49 year-old female
Cognitive impairment more than 2 years of evolution

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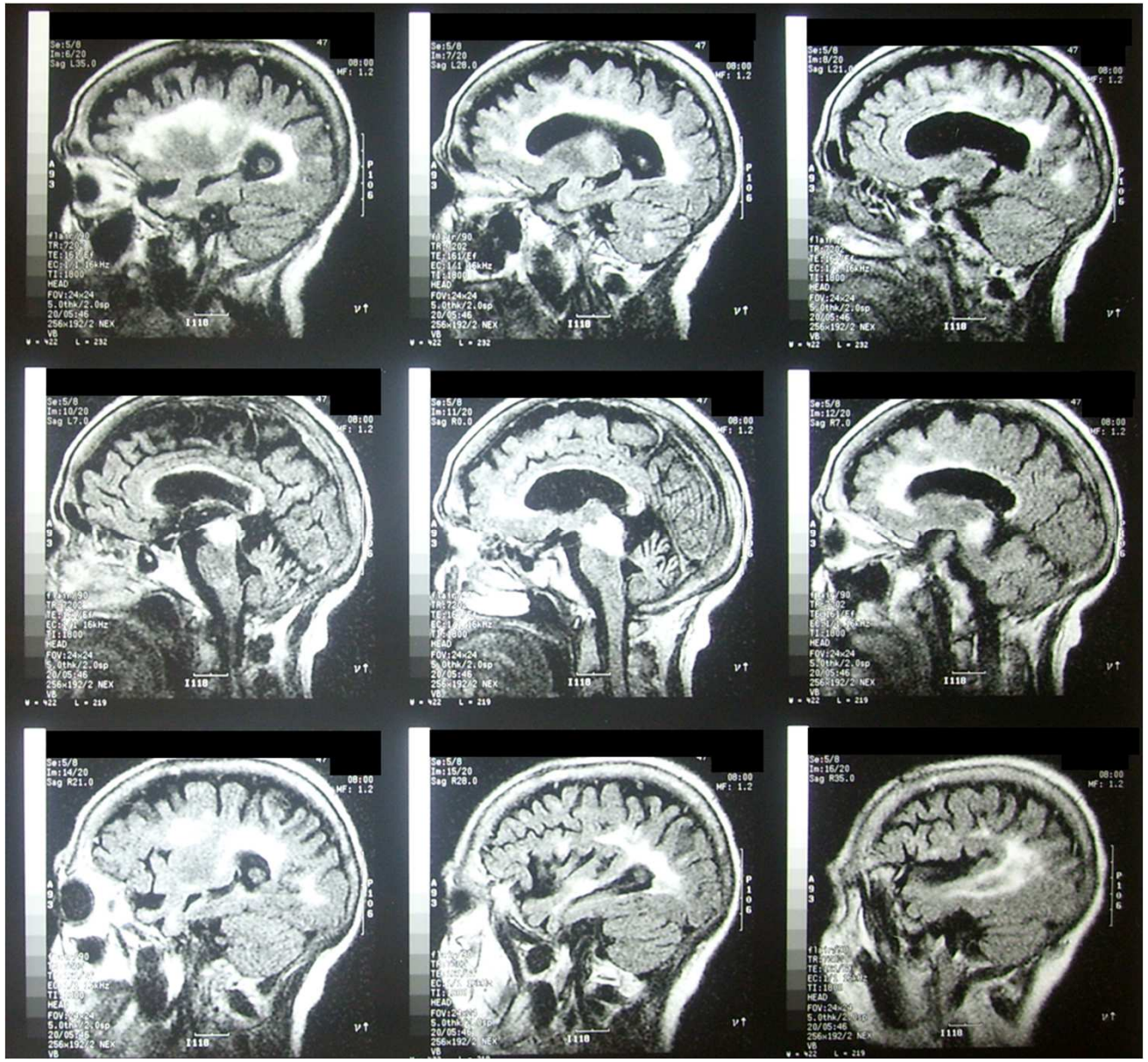
Clinical statement

- 49 y/o female.
- No vascular risk factors.
- Father died of dementia. Older brother with advanced dementia catalogued as vascular (Binswanger's disease).
- Cognitive impairment more than 2 years of evolution.
- Neuropsychological study: Dementia. Probably Alzheimer's Disease type.
- No other neurological symptoms

MRI T1 Flair. Severe cortical atrophy. Corpus callosum atrophy. Extense diffuse white matter hyperintensities. Probable Demyelinating Disease.



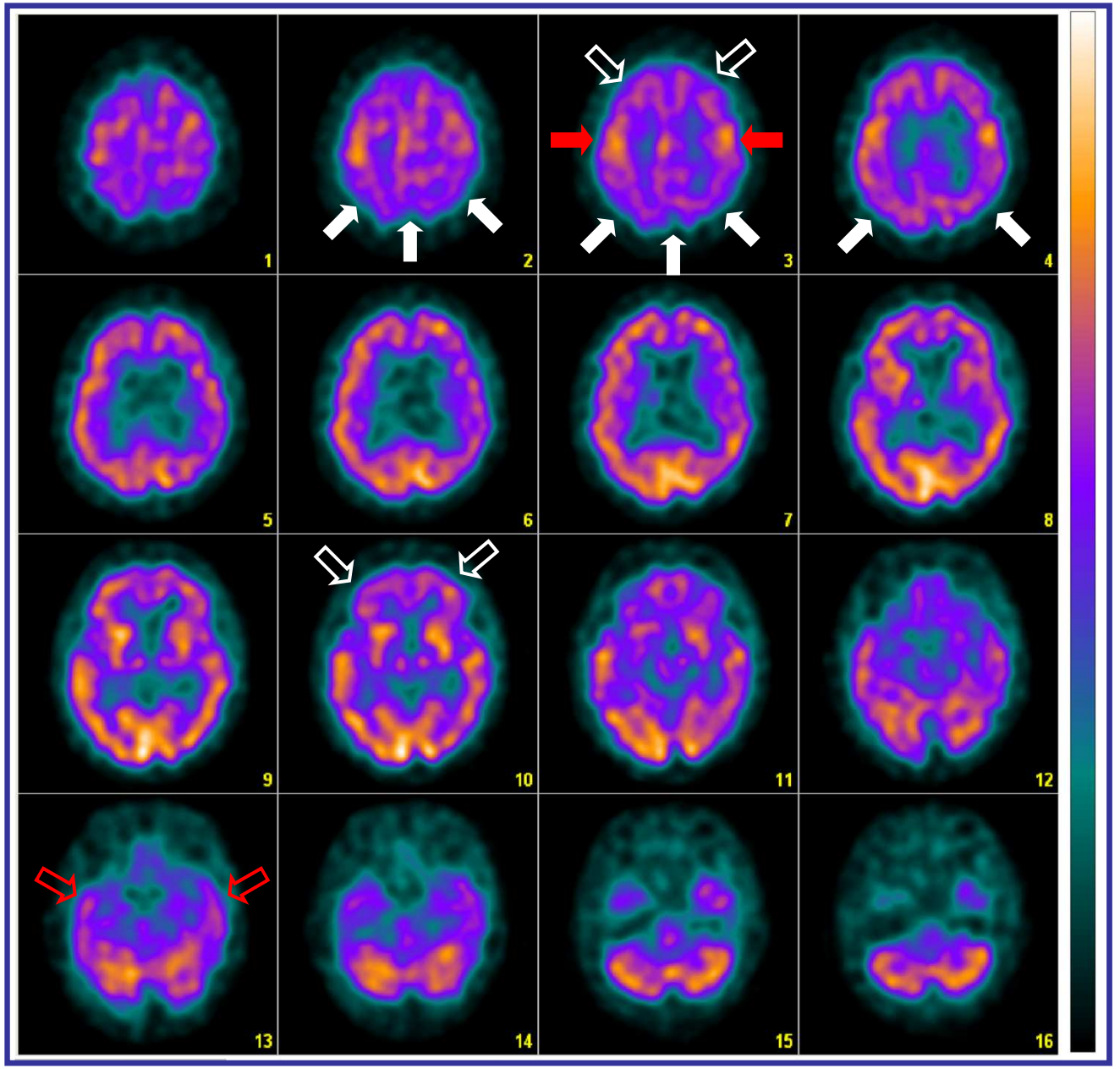
- Brain SPECT is indicated for further evaluation in a young patient with dementia with neuropsycological study consistent with AD and MRI discordant findings.

- Images were acquired in a dual head gammacamera 60 min. p.i. of 99mTc-ECD (925 MBq).

- 128 steps, 25 seconds each. 128×128 matrix. 2.9 mm pixel size. No scatter correction was performed.

- OSEM reconstruction (5 cycles 2 subsets). Prefiltering with Butterworth order 10, cut-off frequency 0.25. Attenuation correction 12 cm-1. Transaxial slices parallel to AC-PC line.

Bilateral posterior parietal, precuneus and posterior cingulate hypoperfusion (white arrows). Bilateral temporal (red), prefrontal (white) and thalamic hypoperfusion. Preservation of primary sensoriomotor cortex (red), occipital cortex, basal ganglia and cerebellum



Interpretation

- Pattern typical of AD (highly specific) unless thalamic involvement (correlated with MRI abnormalities).
- Dementia in a young patient with family history: Early onset familial AD.

Discussion

- Demyelinating disease very unlikely to present with this functional pattern.
- Unusual clinical findings may be present in familial AD (myoclonus, early aphasia or change in behavior, motor deficits, other neurologic symptoms). Diffuse white matter hyperintensities have been previously described.
- Brother of the patient likely to have the same type of dementia (white matter hyperintensities misinterpreted as Binswanger).
- Presenilin-1 gene mutation was detected.

Conclusion

- Brain SPECT is indicated in the diagnosis of dementia when the etiology remains uncertain after a complete clinical evaluation including neuropsycological study, laboratory tests and structural imaging.
- Early onset familial AD can often present with unusual findings and SPECT is likely to add valuable additional information in these patients.

Teaching points

- Brain SPECT in the diagnosis of dementia
- Brain SPECT in early onset dementia – familial AD

References

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