Case Report

Fronto-temporal lobar degeneration

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Abstract

Fronto-temporal lobar degeneration is a clinically and pathologically heterogenous syndrome characterised by progressive decline in behaviour and cognitive functions associated with relatively selective atrophy of frontal and temporal lobes that characterises most, but by no means all, *Fronto-temporal dementias.*

Keywords: Fronto-temporal.

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INTRODUCTION

Fronto-temporal dementias are considered as a separate group of dementing disorders in which abnormalities of personality and behavior overshadow symptoms of memory impairment. Alternatively, the salient presentation may be a language disturbance (e.g., primary progressive aphasia) or motor abnormalities with asymmetrical apraxia and Parkinson's features (e.g., corticobasal degeneration)1) Eventually, a global dementia ensues with progression that in some instances may be more rapid than what is customary for AD.

Three distinct varieties of frontotemporal lobar degeneration has been described

- Behavioural variant: This variant is characterised by changes in behaviour and personality in associates with frontal predominant cortical degeneration.
- Semantic dementia: It is a syndrome of progressive loss of knowledge about words and objects associated with anterior temporal neuronal loss.
- Progressive nonfluent aphasia: It is characterized by effortful language output, loss of grammer and motor speech deficits in the setting of left perisylvian cortical atrophy

CASE REPORT

Sixty five years old female patient presented with chief complaints of behavioural and cognitive dysfunctions since 2yrs.No other significant history was seen. Patient was subjected to undergo MRI brain plain study with basic sequences [T1W,T2W, FLAIR images, DW images]. The MRI study revealed the following features

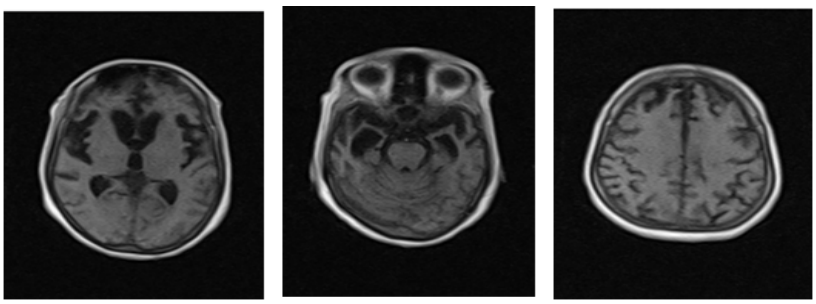

Figure 1: T1w images a) frontal b) temporal c) parietal

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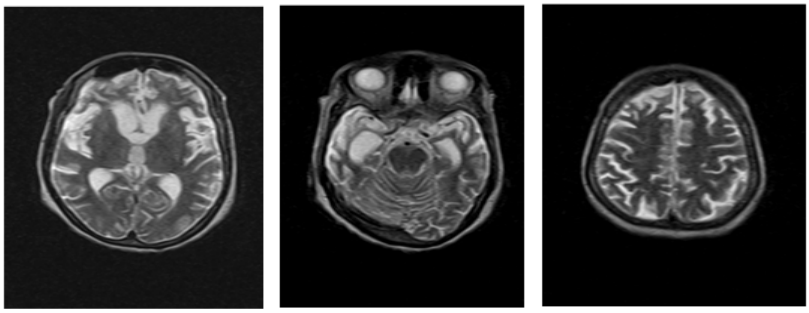

Figure 2: T2W IMAGES: d) FRONTAL e) TEMPORAL f) PARIETAL

The images reveal severe volume loss of bilateral frontal and temporal lobes reflected by dilatation of sulcul CSF spaces, basal cisterns, frontal and temporal horns of lateral ventricle. There is typically sparing of bilateral parietal and occipital lobes. Above features are suggestive of Fronto-temporal lobar degeneration.

DISCUSSION

Behavioural variant FTLD – most common variant

This syndrome is characterised by progressive decline in interpersonal and executive skills with altered emotional responsibility and emergence of variety of abnormal behaviours including apathy, disinhibition, obsessions, rituals and stereotypes.[2] It can develop indolently and early detection may depend on subtle changes of social circumstances. Median age of onset is between 45 and 65yrs of age. They were formerly called pick disease but this term is now reserved for cases with widespread presence of so called pick bodies and is based on pathological and histological features. The normal brain contains 6 isoforms of Tau proteins with either (3R-) or (4R-) microtubule binding repeats. Mutation leads to abnormal accumulations in neurons and glia. Approximately 10% are caused by mutation in microtubule associated protein Tau gene (MAPT) and 10% have mutations in the progranulin gene (GRN).

Neuroimaging

MRI:-Standard T1W images show severe volume loss(atrophy) involving the frontal and the anterior temporal lobes.[3]. The sulci of these lobes become so atrophic that they have been described as knife like appearance. There is usually sparing of bilateral parietal and occipital lobes. Altered white matter signal may be prominent usually in close proximity to areas of cortical atrophy where it reflects gliosis.[4] DWI shows elevated mean diffusivity in the superior frontal gyri, orbitofrontal gyri and anterior temporal lobes. DTI with reduced FA in the superior longitudinal fasciculus is common finding. MRS shows decreased NAA and increased MI in frontal lobes. FDG PET scans show hypoperfusion and hypometabolism in frontal and temporal lobe.[5] CT represents late stage changes suggestive of severe atrophy as described.

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