

Unusual cognitive presentation of frontotemporal brain sagging syndrome (FBSS)

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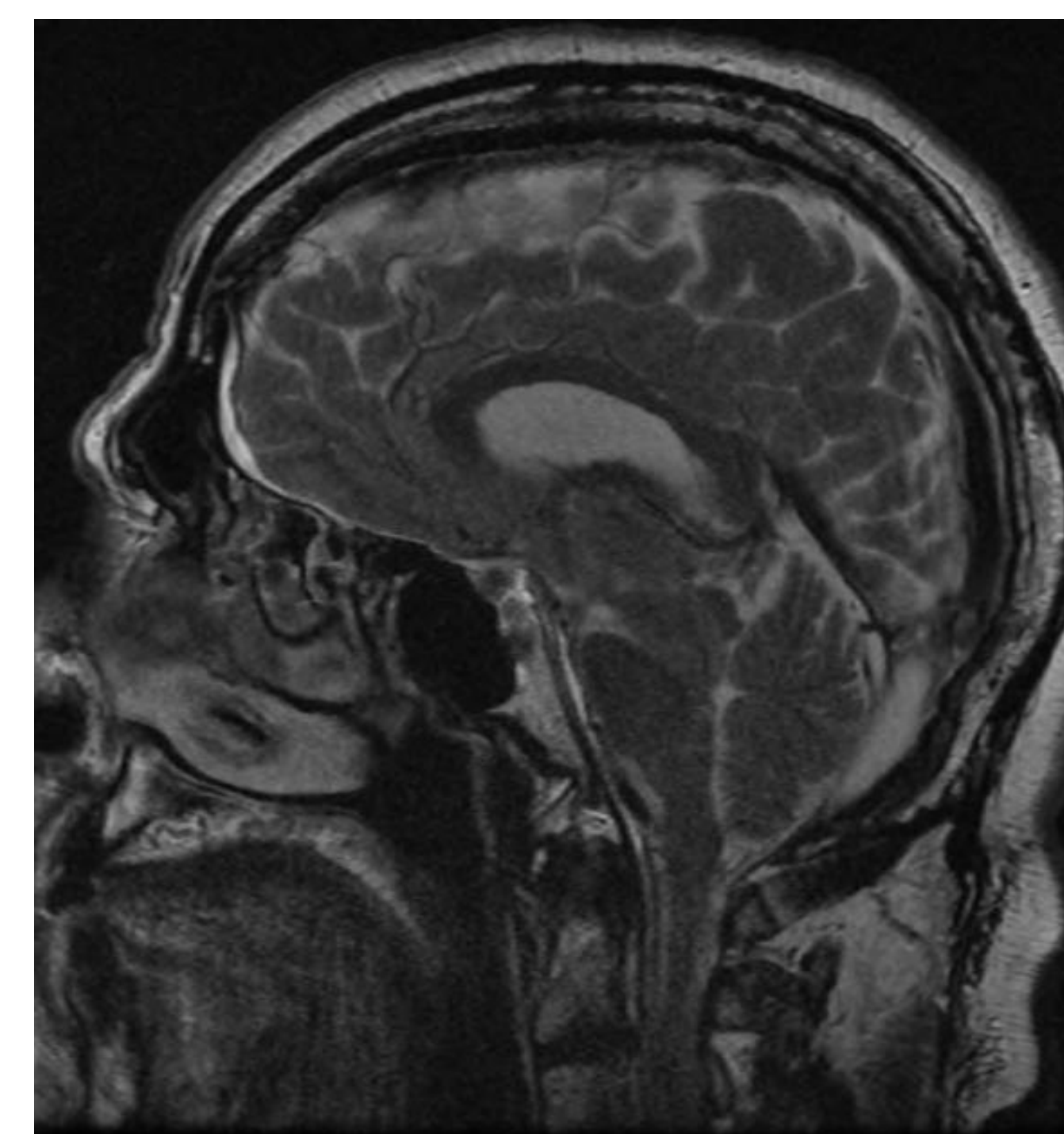
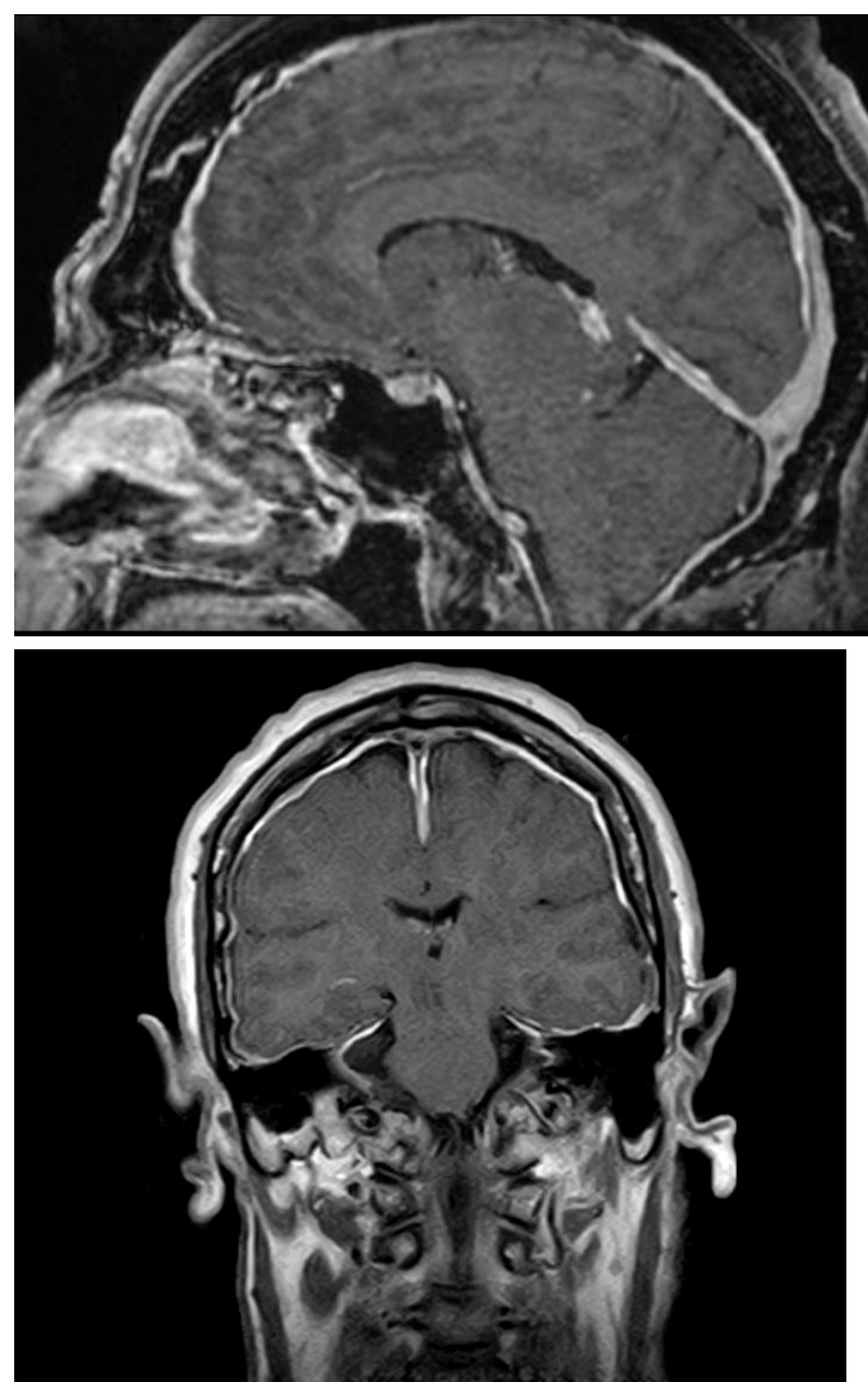
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Objective: to highlight the importance to recognize potentially reversible/treatable syndromes mimicking bvFTD (behavioural variant frontotemporal dementia).

Materials and Methods:

A 72 year-old man came to our attention with a diagnosis of bvFTD. Behavioural symptoms (disinhibition, depression associated to mild cognitive dysfunction) appeared 5 years before; mild gait disturbances were reported. Valproate and duloxetine were not efficacious; brain MRI without gadolinium was reported normal except for Arnold Chiari type I malformation. After 1 year he complained of sudden severe headache and dysarthria, with evidence of venous sinus thrombosis, treated with warfarin with remission; monoclonal k MGUS and high levels of S protein emerged. Due to mild worsening of behavioral symptoms, brain perfusion SPECT was performed, with evidence of frontotemporal bilateral hypoperfusion. Neuropsychological evaluation demonstrated compromise of frontal functioning, supporting a diagnosis of bvFTD; treatment with memantine and rivastigmine was recommended, with partial benefit. Cognitive and behavioral profile remained substantially unchanged during the following years. Brain MRI performed after our first evaluation showed pachymeningeal enhancement with increased thickness and findings resembling frontotemporal brain sagging syndrome (FBSS); revision of previous brain MRIs pointed out that initial radiological signs were already recognizable. Spinal MRI did not recognize possible sites of CSF leak.

Results and Discussion: FBSS is characterized by downward displacement of the brain ("sagging") and may have iatrogenic/traumatic etiology or occur spontaneously; dural rupture and CSF leakage can be demonstrated in a minority of cases. As recently reported, bvFTD can be a clinical presentation of FBSS, usually with slow progression and insidious onset. Hypometabolism of frontal/temporal lobes is commonly observed, male sex is prominent and headache is frequently reported; symptoms suggestive of dysfunction of deep midline structures can be associated.



Figures: Coronal and sagittal view post-contrast showing diffuse dural thickening and enhancement. Both T1 and T2 sagittal view show descent of the central core brain structures (severely "sagging" brain) determining decreased amplitude of the suprasellar and peri-truncular cisterns, splenium depression, midbrain "slumping" and closed pons-midbrain angle.

Conclusions: few atypical clinical characteristics of our patient (slow evolution, associated gait disorders) and lack of frontotemporal atrophy, commonly described in bvFTD even if not mandatory, suggested further investigations and led to a diagnosis of possible FBSS. Our experience highlights the importance to recognize syndromes mimicking bvFTD as soon as possible, since they are potentially reversible only in early phases of the disease.

Bibliography:

- Frontotemporal brain sagging syndrome. An SIH-like presentation mimicking FTD. Wicklund M. R., et al. Neurology 2011.
- Uplifting news for frontotemporal dementia. Lippa C. F., et al. Neurology 2011.