

Idiopathic normal pressure hydrocephalus and behavioral variant of frontotemporal lobar degeneration: an unexpected association

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Introduction

Idiopathic normal pressure hydrocephalus (iNPH) has a complex multifactorial pathogenesis probably linked with neurodegeneration. Several studies have underlined the association with Alzheimer's disease (AD). In contrast, the association with behavioural variant of frontotemporal lobar degeneration (bv-FTLD) has only been suggested by one case-report (Korhonen et al. *Fluids Barriers CNS* 2017 14:10) and a genetic study (Korhonen et al. *Dement Geriatr Cogn Disord* 2019;47:91–103). In order to provide further evidence of this link, we set out to compare prevalence of iNPH between cohorts of AD and bv-FTLD patients.

Methods

Patients with AD and bv-FTLD, seen in consultation at the Memory Center of Nantes in 2019, were retrospectively compared according to iNPH prevalence. In a second step, three groups of 9 patients with either bv-FTLD, iNPH or the double diagnosis (iNPH-bv-FTLD) were thoroughly compared on clinical, radiological and biological data.

Results

5 from 69 (7,25%) bv-FTLD patients displayed iNPH diagnosis while it was only diagnosed in 2 from 178 (1,1%) AD patients ($p=0.02$). iNPH-bv-FTLD patients show characteristic features of both iNPH (notably basal ganglia hypometabolism, decreased CSF AD biomarkers and gait improvement with CSF derivation) and bv-FTLD patients (notably hyperorality and perseverations).

Conclusion

Our data are the first from cohorts of patients with neurodegenerative diseases to support the link between iNPH and bv-FTLD. They should encourage neurologists to look for iNPH in there bv-FTLD patients in case of gait trouble occurrence. Further research is needed to prospectively confirm these results and pathologically assess these newly described patients.

Conflicts of interest

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