserve.

Structural damage could be directly linked to the time of consumption, and not so much to the severity of clinical symptoms, in young adults the diagnostic resource of functional imaging becomes very relevant. In addition, vitamin deficiency that responds to multiple factors is not a sufficient condition for the development of encephalopathy; individual vulnerability also responds to other environmental and/or congenital comorbidities. We also emphasize that the absence of structural damage referred in MRI is not an indicator of good prognosis in these cases.

In subjects with neurological pathology secondary to chronic alcohol toxicity, structural imaging techniques such as MRI show low sensitivity. As in the case presented, the absence of alterations in this image does not exclude the diagnosis, so studies such as brain SPECT are of greater utility and certainty in patients with WKS as they are more sensitive.

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Conflicts of interest

The authors declare no conflicts of interest.

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