# CASE 3

# 73 Y/O WOMAN

# Brief history

- **Past history**
  Type 2 DM

- **Chief complain**
  Progressive memory loss & delusional jealousy

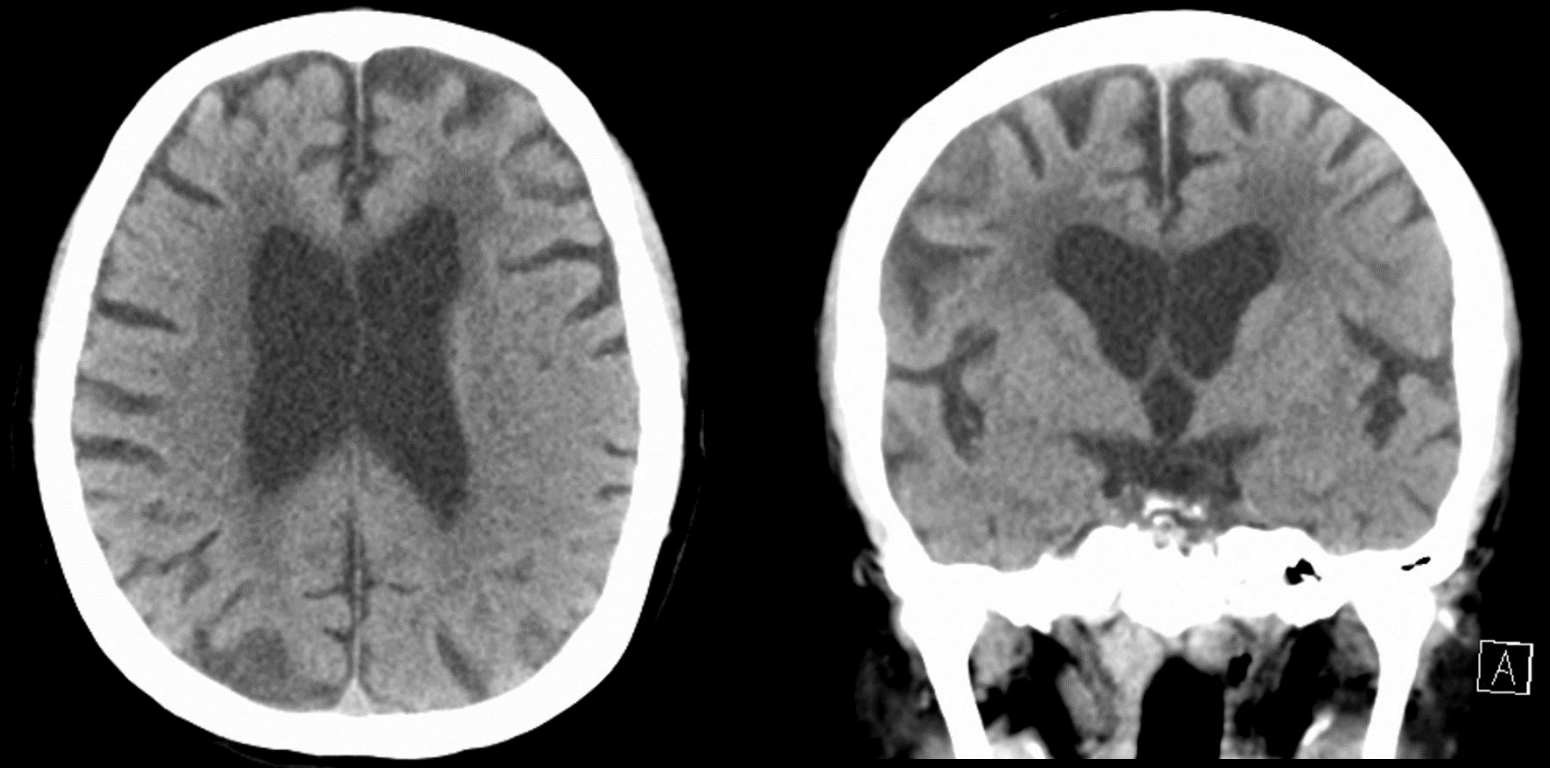
# Imaging studies

Brain CT (2015.10.29)

Brain MRI (2020.03.28)

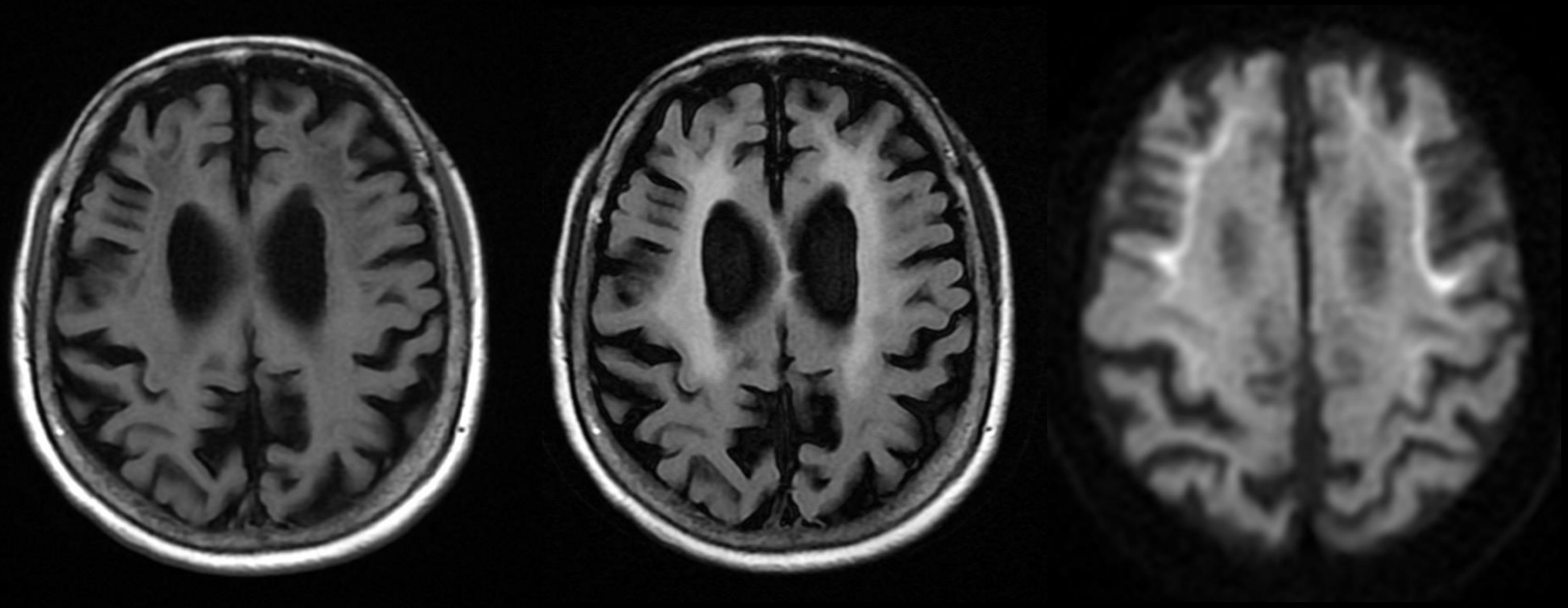
# Brain CT

- ✓ Brain atrophy and chronic periventricular white matter ischemic change



# Brain MRI

- ✓ Senile changes with cortical atrophy over the bilateral fronto-temporal regions with sulcal widening and mild ventricular dilatation
- ✓ Frontal periventricular WM hyperintensities on T2/FLAIR and subcortical hyperintensities along the U fiber on DWI



# Differential Diagnoses

- Adult- onset neuronal intranuclear inclusion disease (NIID)
- Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL)
- Hereditary diffuse leukoencephalopathy with spheroids (HDLS)

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# Adult- onset NIID

- **Rare progressive neurodegenerative disorder** characterized by eosinophilic ubiquitin-positive intranuclear inclusions within neuronal and somatic cells
- Associated with genetic changes in the **NOTCH2NLC gene**
- Typical symptoms include **dementia**, **cognitive dysfunction**, **autonomic impairment** and **behavioral abnormalities**
- **Bilateral curvilinear lace-like hyperintensities along the cortico-medullary junction** in DWI, along with **brain atrophy** and **confluent symmetrical leukoencephalopathy** in T2/FLAIR images
- Pathologic diagnosis can be made via **skin or rectal biopsies**

# Adult- onset NIID