International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Case Report of Frontotemporal Dementia with Motor Neuron Disease

Dr. J. Sai Sharanya[1], Dr. G. Shirisha Laxmi[2], Dr. Ranjith Kumar[3], Dr. K. Ravinder Reddy[4]

[1]Junior Resident Department of General Medicine, Prathima Institute of Medical Sciences.

[2]Junior Resident Department of General Medicine, Prathima Institute of Medical Sciences.

[3]Consultant Neurologist, Professor of Neurology, Prathima Institute of Medical Sciences.

[4]Consultant Cardiologist, Professor and HOD of Department of General Medicine, Prathima Institute of Medical Sciences

Abstract: ***This article details a case study of a 40 - year - old male diagnosed with frontotemporal dementia FTD with motor neuron disease MND features, presenting with a complex array of symptoms including progressive slowness, incontinence, difficulty in walking, chewing, and swallowing, alongside significant muscle wasting and emotional liability. The patients diagnostic journey is explored through physical and neurological examinations, imaging studies, and other investigative procedures, which revealed multiple foci of micro hemorrhages, diffuse cerebral and cerebellar atrophy, and involvement of both upper and lower motor neurons indicative of a combined FTD - MND syndrome. The discussion highlights the diagnostic challenges posed by the overlap of symptoms with other conditions such as ALS and normal pressure hydrocephalus, underscoring the importance of comprehensive clinical assessments and longitudinal monitoring in achieving a conclusive diagnosis. Management strategies including conservative treatments and nutritional support are also addressed.***

Keywords: Frontotemporal Dementia, Motor Neuron Disease, Neurodegenerative Disorders, Diagnostic Challenges, Clinical Management

1. Introduction

Frontotemporal dementia (FTD) is the second most common cause of early - onset dementia in patients younger than 65 years.3 major clinical presentations are recognized in FTD: behavioural variant, progressive non - fluent aphasia and semantic dementia.

Motor neuron diseases (MNDs) are a group of disorders characterized by the dysfunction of either upper motor neurons (UMNs), or lower motor neurons (LMNs), or both, leading to progressive weakness, muscle atrophy with eventual paralysis.

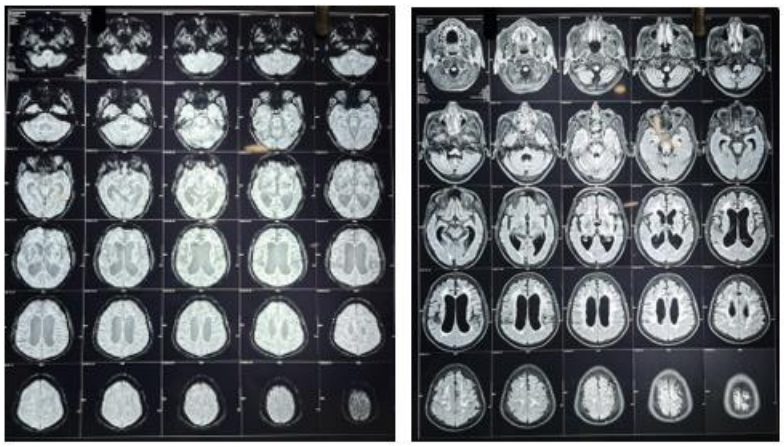
2. Case Report

A 40 yr old male presented with history of progressive slowness of activities, bowel and bladder incontinence since 5 years. H/O progressive difficulty in walking since 2yrs, difficulty in chewing and swallowing of food, drooling of saliva and regurgitation of feeds and cough since 20days.
General physical examination: the patient was conscious and had a vacant staring gaze, along with notable memory deficits and emotional liability, with stable vitals.

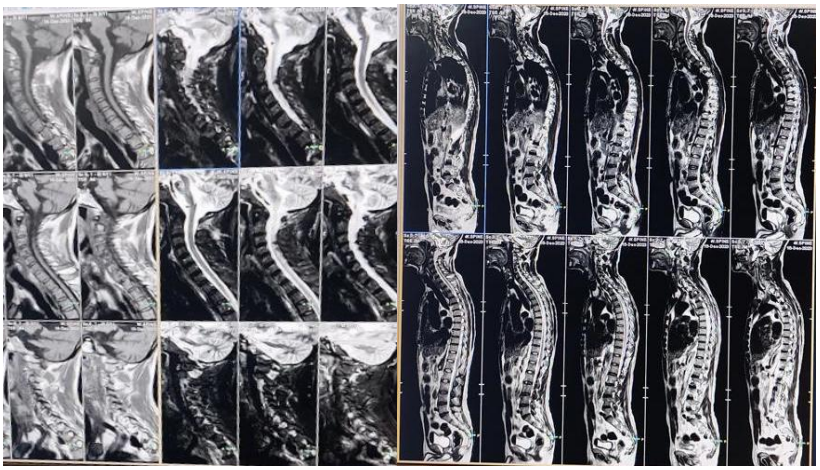
The neurological examination revealed pronounced muscle wasting predominantly in the upper limbs, contrasting with hypertonia observed in the upper limbs and hypotonia in the lower limbs. Additionally, heightened reflexes, brisk jaw jerk, and upgoing plantars were noted. Notably, primitive reflexes such as the glabellar tap and palmomental reflexes remained intact.

There were no observed sensory, cerebellar, or autonomic nervous system deficits.

Initial Workup:
MRI BRAIN showed multiple foci of blooming micro hemorrhages in b/l cerebral, cerebellar, brainstem and basal ganglia with diffuse cerebral and cerebellar atrophy, with dilated ventricles.



MRI WHOLE SPINE revealed no features suggestive of compressive myelopathy or syrinx.



ENMG STUDY: No spontaneous activity noted in B/L deltoid, biceps, FDI muscles.

NCS STUDY: Normal

OTHER INVESTIGATIONS: Sr. electrolytes, CBP and B12 levels normal, ESR 60mm/hr, normal thyroid profile and LFT.

3. Discussion

The MND associated with behavioural variant of FTD is generally similar to that seen in classic ALS, with a few exceptions. Here as there is involvement of both upper motor neurons (resulting in pyramidal signs such as spasticity and hyperreflexia) and lower motor neurons (resulting in weakness, atrophy, and wasting) along with bulbar involvement (facial or tongue weakness or dysphagia). Pseudobulbar affect, or uncontrolled bouts of laughter or crying Presence of behavioural symptoms, progressive changes in personality, Disinhibition and apathy lead us to the diagnosis of FRONTOTEMPORAL DEMENTIA WITH MND features. The patient has been managed conservatively with Ryles tube feeding, IV Edaravone, Antioxidants, riboflavin and COQ supplementation. Advised PEG tube insertion for long term feeding assistance and prevention of aspiration risk.

4. Conclusion

This case presents unique challenges and considerations in its diagnosis with progressive behavioural disturbances, gait disturbances and bowel and bladder disturbances mimicking normal pressure hydrocephalus and also ALS features like UMN and LMN findings.

Concluding a diagnosis of FTD - ALS requires thorough clinical assessments, including neurological examinations, neuropsychological testing, imaging studies, and genetic testing if warranted. Additionally, longitudinal monitoring may be necessary to track the progression of symptoms, ensuring an accurate and timely diagnosis.

References

[1] Bang J, Spina S, Miller BL. Frontotemporal dementia. Lancet 2015; 386: 1672–1682. [PMC free article] [PubMed] [Google Scholar]
[2] Kurz A, Kurz C, Ellis K, et al. What is frontotemporal dementia? Maturitas 2014; 79: 216–219. [PubMed] [Google Scholar]
[3] Bott NT, Radke A, Stephens ML, et al. Frontotemporal dementia: diagnosis, deficits and management. Neurodegener Dis Manag 2014; 4: 439–454. [PMC free article] [PubMed] [Google Scholar]

[4] World Health Organization. Dementia fact sheet, http: //www.who. int/mediacentre/factsheets/fs362/en/ (2016). Accessed January 22, 2017.
[5] Hardiman O, Al - Chalabi A, Chio A, Corr EM, Logroscino G, Robberecht W, Shaw PJ, Simmons Z, van den Berg LH. Amyotrophic lateral sclerosis. Nat Rev Dis Primers.2017 Oct 05; 3: 17071. [PubMed]
[6] Statland JM, Barohn RJ, McVey AL, Katz JS, Dimachkie MM. Patterns of Weakness, Classification of Motor Neuron Disease, and Clinical Diagnosis of Sporadic Amyotrophic Lateral Sclerosis. Neurol Clin.2015 Nov; 33 (4): 735 - 48. [PMC free article] [PubMed].