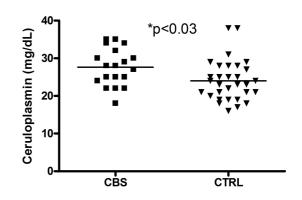
Corticobasal syndrome and copper metabolism: a pilot study

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Introduction

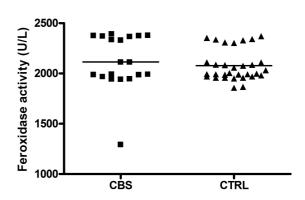
Corticobasal syndrome (CBS) is an heterogeneous degenerative disorder where paramagnetic deposits are often found in basal ganglia and deep cerebellar nuclei at brain MRI.¹ Furthermore, its clinical expression with parkinsonism and hyperkinetic features (dystonia, myoclonus) reminds disorders due to heavy metal deposition (e.g., aceruloplasminemia).



Methods

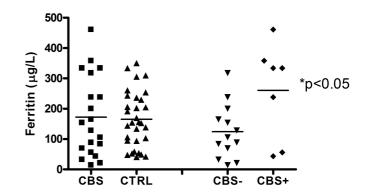
Aim of this pilot work was to assess selected parameters of heavy metal metabolism in these patients.

Serum levels of ceruloplasmin (CP), ferroxidase activity and ferritin were assessed in 20 CBS patients negative for progranulin mutations and 20 matched healthy controls. Brain MR scan was analyzed in order to define the presence of signal alterations compatible with paramagnetic depositions (CBS+ vs. CBS-, n=7 vs. 13).



Results

- •Serum CP was mildly elevated (~15%, p< 0.03) in CBS patients with respect to CTRL
- •Serum ferroxidase activity and ferritin levels were not different between the two groups
- •Seven CBS patients (35%) displayed at brain imaging various degrees of signal alterations compatible with paramagnetic material
- •Analyzing separately this subgroup (CBS+), ferritin levels were doubled with respect to CBS- patients (p<0.05), while no differences were present for the other two parameters.



Discussion

These preliminary data do not support the idea that altered copper metabolism plays a major role in CBS, albeit altered iron metabolism might play a role in a subgroup of patients. However, considering both CP and ferritin roles as markers of the inflammatory response, we cannot exclude that the reported changes reflect differences related to this latter phenomenon.

Ref. ¹Hauser RA, Murtaugh FR, Akhter K, Gold M, Olanow CW. Magnetic resonance imaging of corticobasal degeneration. J Neuroimaging. 1996;6:222-6.



