P02-196 - **CASE REPORT: BEHAVIOUR ALTERATION WITH BRAIN IRON ACCUMULATION**

M. Lazaro, N. Echeverria, J.R. Rodriguez, F. De La Torre, S. Escribano, J.I. Bango

Nuestra Señora de Sonsoles hospital, Avila, Spain

Objectives: To analyze psychiatric symptoms in relation to a case of Hallervorden-Spatz disease (*neurodegeneration with brain iron accumulation* (NBIA) or *pantothenate kinase-associated neurodegeneration* - familial brain degeneration with iron deposition in brain).

Methods: Pubmed revision on behaviour alteration and its relation to brain iron accumulation. Review of patient medical records, including image studies.

Results: A 52-year-old female with diagnosis of mental retardation and psychosis was treated and followed since 1999. Severe behaviour alteration motivated hospital admission in 2009. Over the last two years, the patient had developed progressive dementia, choreoathetosis, mutism, ideomotor apraxia, urinary and fecal incontinence, and corticospinal signs.

A brain MRI (2008) revealed iron deposits in basal ganglia, hypointensity with an area of central hyperintensity ("eye-of-the-tiger"-sign) in both globus pallidi on T2. Based on clinical and MRI findings a diagnosis of Hallervorden Spatz Disease was made.

Classic form of the disease is characterized by early onset and rapid progression, culminating in early death. Atypical disease, as in our case report, has a later onset and more slowly progressive course.

Systemic chelating agents have not proved beneficial. Treatment remains symptomatic. Our patient has experienced a favourable response to low-dose trazodone and quetiapine.

Conclusions: At times, behavioral changes may predate neurologic manifestations, whereas at other times disturbances in mental status and physical functioning may coexist. Among patients with NBIA, those with atypical disease are much more likely to have psychiatric symptoms with cognitive decline. These features present in our case report usually make a difficult and late diagnosis and treatment.

https://doi.org/10.1016/S0924-9338(10)70811-7 Published online by Cambridge University Press