Teaching Neuro*Images*: Nonfluent variant primary progressive aphasia

A distinctive clinico-anatomical syndrome

OPEN

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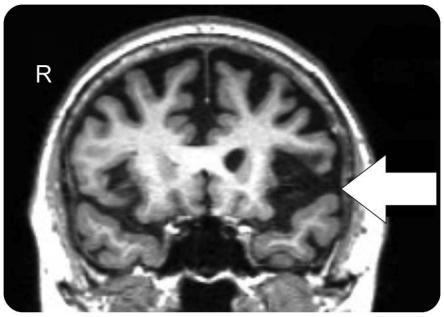
A 66-year-old woman presented with 4 years of progressive speech difficulty. She had nonfluent speech with phonemic errors but intact single-word comprehension and object knowledge. Her grammar was impaired in both speech and writing, and she exhibited orofacial apraxia. A clinico-radiologic (see figure) diagnosis of nonfluent variant primary progressive aphasia was made.

Figure Magnetic resonance image



Coronal volumetric T1-weighted MRI showing asymmetric atrophy of left insula and opercular inferior frontal gyrus (arrow), a pattern typical of nonfluent variant primary progressive aphasia.

Nonfluent variant primary progressive aphasia is a neurodegenerative disease within the spectrum of frontotemporal dementia, characterized by the typical language and brain atrophy patterns seen here.[1] It is most frequently due to tau pathology, and clinicians should be alert to the potential development of progressive supranuclear palsy or corticobasal syndrome.[2]

AUTHOR CONTRIBUTIONS

C.R.M.: image selection and drafting of the manuscript. C.J.D.H., M.N.R., J.D.W.: critical revisions of the manuscript.

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DISCLOSURE

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REFERENCES

1. Gorno-Tempini ML, Hillis AE, Weintraub S, et al. Classification of primary progressive aphasia and its variants. Neurology 2011;76:1006–1014.
2. Josephs KA, Duffy JR. Apraxia of speech and nonfluent aphasia: a new clinical marker for corticobasal degeneration and progressive supranuclear palsy. Curr Opin Neurol 2008;21:688–692.

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