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Huntington's disease and dementia: from structural changes to clinical issues

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Background

Huntington's disease (HD) is an autosomal dominant neurodegenerative disorder, which occurs in patients with a mutation in the IT15 gene (huntingtin). Clinically, the disease presents itself with not only motor symptoms but also dementia. The aim of this study is to review and present all recent data, specifically those related to the neuropathology, biochemistry and diagnostic methods of dementia in HD.

Materials and methods

Recent advances in molecular and genetic research of HD, derived from articles published in journals indexed in PubMed and other Entrez medical databases are being reviewed in an effort to elucidate mechanisms of cell death in the striatum and disruption of cortical-striatal circuitry. Neurophysiological and imaging diagnostic methods are analytically described, while the utility of other rating scales, like the Unified HD Rating Scale, in the differential diagnosis between HD and other dementias is evaluated according to several clinical trials, described in various biomedical literature citations.

Results

HD causes widespread CNS changes and systemic abnormalities, while cell death mechanisms involve variable processes, such as mitochondrial abnormalities, excitotoxicity, neuroinflammation and abnormal protein degradation [1]. Genetic testing, consideration of the inherited disease risk, clinical assessment, neuroimaging techniques, cognitive and psychological rating scales contribute significantly not only to HD diagnosis, but also prognosis [2].

Conclusions

HD is a devastating neurological condition of long duration. Many factors likely contribute to neuron death and dysfunction, making the systematic address of its pathology difficult. The first signs and symptoms are often present before impairments reach a point where the neurologic disease manifests itself, but usually are subtle and remain unnoticed [3,4]. For this reason, clinical rating should focus not only on motor difficulties, but also on the neuropsychological performance of patients as a whole, aiming to detect different types of impairment within this neuropsychiatric disorder. Further research in relation to the understanding of the mechanisms involved in the memory and cognitive impairment of HD and enrichment of the diagnostic tools of all cognitive and emotional declines early in the disease process can have implications for prognostic assessment of persons at risk and eventually assist with early interventions.

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