Normal Pressure Hydrocephalus presenting as behavioural-variant FrontoTemporal Dementia: a case report.

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Background. Idiopathic Normal Pressure Hydrocephalus (iNPH) is a syndrome characterized by a varying combination and degrees of gait disturbance, urinary incontinence and dementia. It is caused by the enlargement of cerebral ventricles and the consequent interstitial edema in periventricular white matter, with suffering of prefrontal pathways.
Sometimes, cognitive and/or neuropsychiatric involvement can be predominant and differential diagnosis with neurodegenerative or neuropsychiatric disorders could be arduous.

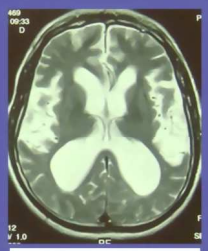
Aims. To describe a case of behavioural-variant Frontotemporal Dementia-like syndrome due to iNPH.

Methods and Results. In 2008, a diabetic and dyslipidaemic 57-years old man, after been laid off, started complaining of apathy, aboulia and diurnal somnolence. A diagnosis of depression of mood was done.

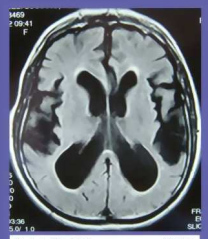
Subsequently, decline in social manner, impairment in regulation of personal conduct, emotional blunting, behaviour and speech change insidiously arose and slowly progressed. General and neurological examination were normal and no urinary difficulties were present. A clinical diagnosis of behavioural-variant Frontotemporal Dementia was done and patient started assuming galantamine extended release until 16mg per day.

In 2012 patient presented to our ambulatory referring a mild worsening of behavioural and cognitive problems, unclear gait difficulties and urinary incontinence. Neurological examination revealed slow, wide-based and shuffling gait with mild disequilibrium. Patient underwent to complete neuroradiological assessment and neuropsychological battery. Cerebral MRI showed ventriculomegaly, thinning and upward elevation of corpus callosum and periventricular hyperintensities in FLAIR-sequences.
Diagnosis of probable iNPH was made; iNPH-grading scale (assessed according to Kubo et al) was 8/12 (2+3+3).

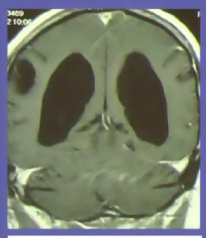
A six months follow-up revealed normalization of gait and urinary disturbances and a remarkable improvement of global neuropsychological profile, especially executive functions (iNPH-grading scale was 3/12: 1+1+1).



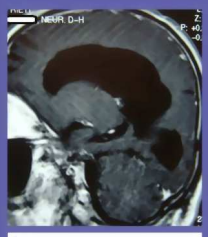
Axial T2-weighted sequences MRI: enlargement of cerebral ventricles.



Axial FLAIR sequences MRI: transependimal edema.



Coronal T2-weighted sequences MRI: narrowing of CSF spaces.



Sagittal T1-weighted sequences MRI: thinning and upward elevation of corpus callosum.

Neurosurgical assessment.

- ✓ phase-contrast MRI identified an increased *stroke volume* (4.8µL), suggestive of iNPH;
- ✓ trought a right frontal ventriculostomy, a "Rickham" catheter was positioned in the right frontal horn;
- ✓ intraventricular *infusion test* confirmed the decrease of intracranial compliance (*elastance*).

Ventriculo-peritoneal shunt was performed.

Neuropsychological evaluation.

Cognitive functions were assessed using tests as follows:

- MMSE (Mini Mental State Examination);
- Memory:
  - Rey Auditory Verbal Learning Test (RAVLT): immediate recall and delayed recall, and recognition;
  - Digit Span Test (Verbal and Spatial);
- Attention and Executive Functions: Trail Making Test (TMT), Visual Search, Frontal Battery Assessment (FAB), Verbal Fluency (phonemic and semantic);
- Deductive Intelligence: Raven Matrices;
- Language: Boston Naming Test.

Behavioural disorders were assessed using behavioural scales, as follows:

- FBI (Frontal Battery Inventory);
- NPI (Neuropsychiatric Inventory)

Pre-shunt. Severe deficits were observed in many cognitive functions: attention/executive functions, memory and visuo-spatial functions. Patient showed psychomotor slowing and reduced speech production, and pathological performances were obtained on the delayed recall, spatial span, phoneme fluency, TMT-A and FAB. Indifference, emotional lability, irritability and depression of mood were mainly observed among behavioural disorders. All the cognitive and behavioural pathological patterns were typically associated to subcortical frontal dysfunction.

Post-shunt. A neuropsychological assessment was repeated after 6 months. Clinically, a wide cognitive (i.e. memory, executive functions) and behavioural improvement was observed.

Conclusions. Since iNPH is one of the rare form of potentially reversible dementia, clinicians should keep in mind it can display a pure neuropsychiatric and/or cognitive onset and such remain for a long time. Complete neurological, neuropsychological, neuroradiological and neurosurgical analysis is essential to avoid a late diagnosis.

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