Journal of Clinical Neuroscience 62 (2019) 216–217

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Images

A case of profound asymmetry in motor skills

José Rafael P. Zuzuárregui [a,*], Anna D. Hohler [b], Krishe Menezes [a]

[a] Division of Neurology, University of California, San Francisco, Fresno Center for Medical Education and Research, 155 N. Fresno St., Fresno, CA 93701, United States
[b] Department of Neurology, Saint Elizabeth's Medical Center, 736 Cambridge St., Brighton, MA 02135, United States

1. Clinical scenario

A 71 year-old female with thyroid disease presented for rapidly progressive cognitive dysfunction. Three years prior, the patient began to experience difficulty with performing motor tasks and neglecting the right side of her body. Attention and concentration also declined. Gait slowed with shortening of stride length. Memory and ability to name objects remained intact. She had no hallucinations, gaze limitation, dream enactment behavior or myoclonus. She had been placed on donepezil and paroxetine with no improvement.

Neurologic examination revealed frontal release signs, ideomotor apraxia and hypomimia. Intermittent dystonia, rigidity and astereognosis of the right hand were seen. Gait examination revealed retropulsion and a slowed gait. Initial testing was within normal limits, including a basic metabolic panel, complete blood count, vitamin B12, thyroid stimulating hormone, Venereal Disease Research Laboratory test, C-reactive protein and erythrocyte sedimentation rate.

Cerebrospinal fluid (CSF) testing revealed one red blood cell/uL, one white blood cell/uL, protein of 29 mg/dL, glucose of 54. CSF culture, gram stain and 14-3-3 protein were both negative. Further CSF analysis revealed A-beta 42 (734.75 pg/ml), phospho-tau (70.55 pg/ml) and total-tau levels (453.95 pg/ml) with A-beta 42/total-tau index (ATI; 0.95), borderline for Alzheimer's disease (Values for Alzheimer's: phospho-tau >68 pg/ml; ATI >1.2). Magnetic resonance imaging (MRI) of the brain showed atrophy of the left parietal lobe (Fig. 1). Positive emission tomography (PET) demonstrated reduced uptake in the left parietal lobe (Fig. 2).

The most likely diagnosis is

A: Rasmussen's encephalitis
B. Corticobasal degeneration
C. Idiopathic Parkinson's disease
D. Progressive Supranuclear Palsy
E. Alzhemier's disease

2. Answer

B. Corticobasal degeneration

3. Discussion

In the setting of asymmetric parkinsonism and dystonia, as well as ideomotor apraxia and a cortical sensory deficit on examination, the patient met criteria for probable corticobasal degeneration of the corticobasal syndrome subtype. This was further supported by asymmetric MRI and PET findings, as well as neuropsychological testing.

Corticobasal degeneration (CBD) is an atypical parkinsonian syndrome with marked asymmetry of symptoms related to deposition of hyperphosphorylated tau in the cortical and basal ganglia [1,2]. Onset of symptoms typically occurs in the sixth decade and includes asymmetric hand clumsiness, bradykinesia, tremor, and rigidity [1,2]. Typical features on examination include tremor, dystonia, alien limb phenomenon, apraxia, myoclonus, cortical sensory loss, postural instability and gait dysfunction [1,2].

Previously a pathologic diagnosis, CBD has since been divided into probable and possible categories [1]. This is further subdivided into four phenotypes: corticobasal syndrome (CBS), frontal behavioral-spatial syndrome (FBS), nonfluent/agrammatic variant of primary progressive aphasia (naPPA), and progressive supranuclear palsy syndrome (PSPS) [1,2]. At this time, imaging findings are not included as part of the proposed diagnostic criteria for CBD [1]. However, T2-weighted and FLAIR images may reveal asymmetric atrophy of the posterior frontal and parietal lobes [3]. Asymmetric hypometabolism can also be seen on ^{18}F-fluorodeoxyglucose PET imaging [3].

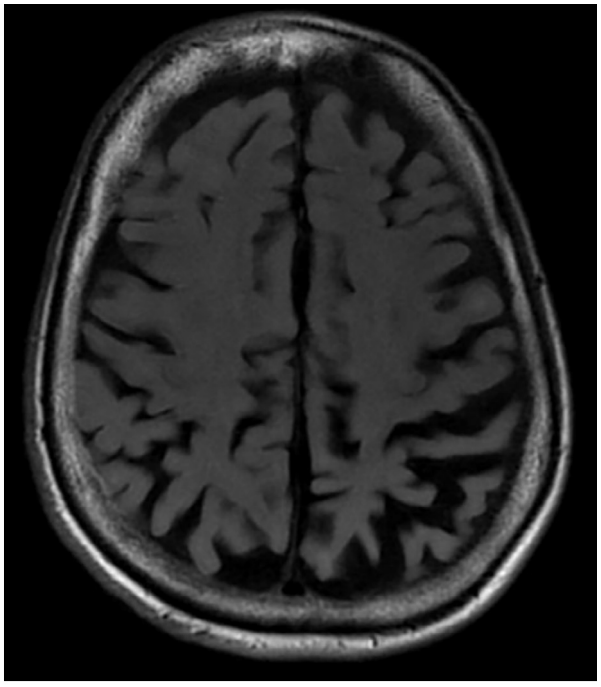
Corticobasal degeneration is poorly-responsive to levodopa and there are no disease-modifying treatments or approved pharmacological or therapeutic at this time [4]. Treatment is typically supportive and aimed at optimizing quality of life [4].

4. Contributors

Drs. Zuzuárregui and Menezes were responsible for the concept design. All authors participated in the drafting of the manuscript.

* Corresponding author at: University of California, San Francisco, Fresno Medical Education Program, 155 N. Fresno St., Fresno, CA 93701, United States.
E-mail addresses: jzuzuarregui@ucsf.fresno.edu (J.R.P. Zuzuárregui), Anna.Hohler@steward.org (A.D. Hohler), krishe.menezes@ucsf.fresno.edu (K. Menezes).

https://doi.org/10.1016/j.jocn.2018.12.021



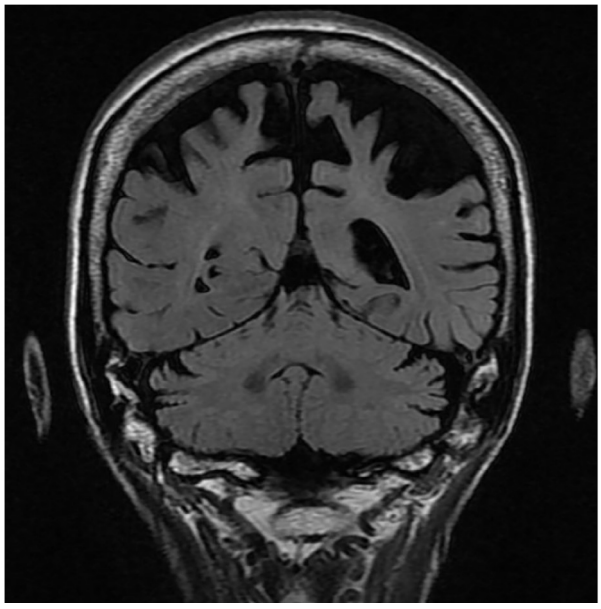


Fig. 1. An axial (A) and coronal (B) T2-weighted brain MRI showed atrophy of the left parietal lobe out of proportion to that seen in the right hemisphere.

5. Conflicts of interest/disclosures

The authors do not have any financial or other conflicts of interests to declare.

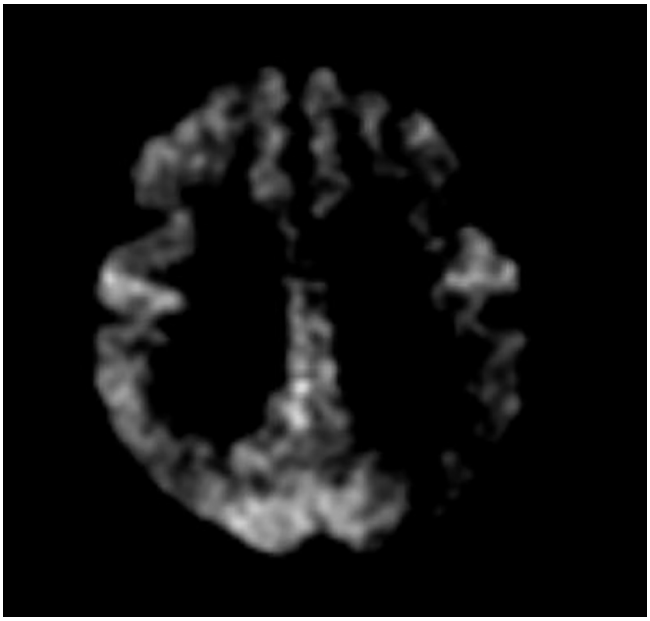


Fig. 2. Brain PET imaging showed reduced fluorodeoxyglucose uptake in the left parietal lobe.

References

[1] Armstrong MJ, Litvan I, Lang AE, et al. Criteria for the diagnosis of corticobasal degeneration. Neurology 2013 Jan 29;80(5):496–503.
[2] McFarland NR. Diagnostic approach to atypical parkinsonian syndromes. Continuum (Minneap Minn) 2016:1117–42 (4 Movement Disorders).
[3] Ogawa T, Fujii S, Kuya K, et al. Role of neuroimaging on differentiation of parkinson's disease and its related diseases. Yonago Acta Med 2018;61 (3):145–55.
[4] Lamb R, Rohrer JD, Lees AJ, et al. Progressive supranuclear palsy and corticobasal degeneration: pathophysiology and treatment options. Curr Treat Options Neurol 2016 Sep;18(9):42.