Annals of Medicine and Surgery 81 (2022) 104545

Contents lists available at ScienceDirect

Annals of Medicine and Surgery

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

Case Report

An Indonesian elderly with primary progressive aphasia and behavioral variant of frontotemporal dementia: A case report and review article

Aditya Kusumo Riswanto *, Wendy Amelia Sihombing, Yudha Haryono

Department of Neurology, Faculty of Medicine, Universitas Airlangga, Dr. Soetomo General Academic Hospital, Surabaya, Indonesia

ARTICLE INFO

Keywords:
Abnormal behavior
Dementia
Frontotemporal
Mental health

ABSTRACT

Background: Frontotemporal dementia (FTD) or Pick's disease, is the second most frequent cause of primary degenerative dementia in those between 55 and 65 years old.
Case presentation: A 57-year-old Indonesian female reported family that six months until one year prior to the presentation of her first symptoms, the patient had problems with memory, particularly short-term memory loss, with the patient unable to remember the task she was doing on time. The electroencephalogram revealed slowing background cerebral activity and diffuse slowing activity, indicating encephalopathy diffuse moderate state. CSF showed no pleocytosis and no elevated CSF Protein, but we did not perform tau level. She underwent brain magnetic resonance imaging (MRI) because of her aggression and impulsiveness. Brain MRI was notable for bilateral frontal and temporal atrophy. Incidentally, there was the leptomeningeal enhancement of the bilateral frontotemporal lobe. The patients were administered Haloperidol 0.5 mg orally twice daily, Donepezil 5 mg oral once daily, Aripiprazole 2.5 mg once daily, and Memantine 10 mg twice daily. The patient was discharged one week after admission and was started on antiviral therapy Acyclovir 800 mg 5 times a day for 14 days. The patient had shown more cooperative and less agitative.
Discussion: We report that FTD aims to help improve effective management.
Conclusion: Awareness of FTD needs to be increased even though this case is sporadic because it does not demand the possibility of this case occurring at a young age.

1. Introduction

Frontotemporal dementia (FTD) is a clinically, neuroanatomically, and pathologically heterogeneous group of neurodegenerative diseases that share a propensity to target the frontotemporal lobes of the brain [1]. From a clinical perspective, FTD usually presents as a disturbance of complex behavior, affecting pre-dominantly interpersonal conduct or language (primary progressive aphasia, PPA), memory and navigational skills and other aspects of general intellect are often well maintained initially, often present in middle life [2,3]. It was reported that 0.1–0.3 out of 1000 people experienced FTD [4,5]. This report presents a case of a middle-aged woman who was diagnosed with Dementia, Alzheimer and organic mental disorders for many years before developing and being diagnosed with FTD, behavior and primary progressive aphasia. We report based on SCARE guideline 2020 [6].

2. Case presentation

A 57-year-old Indonesian female reported that six months until one year before the presentation of her first symptoms, the patient had problems with memory, particularly short-term memory loss, with the patient unable to remember the task she was doing at a time. She was less organized in managing her finances, and the patient was found to have a car accident, drive out alone and get lost. The patient also frequently misplaced things. There were continued behavioral changes, with apathy, lack of interest and difficulty planning. She lost interest in her hobbies and spent hours sitting on the couch staring at the television or wall, and her effect became flat. She has decreased speech ability, sudden aggression, impulsiveness several times, and insomnia. The family report patient has no history of previous health problems, especially syphilis is also denied.

Her pulse, blood pressure, and respiratory rate were regular on physical examination, and no medical signs were evident. Even her

* Corresponding author. Department of Neurology, Faculty of Medicine, Universitas Airlangga, Dr. Soetomo General Academic Hospital, Jl. Mayjend Prof. Dr. Moestopo No. 6-8, Airlangga, Gubeng, Surabaya, East Java, 60286, Indonesia.
E-mail address: aditya.riswanto0622@gmail.com (A.K. Riswanto).

https://doi.org/10.1016/j.amsu.2022.104545
Received 3 July 2022; Received in revised form 22 August 2022; Accepted 27 August 2022
Available online 2 September 2022

systemic examination was regular, but she was conscious and uncooperative such as her eye-to-eye contact and attention were brief. Her last hospital visit showed poor grooming and hygiene, a flat affect, and appeared apathetic. She had no speech and not paying attention to her surroundings, and her mood was labile and regular psychomotor activity. In the clinical exam, we found severe cognitive impairment, as revealed by the neuropsychological assessment, optimistic glabellar and bilateral palm omental reflex, although these frontal release signs are not sensitive or specific for FTD. All cranial nerves were found intact, and the remainder of reflex examinations are typically standard. Blood analysis test showed BUN of 13 mg/dL, creatinine serum of mg/dL, AST of 26 U/L, ALT of 33 U/L, Hb of 13.2 g/dL, RBC of 4.31 $\times$ 10^6/μL, hematocrit of 38.5%, WBC of 15.91 $\times$ 10^3/μL, Neutrophile of 83.3% and platelet of 283,000/μL. However, urine analysis showed pH of 7.5 and leucocytes of 3+. The patient's complete blood count, renal functions, liver functions, and routine urine examinations were average. Her serum electrolytes were normal. He tested positive for IgG Cytomegaly virus (CMV) and found IgM-IgG positive on Herpes simplex virus type I.

The electroencephalogram revealed slowing background cerebral activity and diffuse slowing activity, indicating encephalopathy diffuse moderate state (Fig. 1). CSF showed no pleocytosis and no elevated CSF Protein, but we did not perform tau level. She underwent brain magnetic resonance imaging (MRI), given her aggression and impulsiveness. Brain MRI was notable for bilateral frontal and temporal atrophy. Incidentally, there was the leptomeningeal enhancement of the bilateral frontotemporal lobe (Fig. 2 & 3). The patients were administered Haloperidol 0.5 mg orally twice daily, Donepezil 5 mg oral once daily, Aripiprazole 2.5 mg once daily, and Memantine 10 mg twice daily. The patient was discharged one week after admission and was started on antiviral therapy Acyclovir 800 mg 5 times a day for 14 days. The patient had shown more cooperative and less agitative.

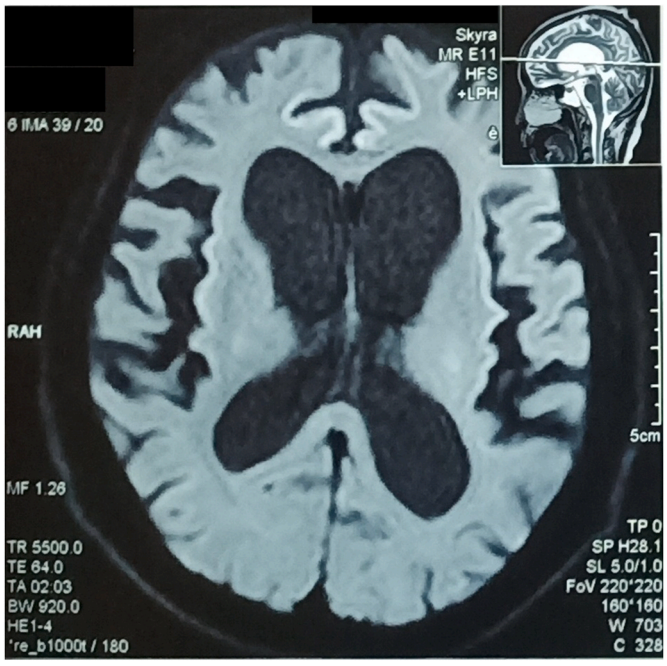

Fig. 2. Bilateral prefrontal cortex atrophy and insular cortex atrophy.

3. Discussion

There are currently no therapies approved explicitly for FTD. Thus, education and supportive management of safety and behavioral issues for patients and caregivers are essential in supporting patients with FTD [7]. Nonpharmacologic interventions should be considered the first line of treatment for patients with FTD [8]. The primary purpose of non-pharmacological interventions is to prevent disruptive behaviors, provide symptom relief, and lessen caregiver distress [9,10]. Environmental approaches aim to decrease anxiety, aggression, and irritability that arise from patients' difficulty processing information from the array of daily stimuli. These interventions include reducing noise, limiting stimulation, simplifying social parameters by limiting interaction to small groups of people, and facilitating the removal of complicated daily activities that may confuse, and therefore agitate, patients. Exercise has also been suggested to reduce behavioral symptoms [9,11].

Speech and swallowing assessments are indicated to optimize communication strategies and screen for dysphagia, which is common, particularly in patients with noneffluent agrammatic PPA. Physical therapy evaluations of gait when falls or balance problems are reported and occupational therapy assessments of home safety are also indicated. Caregivers should be referred to local FTD or dementia support groups for support and education on behavioral management strategies [7]. Non-pharmacological therapy, one of which is being developed, is occupational therapy. Occupational therapy has a role in improving functional ability and independence by using a combined approach. The role of occupational therapy in Cognitive rehabilitation uses an individual-centred perspective approach, involving not only the patient but family and people around the patient [12].

Serotonin is a selective serotonin reuptake inhibitor, an essential

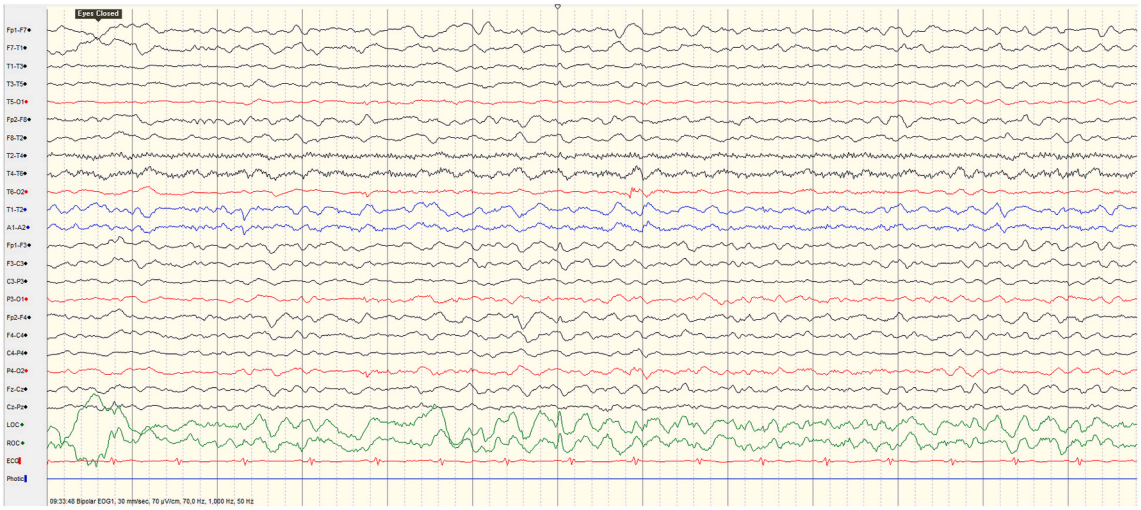

Fig. 1. The electroencephalogram: slowing background cerebral activity and diffuse slowing activity.

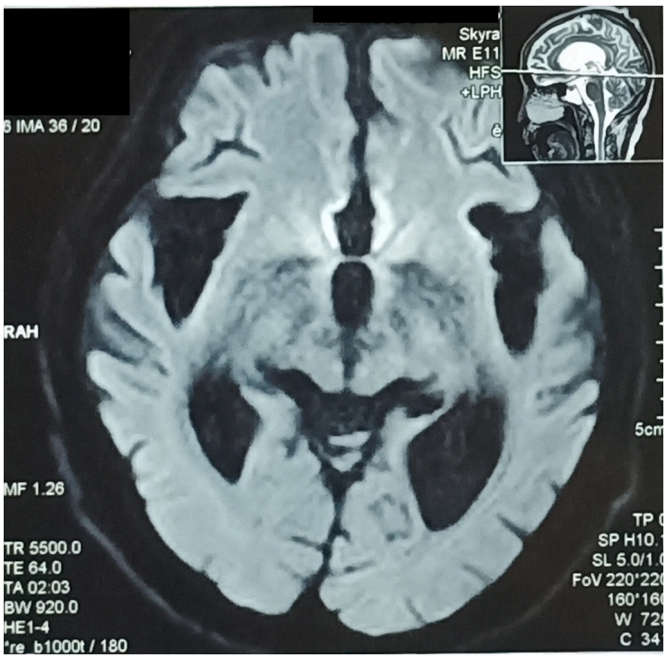

Fig. 3. Bifrontal temporal atrophy.

neurotransmitter in frontal subcortical pathways, and loss of serotonergic binding is documented in bvFTD [8,10]. Early case reports and case series studies using selective serotonin reuptake inhibitors (including fluoxetine, sertraline, and paroxetine) have some efficacy in treating compulsion, hyperorality, depression, inappropriate sexual behavior, and disinhibition of patients with bvFTD [8,12]. There is limited evidence that antidepressants such as selective serotonin reuptake inhibitors (SSRIs), such as citalopram, or trazodone, are used to improve behavioral symptoms, including impulsivity, disinhibition, agitation/irritability, or compulsive behaviors [7,9]. bvFTD patients who have been prescribed 30 mg daily dosages of citalopram demonstrated improved disinhibition symptoms and a partial restoration of serotonergic neurotransmission in dysfunctional prefrontal cortical systems [9,11]. Electroencephalography (EEG) is a device that can record electrical activity along the scalp. EEG measures the voltage fluctuations produced by ion currents in brain neurons [13].

Alternatively, treatment with trazodone at dosages of at least 300 mg/day over 12 weeks has been reported to help decrease symptoms of problematic eating, agitation, irritability, dysphoria, and depression, although adverse events were more prominent in subjects being prescribed the psychotropic [8,9]. Memantine (NMDA antagonist) and cholinesterase inhibitors have no clinically significant efficacy in treating FTD, and they potentially hasten cognitive decline or worsen behavioral symptoms [9,10]. This is likely because cholinergic deficit may not contribute toFTDe pathophysiology [10,14]. Antipsychotics and mood stabilizers are occasionally used to treat psychosis and agitation of patients with FTD, but they are not FDA-approved and could be detrimental in dementia-associated agitation due to increased risk of mortality and unwanted side effects[3]. Risperidone and olanzapine have some documented utility in treating behavioral disturbance (agitation or impulsivity) compared with other antipsychotics [8–10]. However, these medications could have side effects and increase mortality risk in patients with FTD [10,15].

Psychosis and aggression may require neuroleptic medications, although gold-standard randomized clinical trials of these agents are not available in patients with FTD. If needed, initiation at a low dose and frequent re-assessment of efficacy and need for continued use is required given black box warnings for this class of medications due to increased mortality. For patients with CBS or noneffluent agrammatic variant PPA who have memory deficits, approximately 30–40% may have underlying AD pathology, and therefore a 2–3-month trial of a cholinesterase inhibitor is warranted. A double-blind, placebo-controlled randomized trial of memantine for cognitive and behavioral symptoms of FTD showed no benefit [7]. Over the last few decades, there has been a renewed interest in focal neuromodulation as a treatment approach for neuropsychiatric conditions. The neuromodulation-based interventions discussed include Transcranial Magnetic Stimulation (TMS) and Transcranial Direct Current Stimulation (tDCS), which are non-invasive intervention therapy and Vagus Nerve Stimulation (VNS) and Deep Brain Stimulation (DBS), which are invasive interventional therapies [16,17].

Frontotemporal dementia is essentially assessment, as in our case. Neuroimaging studies are essential to provide supportive evidence for diagnosis and exclude other structural diseases. The treatment mode available for frontotemporal dementia is minimal as there are no currently FDA-approved disease-modifying treatments.

4. Conclusion

Although uncommon, clinicians must be mindful of young onset FTD. Clinicians should consider FTD when assessing patients with lack of judgment, erratic behavior, and unresponsiveness. It is essential to recognize that there is no positive anticipation or treatment accessible for FTD.

Ethical approval

Not applicable.

Sources of funding

None.

Author contribution

All authors contributed toward data analysis, drafting and revising the paper, gave final approval of the version to be published and agree to be accountable for all aspects of the work.

Registration of research studies

Name of the registry:-.
Unique Identifying number or registration ID:-.
Hyperlink to your specific registration (must be publicly accessible and will be checked):-.

Guarantor

Aditya Kusumo Riswanto is the person in charge of the publication of our manuscript.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

Aditya Kusumo Riswanto, Wendy Amelia Sihombing, and Yudha

Haryono declare that they no conflict of interest.

Acknowledgement

We would like to thank our editor, “Fis Citra Ariyanto”.

References

[1] H. Sivasathiaseelan, C.R. Marshall, J.L. Agustus, E. Benhamou, R.L. Bond, J.E. P. van Leeuwen, et al., Frontotemporal dementia: a clinical review, Semin. Neurol. 39 (2) (2019) 251–263, https://doi.org/10.1055/s-0039-1683379.

[2] J.D. Warren, J.D. Rohrer, M.N. Rossor, Clinical review. Frontotemporal dementia, BMJ (Clin. Res. ed) 347 (2013) f4827, https://doi.org/10.1136/bmj.f4827.

[3] J.P. Swatan, Y. Haryono, Are abbreviated mental test (AMT) and ascertain dementia 8 Indonesia (AD8-INA) questionnaires more superior than mini-mental state examination (MMSE) as dementia screening instrument among elderly in rural areas? AKSONA 2 (1) (2022) 6–10, https://doi.org/10.20473/aksona.v2i1.210.

[4] D.B. Hogan, N. Jetté, K.M. Fiest, J.I. Roberts, D. Pearson, E.E. Smith, et al., The prevalence and incidence of frontotemporal dementia: a systematic review, The Canadian journal of neurological sciences Le journal canadien des sciences neurologiques 43 (Suppl 1) (2016) S96–s109, https://doi.org/10.1017/cjn.2016.25.

[5] A.H. Pramudita, S. Ermawati, F. Zahira, N. Hafizah, Q.A. NP, J.P. Swatan, et al., Profile of dementia in rural area of Indonesia, EurAsia J. BioSci. 14 (1) (2020).

[6] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230, https://doi.org/10.1016/j.ijsu.2020.10.034.

[7] E.C. Finger, Frontotemporal dementias, Continuum 22 (2 Dementia) (2016) 464–489, https://doi.org/10.1212/con.0000000000000300.

[8] P.A. Ljubenkov, B.L. Miller, A clinical guide to frontotemporal dementias, Focus 14 (4) (2016) 448–464, https://doi.org/10.1176/appi.focus.20160018.

[9] J.J. Young, M. Lavakumar, D. Tampi, S. Balachandran, R.R. Tampi, Frontotemporal dementia: latest evidence and clinical implications, Ther. Adv. Psychopharmacol. 8 (1) (2018) 33–48, https://doi.org/10.1177/2045125317739818.

[10] M.N. Liu, C.I. Lau, C.P. Lin, Precision medicine for frontotemporal dementia, Front. Psychiatr. 10 (2019) 75, https://doi.org/10.3389/fpsyt.2019.00075.

[11] M. Lowrani, R. Indarwati, P. Lestari, Non-pharmacological therapy for the elderly to prevent dementia through cognitive stimulation therapy: a systematic review, Jurnal Ners 15 (1Sp) (2020) 221–229, https://doi.org/10.20473/jn.v15i1Sp.19018.

[12] D.W. Sari, A. Igarashi, M. Takaoka, R. Yamahana, M. Noguchi-Watanabe, C. Teramoto, et al., Virtual reality program to develop dementia-friendly communities in Japan, Australas. J. Ageing 39 (3) (2020) e352–e359, https://doi.org/10.1111/ajag.12797.

[13] A.F. Setiawan, A.D. Wibawa, M.H. Purnomo, W.R. Islamiyah, Monitoring Stroke Rehabilitation Re-Learning Program using EEG Parameter: a preliminary study for developing self-monitoring system for stroke rehabilitation during new normal, in: 2020 International Seminar on Application for Technology of Information and Communication (iSemantic), 2020, pp. 620–624.

[14] L.R. Garrett, T. Niccoli, Frontotemporal dementia and glucose metabolism, Front. Neurosci. 16 (2022), 812222, https://doi.org/10.3389/fnins.2022.812222.

[15] M. Castro, N. Venkateswaran, S.T. Peters, D.R. Deyle, M. Bower, M.D. Koob, et al., Case report: early-onset behavioral variant frontotemporal dementia in patient with retrotransposed full-length transcript of matrin-3 variant 5, Front. Neurol. 11 (2020), 600468, https://doi.org/10.3389/fneur.2020.600468.

[16] A. Vlaicu, M. Bustuchina Vlaicu, New neuromodulation techniques for treatment resistant depression, Int. J. Psychiatr. Clin. Pract. 24 (2) (2020) 106–115, https://doi.org/10.1080/13651501.2020.1728340.

[17] E.C. Prasetya, L. Basoeki, Neuromodulation intervention in resistant depression, Jurnal Psikiatri Surabaya 8 (2) (2020) 39–46, https://doi.org/10.20473/jps.v8i2.17136.