Klüver-Bucy syndrome as onset presentation of extremely young onset behavioural variant frontotemporal dementia

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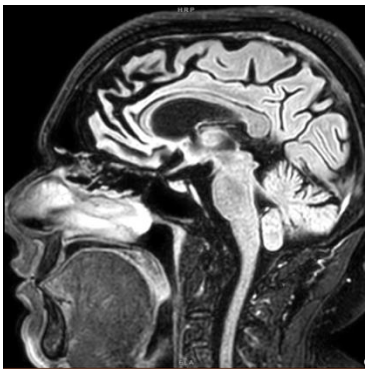
BACKGROUND

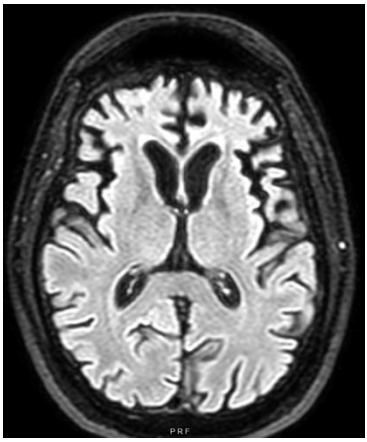
Klüver-Bucy syndrome (KBS) is a neuropsychiatric disorder due to lesions affecting **bilateral temporal lobes**. It is characterized by hyperorality, eating disorders, hypersexuality, placidity and visual agnosia. KBS can be associated to frontotemporal dementia (FTD), the second most common cause of young-onset dementia (YOD), characterized by progressive cortical atrophy involving frontal and anterior temporal lobes. It is classified into two phenotypes: behavioural variant FTD (bvFTD), presenting with early changes in personality, behaviour, and executive function, and primary progressive aphasia (PPA), defined by insidious language decline. Three PPA variants are recognized: semantic, nonfluent/agrammatic, and logopenic. Overlap syndromes involving motor neuron disease (FTD-MND) and parkinsonism have also been described, with C9ORF72 hexanucleotide repeat expansion implicated. Approximately 20% of FTD cases follow an autosomal dominant inheritance, commonly due to mutations in MAPT, GRN, or C9ORF72. Diagnosis is based on clinical criteria supported by neuroimaging, CSF biomarkers, neuropsychological testing, and genetic analysis.



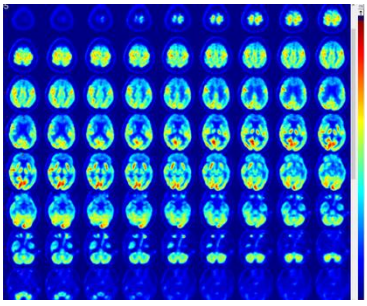
Brain MRI, FLAIR sequence, sagittal

CASE PRESENTATION

A 29-year-old man presented with a two-year history of progressive psychopathological symptoms, including depressive mood, social withdrawal, affective flattening, abulia and anhedonia. He also presented Klüver-Bucy syndrome. Medical and family history were unremarkable. Psychiatric treatment with Aripiprazole and Risperidone yielded no clinical improvement. Despite trials of Clozapine and Olanzapine during psychiatric hospitalization, symptoms worsened. Brain CT was normal, and MRI revealed **severe frontal and parietal cortical atrophy**. Neurological evaluation was prompted by further clinical deterioration. On examination, the patient was alert but poorly cooperative, hypomimic and marked psychomotor slowed. Cranial nerve function was intact. Mild rigidity was noted in the upper limbs with brisk reflexes and preserved strength. He exhibited disorientation, poor hygiene, anosognosia, disorganized speech, and socially inappropriate behaviours. Neuropsychological assessment (MMSE: 14.86/30) revealed impairments in verbal memory, attention, executive functioning, visuospatial reasoning, and constructive praxis. CSF analysis was negative for tau and onconeural antibodies. Brain FDG-PET demonstrated **hypometabolism in frontal, parietal, and temporal regions**. Amyloid PET excluded Alzheimer's pathology. A total-body CT scan ruled out paraneoplastic causes. A comprehensive genetic panel was initiated (results pending). After three months, symptoms of Klüver-Bucy syndrome worsened.



Brain MRI, FLAIR, axonal



Brain FDG-PET

CONCLUSION

Despite pending genetic results, the clinical picture fulfils criteria for **probable bvFTD**, with an **unusually early onset**. This case underlines the differential diagnosis of FTD from primary psychiatric disorders in young adults. Early neuroimaging and multidisciplinary evaluation are crucial for timely diagnosis. The presentation of Klüver-Bucy syndrome further supports anterior temporal lobe involvement, highlighting the need for heightened awareness of neurodegenerative processes in atypical psychiatric presentations.

References
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[2] J. C. Morris and B. F. Boeve, "Behavioral Variant Frontotemporal Dementia," Jun. 01, 2022, *Lippincott Williams and Wilkins*. doi: 10.1212/CON.0000000000001105.