Frontotemporal lobar dementia in a young woman

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A 65-year-old woman was addressed to the Department of Radiology for assessment of dementia. Mnestic tests found moderate memory problems with mini-mental state (MMS) measured at 23/30. Symptoms were essentially marked by behavioral disorders of gradual onset for 2 years (decline in activity and empathy for her family). She was addressed for assessment of Alzheimer’s type dementia. MRI examination (coronal and sagittal T1 weighted sequences) found a severe asymmetrical cortico subcortical atrophy dominant at the right frontal lobe and anterior portion of right temporal lobe (Fig. A and B, white arrows). Hippocampal regions were preserved (Fig. A, asterix). The diagnosis of frontotemporal lobar dementia (FTLD) was therefore suggested.

Comment

Dementias in young patients are characterized by the occurrence of symptoms before the 65 years of age (1). Their etiologies and atypical clinical presentation (behavioral, cognitive, neurological and psychiatric disorders) of dementia are different of older patient. Alzheimer’s disease and lobar frontotemporal degeneration are the most frequent degenerative causes of these dementias. Lobar frontotemporal degeneration includes a set of pathologies characterized by focal atrophy of frontal and temporal lobes. The onset of symptoms is between 45 and 60 years. Symptoms appear insidiously with behavioral disorders, personality changes, loss of empathy and motivation. Three clinical forms are described as dominate signs: FTLD (behavior disorders), non-fluent primary progressive aphasia and semantic dementia (language disorders). In Alzheimer’s disease, memory and spatial orientation disorders predominate. MRI is the key examination in the etiological investigation; it will clarify the mechanism of dementia (degenerative, vascular, infectious, inflammatory, metabolic or toxic). The protocol includes volumetric T1 weighted sequence to search and locate brain atrophy; coronal T2 weighted sequence allowing better temporal analysis, axial Flair weighted sequence searching for leukoaraphy (vascular, metabolic, inflammatory or infectious), axial T2 EG weighted sequence looking for microbleeds (vascular or metabolic etiology), and diffusion weighted sequence to search stroke etiology or arguments for a Creutzfeldt Jacob’s disease (Haute Autorité de santé, France, 2011). In FTLD, MRI examination shows a frontal lobe atrophy (in particular fronto-orbital cortex) and anterior temporal lobe (Fig. A and B). In Alzheimer’s disease, MRI examination shows medial temporal lobe atrophy (hippocampus, tonsils and entorhinal cortex) (Fig. C, black arrows).

Reference