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Frontotemporal Dementia: About A Case Report

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Abstract **Case Report**

***Introduction*:** Frontotemporal dementia is a common form of degenerative dementia. It typically begins between the ages of 45 and 65. However, frontotemporal dementia can often be mistaken for several psychiatric clinical presentations due to behavioral, personality, and language disorders that can be observed in various psychiatric pathology. ***Case Report*:** We report the case of a 61-year-old patient, who presented with behavioral disturbances, neglect of personal hygiene, reduced speech and a progressive onset of personality change over a three-year period, contrasting sharply with his previous state. A cerebral CT scan revealed symmetrical focal atrophy of the frontal and temporal lobes. Family members were informed about the nature and prognosis of the disease. The patient showed partial improvement of symptoms with Quetiapine 300mg/day. ***Conclusion*:** The diagnosis of frontotemporal dementia relies on a good knowledge of the diagnostic approach and the course of the disease, and on neuroimaging. The challenge is to identify targeted therapies to optimize prognosis.

Keywords: Frontotemporal Dementia, Middle Ages, Behavioral Disorders, Personality Changes, Brain Imaging.

INTRODUCTION

Frontotemporal dementia is the most common type of primary degenerative dementia after Alzheimer's disease [1], with an estimated prevalence of 2.6% [2]. It is the second most common cause of dementia in young people, occurring before the age of 65 [3].

Most frontotemporal dementias manifest themselves clinically as one of three variants: the behavioral variant, the most common, or one of two language variants (primary progressive aphasia of the asemantic type or primary progressive apragmatic non-fluent aphasia) [3]. However, unlike Alzheimer's disease, visual-spatial skills are generally preserved [4].

Nevertheless, frontotemporal dementia is a highly diverse group of disorders characterized by relatively selective degeneration of the frontal and/or temporal lobes [3].

Frontotemporal dementia is a focal form of presenial dementia, that remains poorly understood [1]. Diagnosis remains difficult, especially as the insidious onset of this pathology frequently evokes psychiatric pathologies. Nonetheless, diagnostic criteria have been developed by Lund and Manchester [5], to help with the diagnosis of frontotemporal dementia.

CASE REPORT

Mr B.N, aged 61, divorced, father of one daughter, is currently unemployed, hospitalized for alcohol withdrawal treatment with maintenance of abstinence. His brother reports that the onset of the illness seems to date back three years, marked by discreet but progressive personality changes in Mr. B.N. He became isolated, withdrawn, communicating little, neglecting his corporal care. He also began to show little interest in his hobbies and obligations. He no longer cared about his family members, and spent his days alone at home. Subsequently, he resumed drinking alcohol, which he had weaned off several years earlier, to remedy his sleep problems. As a result of these behavioral changes, the patient was obliged to give up his job (as a company shopkeeper), which he had held for several years. During the past year, he began experiencing memory problems, particularly with short-term memory loss. He often forgot the tasks he was performing and frequently misplaced his belongings. Nevertheless, he was able to recognize his family members and was able to go out on his own without getting lost. He has no medical or surgical history.

The psychiatric interview found a calm patient, well oriented in time and space, his attention distracted, his personal hygiene was neglected, and his speech was

characterized by brief answers. There were no apparent delusions or perceptual disturbances. His mood was neither sad nor euphoric, and his affect was concordant. His insight was negative, and his appetite and sleep were diminished. The Mini Mental State Examination score was 22/30 on neuropsychiatric examination.

Blood tests (hematology, biochemistry, ionogram, thyroid hormones, B12, folates) came back normal. Viral serologies (syphilis, HIV, hepatitis B and C) were negative. Electrocardiogram was normal: regular sinus rhythm, no conduction or repolarization disorders. A cerebral CT scan was performed, which revealed symmetrical focal atrophy of the frontal and temporal lobes.

The diagnosis of frontotemporal dementia comorbid with an alcohol use disorder was made. The announcement of the diagnosis enabled the family to better understand and then accept the patient's symptomatology.

During his hospital stay, the patient preferred to stay alone in his room, spoke very little, frequently misplaced his belongings, and exhibited slowed behavior.

Quetiapine was administered to the patient to manage his behavioral problems at low doses, gradually increasing to 300 mg/day. His condition improved slightly, his sleep was reportedly restored, he began to take care of himself, and he began to talk and participate in ward activities with the other patients. However, his behavioural and personality issues persisted.

With regard to his alcohol use disorder, the patient benefited from psycho-education and motivational therapy, with a focus on relapse prevention (identification of relapse factors and implementation of valid alternatives/choice and evaluation of a solution, work on automatic thoughts, social skills training, problem-solving, relaxation, etc.).

DISCUSSION

Frontotemporal dementia is among the most common causes of early-onset dementia [6]. Onset of the disease occurs at an average age of 56 years [7], with a sex ratio of 1 [3]. Depending on the patient's age, frontotemporal dementia may present neurologically similar to Parkinson's or Alzheimer's disease [8-10]. Our patient developed frontotemporal dementia at the age of 58.

The positive and etiological diagnosis of frontotemporal dementia remains a challenge. Recent studies suggest that genetics play an important role in the etiology of the disease [3-11]. A family history is found in 20-50% of cases [3], with the autosomal dominant mode (chromosome 17) being the most frequent [7].

Patients with frontotemporal dementia present a heterogeneous clinical picture that may include behavioral, cognitive and motor manifestations [6]. Our patient exhibited progressive changes in personality, behavioral disturbances, neglect of personal hygiene, loss of social awareness, loss of empathy and progressive language decline. As a result of these behavioral changes, the patient was obliged to stop work. Our patient's clinical manifestations met the Lund and Manchester criteria [5].

The diagnosis of frontotemporal dementia is based primarily on a thorough clinical assessment, and on the course of the disease [3]. Radiological investigations are also important in making the diagnosis and ruling out other structural diseases. Our patient presented with frontal and temporal lobe atrophy on cerebral CT.

Post-mortem histopathological analysis is the only examination capable of formally confirming the diagnosis of frontotemporal dementia [3]. Macroscopic examination of the brain of a frontotemporal dementia patient generally shows symmetrical focal atrophy of the frontal and temporal lobes [3]. Nonetheless, microscopic examination of the cerebral cortex in most cases shows microvacuolation and neuronal loss. When staining neuronal deposits, abnormal aggregates of Tau protein (46%) and positive ubiquitin inclusions (29%) were found [11].

Therapeutically, there is no curative or stabilizing treatment for frontotemporal dementia. The drugs used are purely symptomatic (behavioral disorders) [12], with selective serotonin reuptake inhibitors being recommended for frontotemporal dementia [13], due to a reduction in serotonin 5HT2A receptors revealed in studies [3-6].

According to the literature, hypersensitivity to neuroleptics exists in frontotemporal dementia patients [14], so they should be prescribed with caution. In our patient, quetiapine was administered to manage his behavioral disturbances at low doses, with progressive dose escalation up to 300 mg/day. His condition improved slightly. However, his behavioral and personality disorders persisted. Similarly, Kuğu *et al*., [6], reported partial improvement in behavioral symptoms in a patient with insidious onset frontotemporal dementia, presenting with behavioral disturbances and personality modification.

Acetylcholinesterase inhibitors (tacrine, donepezil, rivastigmine), and their possible combination with N-methyl-D-aspartate receptor antagonists (memantine), have shown some efficacy in the management of frontotemporal dementia, but further studies are still needed [15].

CONCLUSION

Frontotemporal dementia can be easily mistaken for various psychiatric conditions, highlighting the importance of comprehensive screening, understanding symptoms, and multidisciplinary management.

The benefits of diagnosis include improved therapeutic management and a more reliable prognostic approach, leading to a better therapeutic alliance.

REFERENCES

1. Snowden, J. S., Neary, D., & Mann, D.M. (2002). Frontotemporal dementia. *The British Journal of Psychiatry*, *180*, 140-143.
2. Ikejima, C., Yasuno, F., Mizukami, K., Sasaki, M., Tanimukai, S., & Asada, T. (2009). Prevalence and causes of early-onset dementia in Japan: a population-based study. *Stroke*, *40*(8), 2709-2714.
3. Lebouvier, T., Bertoux, M., Leroy, M., Lebert, F., Deramecourt, V., & Pasquier, F. (2019). Diagnostic positif et étiologique des démences frontotemporales. *Pratique Neurologique-FMC*, *10*(2), 101-111.
4. Ting, H. S., Yashodhara, B. M., Dar, B. A., Ali, U., Yousuf, M., & Abas, A. L. (2013). A case of frontotemporal dementia. *International Journal of Case Reports and Images (IJCRI)*, *4*(3), 175-179.
5. Englund, B., Brun, A., Gustafson, L., Passant, U., Mann, D., Neary, D., & Snowden, J. S. (1994). Clinical and neuropathological criteria for frontotemporal dementia. *J Neurol Neurosurg Psychiatry*, *57*(4), 416-8.
6. Kuğu, N., Doğan, O., Kavakcı, Ö., & Terlemez, İ. (2010). Frontotemporal dementia: a case presentation. *Dusunen Adam Journal of Psychiatry and Neurological Sciences*, *23*(4), 293.
7. Chow, T. W., Miller, B. L., Hayashi, V. N., & Geschwind, D. H. (1999). Inheritance of frontotemporal dementia. *Archives of neurology*, *56*(7), 817-822.
8. Rosen, H. J., Hartikainen, K. M., Jagust, W., Kramer, J. H., Reed, B. R., Cummings, J. L., ... & Miller, B. L. (2002). Utility of clinical criteria in differentiating frontotemporal lobar degeneration (FTLD) from AD. *Neurology*, *58*(11), 1608-1615.
9. Gigi, A., Pirrotta, R., Kelley-Puskas, M., Lazignac, C., & Damsa, C. (2006). Troubles du comportement aux urgences, ou démence fronto-temporale? Un défi pour les psychiatres: Behavior disturbances in emergency psychiatry or fronto-temporal dementia diagnosis? A challenge for psychiatrists. *L'Encéphale*, *32*(5), 775-780.
10. Wider, C., & Wszolek, Z. K. (2008). Etiology and pathophysiology of frontotemporal dementia, Parkinson disease and Alzheimer disease: lessons from genetic studies. *Neurodegenerative Diseases*, *5*(3-4), 122-125.
11. Forman, M. S., Farmer, J., Johnson, J. K., Clark, C. M., Arnold, S. E., Coslett, H. B., ... & Grossman, M. (2006). Frontotemporal dementia: clinicopathological correlations. *Annals of Neurology: Official Journal of the American Neurological Association and the Child Neurology Society*, *59*(6), 952-962.
12. Lebert, F., Stekke, W., Hasenbroekx, C., & Pasquier, F. (2004). Frontotemporal dementia: a randomised, controlled trial with trazodone. *Dementia and geriatric cognitive disorders*, *17*(4), 355-359.
13. Chauvire, V., Even, C., Thuile, J., Rouillon, F., & Guelfi, J. D. (2007). La démence frontotemporale: revue de la littérature. *L'Encéphale*, *33*(6), 933-940.
14. Lebert, F. (2008). Pasquier F. Symptômes comportementaux et psychologiques lors de démence. *EMC Neurologie Paris: Elsevier*, 17-057.
15. Moretti, R., Torre, P., Antonello, R. M., Cattaruzza, T., Cazzato, G., & Bava, A. (2004). Rivastigmine in frontotemporal dementia: an open-label study. *Drugs & aging*, *21*, 931-937.