

## CASE REPORT

# BEHAVIOURAL VARIANT FRONTOTEMPORAL DEMENTIA: A CASE REPORT

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## Abstract

**Objective:** This case report highlights the challenges encountered in arriving at the diagnosis of a case with 'behavioural variant Frontotemporal Dementia'. **Methods:** We report a case of 'behavioural variant Frontotemporal Dementia' diagnosed in a 49 year old Chinese lady. **Results:** This patient was misdiagnosed as Obsessive Compulsive Disorder (OCD), Parkinson Disease and Psychotic Disorder Not Otherwise Specified (NOS), deemed not responding to medications which was later complicated with Neuroleptic malignant syndrome (NMS). Her diagnosis was revised only after 2 years, after which her behavioural problems stabilised, bringing some relief to her family's distress. **Conclusion:** Though not rare, 'behavioural variant Frontotemporal dementia' is often misdiagnosed and patients and their families suffer unnecessary suffering before the condition is finally diagnosed. *ASEAN Journal of Psychiatry, Vol. 14 (1): January – June 2013: XX XX.*

**Keywords:** Behavioural Variant, Frontotemporal, Dementia, Diagnosis

## Introduction

Frontotemporal dementias (FTD) are a group of clinical neurodegenerative syndromes characterised by prominent changes in behaviour, language and frontal functions [1-3]. Unlike dementia of the Alzheimer's type, the onset of FTD occurs earlier, often before the age of 65 years and memory is relatively preserved until late. The prevalence of FTD is estimated to be 3-15 per 100000 [4,5], with 20-40% being familial [3]. On brain imaging, there is focal atrophy over the frontal and anterior temporal regions of the brain [3]. Three distinctive variants have been described: behavioural variant FTD (bvFTD), semantic dementia (SD) and progressive non-fluent aphasia (PNA); of which bvFTD is the commonest. Though not rare, diagnosis is often delayed [6]. Rosnes et al (2008) found that it took an average of 59.2

months and 49.5 months to arrive at a clinical diagnosis of Frontotemporal Dementia in Norway and Sweden respectively [6]. This is likely due to the heterogenous presentations of FTD which can mimic psychiatric disorders [7]. This is a case report highlighting the challenges in arriving at a diagnosis of a case of bvFTD.

## Case Report

T, a 49 year old Chinese seamstress was hospitalised in May 2011 for a 2-year history of progressive 'restlessness and disorganized behaviour' which did not improve on high dose antipsychotic medication. She was initially brought to the neurology clinic a year ago for prominent changes in behaviour associated with some forgetfulness. The neighbours complained that T kept returning to the shops insisting on getting the 'correct change'. She barged into

her neighbours' homes uninvited 'to chat', causing considerable distress when she failed to take hints to leave. She had repetitive behaviour such as taking the laundry in and then putting them out again, rearranging the furniture at home and repeatedly showering. She also hoarded newspapers and bits of string. She was well apart from 'iron deficiency anaemia' secondary to fibroid-associated menorrhagia. T did not have any family history of dementia or neurological diseases.

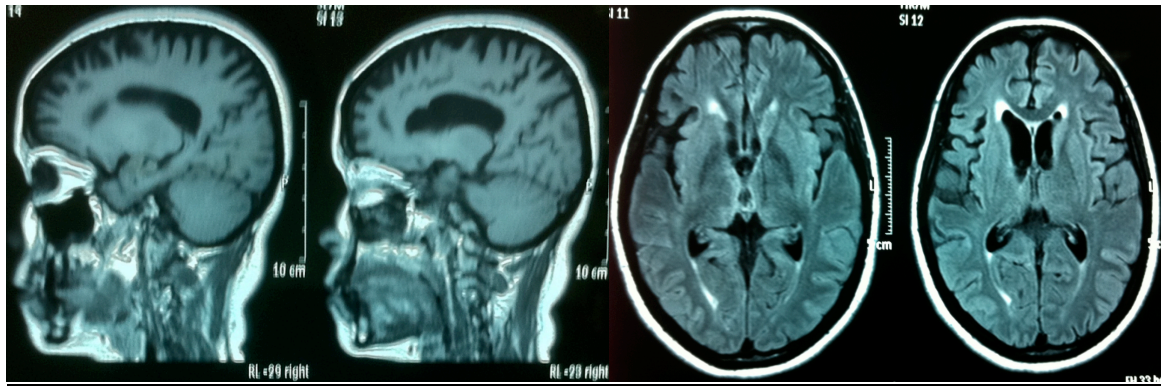
At the time, she was not found to have any physical problems or any neurological deficits. She scored 25/30 in the Mini-Mental State Examination (MMSE) (losing 4 marks to serial 7s and 1 to 5-minute recall), 16/18 in the Frontal Assessment Battery and 13/15 in the Geriatric Depression Scale. Her blood investigations did not reveal any abnormal findings. Lumbar puncture results were normal. MRI Brain was reported as generalised cerebral atrophy.

Within the same year (2010), she lost her job due to the poor quality of her sewing. Premorbidly from a friendly person who was gentle, easy-going and mild mannered; she had become someone who complained a lot, acted on impulse without regarding the opinion or feelings of others and hoarded things. She was not able to perform her usual house chores. She kept buying food even though some had rotted in the fridge. She spent hours recalculating her telephone credits but could not get them right. Her day was spent either collecting bits of paper or string which she hoarded as a compulsion.

She was referred to the psychiatrist and prescribed with Sertraline 100mg OD for

Obsessive Compulsive Disorder. Albeit a lot quieter, she did not improve. Her family had to instruct her on when to shower or eat. On the other hand, she displayed hyperorality - she gobbled up whatever food or drinks on sight and would not stop unless her family stopped her. Her behaviour caused considerable embarrassment to her family as she would impulsively take food or drinks of other patrons when they brought her out for dinner. She displayed utilization behaviour; her continually tipping a cup into her mouth even when the cup was empty. She was not able to sustain any meaningful conversation except to say repetitive words to indicate her needs.

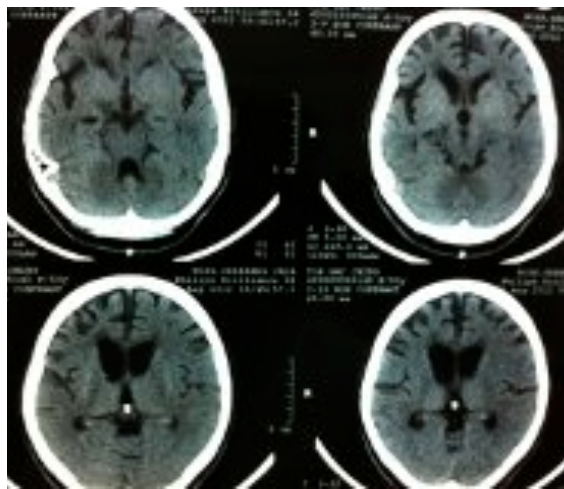
Her diagnosis was revised to 'Psychotic Disorder NOS'. She was treated with Risperidone 3 mg OD but she developed secondary parkinsonism. In between, another doctor started her on generic T. Madopar for presumed Parkinson Disease. Her medication was switched to generic T. Aripiprazole and T. Madopar was stopped in 2011. However, she continued to deteriorate, becoming more impulsive and restless despite maximal dose of Aripiprazole. She showered almost every hour, drank water repetitively and hoarded objects. She had no regard for the frustration of her family. Her affect was blunted. She did not otherwise have any psychotic or mood symptoms. She scored 13/30 on Montreal Cognitive Assessment, and 25/30 in the MMSE. A repeat MRI Brain in 2011 (Figure 1) showed further atrophy of the brain. A provisional diagnosis of Frontotemporal Dementia was made. Aripiprazole was tapered off and T was started on Fluvoxamine 50mg OD.



**Figure 1. MRI Brain of T Images July 2011: reported as generalised atrophy**

Unfortunately, T developed fever, generalised rigidity and markedly elevated serum creatinine kinase 3 days later. Her other laboratory investigations were normal. She was treated for Neuroleptic Malignant Syndrome. She was later restarted on T. Fluvoxamine; and gradually improved; being less impulsive and able to follow instructions. She was referred to the clinical psychologist for behavioural management. Her family was given psycho-education and empowered on how to manage her behavioural symptoms at home. T was finally discharged home with T. Fluvoxamine 25 mg

ON, T. Memantine 5 mg OD and T. Clonazepam 0.25mg PRN. On her follow-up visit, the family reported that she still had repetitive behaviour, but was no longer as impulsive and able to follow instructions. Her family was relieved that they had an answer and were now able to cope with her. Her son even found her a job as a sweeper (closely supervised by her husband) at a nearby apartment block. She is still working now and stable on T. Fluvoxamine 100mg BD. Her CT Brain a year later (Figure 2) revealed preferential frontotemporal atrophy.



**Figure 2. CT Brain Images of T August 2012 showing preferential temporoparietal atrophy**

## Discussion

For T, her diagnosis was reached at after a period of slightly more than 2 years. She fulfilled the International Consensus Criteria for

bvFTD [8] – she had progressive deterioration of her behaviour and cognition; early behavioural disinhibition, early loss of empathy, early repetitive or compulsive behaviour, hyperorality, objective evidence of executive deficits with

relative sparing of memory and visuospatial functions; exhibited significant functional decline and had imaging results consistent with bvFTD. Although her diagnosis was reached earlier compared to the Norwegian and Swedish cohort, she and her family underwent significant distress during the 2 years of uncertainty. T visited at least 4 different doctors, was misdiagnosed with Obsessive Compulsive Disorder, Parkinson Disease and Psychotic Disorder NOS; treated inappropriately with high doses of neuroleptics and later antiparkinsonian medication. She also unfortunately developed Neuroleptic Malignant Syndrome, a life threatening condition in the process.

Why does it take so long to come to a diagnosis of Frontotemporal Dementia? Firstly, the disease may mimic primary psychiatric disorders, misleading doctors into treating patients as such [7]. Secondly, in early bvFTD, changes are often subtle and may not be picked up by insensitive, conventional brain imaging[8]. For T, her MRI was initially reported as generalised atrophy, and only after 2 years did her brain CT began to show the classical frontotemporal preferential atrophy. Thirdly, FTD is a neurodegenerative disease without cure; hence the hesitancy on the part of the attending doctor to proclaim such a 'damning' diagnosis to the patient. Fourthly, although there are existing diagnostic criteria for the diagnosis of FTD such as the Lund-Manchester Criteria and Neary's Consensus Criteria, the former criteria lacked precision while latter criteria was relatively insensitive with ambiguous behavioural descriptors and rigid core criteria [8, 9]. While it is now possible to say that Madam T fulfilled all the criteria, at the time when her symptoms started to emerge, such a conclusion would not have been so clear-cut. Also, the previous 2 criteria catered to the all types of FTD. It is only of recent that the International Consensus Criteria for behavioural variant FTD have been developed [8].

Although there is no cure for FTD at present, symptomatic medication such as SSRIs for the impulsivity and repetitive behaviour; creating activities tailored to the needs of the patient and both educating and empowering the family has dramatically turned their suffering and

uncertainty into relief and some degree of satisfaction that they are able to help their loved one. T had tremendous family support and this has resulted in T's improved quality of life. Instead of being locked at home or restrained in a facility for her impulsivity, she is now earn some income as a sweeper and help with simple chores in the kitchen under the supervision of her devoted husband, while retaining some degree of dignity in her life. Perhaps we should all learn from this case - on how to cope and make the most of life despite the 'damning' nature and course of the disease.

### **Conclusion**

The early symptoms of bvFrontotemporal dementia may resemble those of primary psychiatric disorders and bvFTD is often misdiagnosed. As such, one needs to have a high index of suspicion and greater awareness about this condition. A meticulous and comprehensive review of the available evidence, with frequent communication with the patient's family is essential to come to the diagnosis and prevent unnecessary delay and suffering.

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