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Research Article

Behavioral symptom profile of Parkinson disease dementia with difference with behavioral variant of FTD- A primer study

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ABSTRACT

Patients of behavioral variant of frontotemporal dementia (bvFTD) exhibit prominent behavior abnormalities along with executive dysfunction. Whereas executive dysfunction is also common in subcortical dementias because of disturbance in fronto-subcortical connections but behavior symptoms are less frequent. Objectives: To determine the frequencies of behavior symptomatology of patients with subcortical Parkinson's disease dementia (PDD) and frontal cortical dementia (bvFTD) and to find out the discriminating value of behavioral characteristics. Methods: A total 20 bvFTD and 20 Parkinson's disease dementia (PDD) were recruited. Behavior symptoms were noted as questionnaire prepared by Bathgate et al. (2000) Results: Whereas eating and vegetative behaviors were more frequent in bvFTD (85% vs 45%, p=0.018), the neuropsychiatric behavior were more common in PDD group 80.0% vs 50.0%, p=0.095). Although didn't achieve statistical significance, other symptoms found to be discriminating in two condition are (i) sensory behavior (p=0.056), (ii) cognitive mediated behavior (p=0.794), (iii) affect-social behavior (p=0.162), and (iv) compulsion-ritual behavior (p=0.33). The behavior related to environmental dependency were not found prominent in either groups (p =0.155). Conclusions: Behavioral abnormalities are more common in bvFTD group with prominent abnormality in eating & vegetative, sensory, cognitive mediated, affect-social conduct, compulsion-ritual behaviors. The PDD group showed marked neuropsychiatry behavioral abnormalities

1. Introduction

Frontal lobes play a critical role in human behavior. Some of the most striking neurobehavioral syndromes are coupled with frontal lobe dysfunction. Classical five frontal-subcortical circuits help us to understand wide range of behavioral changes that accompany neurodegenerative disorders.[1,2]

The frontal lobe behavioral dysfunction can occur in both cortical e.g. FTD, AD and subcortical dementia e.g. HD, PDD and VD.[3] Among degenerative neurological disorders bvFTD is presented with prototype of behavioral features. The characteristics of bvFTD are profound alterations in personality and social conduct. These include decline in social conduct, emotional blunting, and loss of insight, decline in hygiene, mental inflexibility, distractibility, hyperorality and dietary changes, preservative and stereotyped and utilization behavior. But there is also petite knowledge as regards diagnostic specificity. These features may mislead initially diagnosis of bipolar disorder or schizophrenia.[4] In PDD the cognitive deficits involves executive, visuospatial, attention and memory function. Behavioral features are also common in this subcortical dementia. Hallucinations and delusion occurs in frequency of 45-65% in PDD.[5] Beside this major depression also seen but has same prevalence as in other degenerative dementia.[3]

Behavioral features specified in consensus criteria of FTD and PDD is currently covering a spectrum of behavioral alteration. e.g. the symptom of dietary change as overeating, food fads, indiscriminate eating, preference

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for sweet and for savory foods. There is currently little data available regarding the degree of consistency and precise form of behavioral changes within an FTD and other dementia. Although comparative studies on behaviors in FTD and other dementias was done, but these not designed to address the range of usual behavior which clinically helpful in diagnosis of FTD and PDD. The respective emphasis on cognitive and neurological data in the diagnosis of these two common forms of dementia might lead to recognized behaviors which are clinically differential.[6]

The purpose of the study was to explore the nature of behavior changes in bv FTD and PDD. We also looked for discriminating value of behavioral characteristics in two conditions.

2. Material & Methods

This is descriptive study. Subjects enrolled from Cognitive Clinic and subjected to a detail history, neurological examination and cognitive functions assessment. Objective frontal lobe testing was also done in form of FAB score.[7,8] Battery of investigations including complete Haemogram, blood sugar, Blood urea, Serum creatinine, LFT, Lipid profile, thyroid function test, vitamin B12 estimation and with Neuroimaging. Patients in whom the diagnosis is equivocal were excluded. Consecutive 20 patients of bvFTD and 20 patients of PDD were included. The frontal lobe behavior assessed employing a set of questionnaire by Bathgate et al. (2002).[9] We divided various behavior in seven categories e.g. affect and social, sensory, eating-vegetative, repetitive-compulsion-ritual, environmental dependency, cognitive mediated behaviors and behaviors related to psychosis. We used the Bengali translated and validated version. It was emphasized that a 'symptom' should reveal a remarkable change from the patient's premorbid condition and not a long-standing trait. Responses for each question were recorded as absence or presence of symptoms. Analysis performed with help of Excel software and *SPSS- Win* statistical packages version 17.

3. Results

Socio demographic features: The mean age of participants in PDD group is 62.2 years and 56.6 years in bvFTD group. The mean duration of illness at presentation was 3.85 years in bvFTD group and 6.7 years in PDD group. bvFTD group was found with relatively poor cognitive side (MMSE= 9.05 $\pm$10.37) and CDR= 1.09$\pm$37) while PDD has good base line cognition (MMSE= 20.60$\pm$ 5.63) and CDR= 0.89$\pm$0.35). On opposite side FAB score of bvFTD group was 1.95$\pm$ 2.43) and of PDD 8.25$\pm$6.02).

Affect and social behavior abnormality more evidently observed in bvFTD. Apathy and loss of em pathy more reported in FTD than PDD(95% v/s 30%). Loss of embracement was present in 13(65.0%) of bvFTD and 3(15.0%) of PDD. bvFTD patient are more likely to exhibit disinhibited behaviors than PDD. (19 (95.0%) v/s 7(35.0%) with *p* value of 0.0001**.** On comparing sensory behaviors, inappropriate response to painful stimuli in 14(70.0%) of the bv FTD group but was found only in 1 PDD (p value =0.0001). The loss of discrimination seen in 17(85.0%) of bvFTD patients v/s 4(20.0%) of PDD patients with p value of 0.0001. Wandering and pacing were reported more frequently in FTD with *p* value- 0.0014. Repetitive behaviors were common in the bvFTD group. Repetitive motor and verbal stereotypes were reported in 1/3 of the bvFTD patients. Among various environments dependency related behaviors significant association observed for mutism in bvFTD.(95% v/s 35 % with *p* value of 0.0001)Psychosis related features are present more profoundly in PDD group. Delusions noted in 9(45%) of PDD patients. Visual ha llucinations are observed in 14 (70.0%) PDD and 4(20%) bvFTD patients with *p* value of 0.0014. There were no differences between the groups in the tendency to exaggerated emotion, loss of insight, aggressiveness and irritability, altered preference for savory or sweet, repeated behaviors, behaviors related to environmental dependency. A loss of sense of smell, change in response to heat and cold sensation, and hyper oral behavioral were present in both groups but numerically more frequent in FTD.

4. Discussion

Behavior changes are the dominant symptom in bvFTD. A number of social and affect related behavior symptoms show ed very high sensitivity to FTD noted in least 50% of cases. The most sensitive feature, being present in virtually all patients are as follows: apathy and lack of empathy, selfishness, disinhibition, neglect of personal hygiene, loss of awareness for pain, loss of discrimination, overeating, wandering-pacing, fails to recognize objects, mutism, delusion- hallucination. Our result matches with previous done studies.

We observed a pathy and lack of empathy has statistically significant association with bvFTD. Similar results observed for symptoms in previous studies in range of 49 to 87%.[9–11] Three past studies found that 5-12% Parkinson patients showed apathy while 28-30% were both apathetic and depressed.[12–15] while study by Pai-Chiu et al showed 55.2 % PDD has apathy and 52.8% has depression.[15] The presence of disinhibited behavior symptoms significantly favors diagnosis of FTD compared to PDD. In bvFTD group result was 70-79 % for disinhibited behavior in various studies which matches our results.[9,11,16] Contrary to our results study Isabelle Le Ber et al. found disinhibited behavior in frequency of 10-29 %.[10] A Pai- Chiu et al[15] group observed[16] disinhibited behavior in 8% PDD while 24% by Yoon Sang et al.[17] Exaggerated emotion,

Table 1: Demographical and cognitive characteristic of study populations

	bvFTD	PDD	P value
Number	20	20	-
Age	56.60 ± 10.26	62.25± 7.26	0.0516
Duration of illness	3.85±1.56	6.70±3.07	0.0007
Education	6.35±5.16	7.35±4.93	0.5349
BMSE	9.05± 10.37	20.60± 5.63	0.0001
FAB score	1.95± 2.43	8.25 ± 6.02	0.0001
CDR	1.09±0.37	0.89±0.35	-

Table 2: Frequency of major behavioral categories in both groups

	bvFTD	PDD	*P value*
Affect & social	20 (100)	16(80%)	0.162
Sensory	13 (65%)	6 (30%)	0.056
Eating & vegetative	17 (85%)	9(45%)	0.018
Environmental dependency	8 (40%)	3(15%)	0.155
Cognitive mediated	14(70%)	12(60%)	0.794
Neuropsychiatry behaviors	10(50%)	16(80%)	0.095
Compulsion & ritual	10(50%)	6 (30%)	0.333

aggressiveness, irritability was present in 55% of bvFTD and 25% PDD. Past studies on FTD noted such behavior between 13-88 %.[18] Similar behavioral noted in 25%-29 % of PDD by others.[13,15]

One interesting result of this study was about insight. Insight was present equally in both groups that are around 80% in each. In past studies 23%-100 % FTD patients reported without any insight.[9,18] Aarsland et al observed PDD 30% patients lost insight about disease.[13]

Abnormal sensory responses were reporte d numerically most often in bvFTD. Loss of pain response has statistically significant for bvFTD. An inappropriate response to stimulus found in 33-41 % in bv-FTD while same studies showed abnormal hot cold responses in 7-11%.[7,19] Abnormal pain and temperature perception including allodynia and hyperalgesia found in 40-46 % PDD patients.[20,21]

Hyposmia was numerically more frequent in FTD 15(75.0%) compare to 12(60.0%) in PDD group. Bathgate et al reported 38 % patients suffered from hyposmia.[9] PD has hyposmia as a prominent non motor symptom, occurring in about 80%-90% of PD patients.[20,21] So despite being more frequent than in PD, olfactory dysfunction has been reported in FTD as well.

Abnormal eating behaviors were reported in 85% bvFTD and 45% of PDD patients. Some eating behavior are more common in bv-FTD. Bathgate et al observed these eating behavior in range of 13-50 %. Contrary to our study they observed sweet tongue in 50 % of patients.[9] But our results are close to results o f Ikeda M and Tanabe H. They found food faddism in 17 % and sweet tongue in 60 %, slat carving in 30 % patients. Other eating behavioral was present in 30-60 % of their study.[22] Such behavior are uncommon in PDD. In various studies, it observed that PDD patient loss weight (20-30 %) related change of food habits, bradykinesia, altered perception of taste and smell or effect of medicines.[15,23,24] There two behavior which achieved statistically significant are loss of discriminations and overeating. The loss of discrimination is seen in 85.0% and overeating observed in 40.0% of bv-FTD. Ikeda M and Tanabe H noted loss of discriminat ion in 60% and overeating in 48%.[22] Bathgate et al also found similar results e.g. loss of discrimination in 48% and overeating in 70 %.[9] Hyper orality was present in 5 bvFTD and 1 PDD patient. In bvFTD hyperorality observed up to 9-20 % in various studies.[9,18,22,25] This primitive behavior not observed in PDD in general.

Wandering and pacing were reported more frequently in bv FTD. In past this behavior noted in range of 58-83%.[9,19] in PDD such behavior not reported commonly. We observed 16(65.0%) of bvFTD patients said about altered sexual behavior predominently hypo sexuality. Similar observation made by Bath gate et al as hyposexuality in 58% while hypersexuality in 19 %.[9] In PDD we observed 7(35.0%) with such symptoms. In past studies hypersexuality in PD noted as a part of impulse control disorder in around frequency of 3.5 %.[26]

In this study 16(80%) of bvFTD group has altered sleep pattern, among majority were suffering from hyposomnia 10(50%). While 6(30%) has somnolence. Somnolence noted in FTD as 30-47 % which is similar to other neurodegerative disease.[9,27] Similarly 15(75%) patients of PDD had altered sleep pattern, among 10(50%) were suffering from hyposomnia while 5(25%) has somnolence. 80% of PD patients suffering from insomnia[28] while 50% experiences excessive daytime sleepiness.[29] REM sleep behavior disorder also noted frequently. (30-90%)[30]

Repetitive behavior were common in the FTD (50%). Repetitives tereotypies were reported in 35% bvFTD

Table 3: Behavior symptoms in bvFTD and PDD group

	FTD Number (n) (%)	**PDD Number (n) (%)**	**Chi-square value**	*p* **-value**
A. **Affect and social behavior**s				
Lack of apathy/ empathy	19(95.0%)	6(30.0%)	18.0267	0.0001*
Exaggerated emotional display/Aggressiveness/ Irritability	11 (55.0%)	5(25.0%)	3.7500	0.0528
Loss of embarrassment / Neglect of hygiene	13 (65.0%)	3(15.0%)	10.4167	0.0012*
Loss of insight	16(80.0%)	16(80.0%)	0.0000	1.0000
Disinhibition/ Selfishness	19(95.0%)	7(35.0%)	15.8242	0.0001*
B. **Sensory Behavior**s				
Loss of awareness of senses including pain	14(70.0%)	1(5.0%)	18.0267	0.0001*
Loss of smell	15(75.0%)	12(60.0%)	1.0256	0.3111
Exaggerated sensory (paresthesia &heat / cold) response	6(30.0%)	4(20.0%)	0.5333	0.4652
C. Eating and vegetative behaviors				
Preference for sweet food	6(30.0%)	4(20.0%)	0.5333	0.4652
Preference for savoury foods	1(5.0%)	1(5.0%)	0.0000	1.0000
Loss of discrimination	17(85.0%)	5(25.0%)	14.5455	0.0001
Overeats	8(40.0%)	2(10.0%)	4.8000	0.0284
Oral exploration of objects	5(25.0%)	1(5.0%)	3.1373	0.0765
Wandering	14(70.0%)	4(20.0%)	10.1010	0.0014
Pacing	10(50.0%)	2(10.0%)	7.6190	0.0057
Altered Sexual behavior	13(65.0%)	7(35.0%)	3.6000	0.0577
Hypersomnia	6(30.0%)	5(25.0%)	0.1254	0.7232
Hyposomnia	10(50.0%)	10(50.0%)	0.0000	1.0000
C. **Repetitive and compulsive behavio r**s				
Stereotypes/ Adherence to daily routine	7(35.0%)	2(10.0%)	3.5842	0.0583
Repetitive acts	10(50.0%)	0	1.0256	0.3111
O bsessive-compulsive behavior	5(25.0%)	1(5.0%)	3.1373	0.0765
E. **Environmental dependency related behavio r**s				
Hoarding and handles objects	2(10.0%)	2(10.0%)	0.0000	1.0000
Echoparexia/ echolalia	2(10.0%)	0	2.1053	0.1467
Utilization phenomena	7(35.0%)	0	8.4848	0.0035
F. **Cognitive mediated behavio r**s				
Mislays/ Fails to recognize/ Difficulty in locating objects	13(65.0%)	8(40.0%)	2.506	0.1133
Lost in familiar surroundings/ Disorientation in own home	9(45.5%)	7(35.0%)	0.4167	0.5186
Use wrong word	9(45.0%)	5(25.0%)	1.7582	0.1848
Mutism	19(95.0%)	7(35%)	15.824	0.0001
G. **Psychosis related behavio r**s				
Delusions	2(10.0 %)	10(50.0%)	7.6190	0.0057
Visual illusion/hallucination	**4(20.0%)**	**14(70.0%)**	**10.1010**	**0.0014** *
Auditory illusion/ hallucination	2(10.0%)	3(15.0%)	0.228	0.632

compare to 10% of PDD. The Bathgate et al record ed these stereotypies in 27%-60%, bvFTD patients.[9] A Mario F et al observed preservatives and stereotyped behavior n 45.3% FTD at presentation and increase up to 88.7% in 2 year follow up.[18] Though impulse control disorder are well document in PD(13.6%)[31] but there is barely any study on repetitive and compulsive behaviors in PDD. Pay - Chiu et al's study showed 16 % PDD patient has repetitive behaviors.[15] Obsessive-compulsive features present in both group in range of 25%. Obsessive behavior observed in 17-40 % FTD in past studies.[9,18,29] There are studies showed these features in PD around 6-22%.[28,32]

Our study revealed environmental dependency related behaviors are equally common. Among these utilization behaviors was attend statistical significant. Utilization behavior was reported in seven patients of bvFTD group. This very interesting clinical sign observed in more than 75% Indian bvFTD patients,[33] while other reported this behavior very less frequently.[9,11,18] These environmental dependency related behavior are barely described in PDD.

Behaviors relating to cognition and spatial dysfunction were more common in bvFTD group(70%) than in PDD group (60%). The impaired executive dysfunction and attention observed in half of patients with PDD in past.[34,35] Interestingly these changes also observed in predemented stages.[36,37] On other side executive dysfunction in bvFTD reported as 23-53 % at initial presentation.[9,16,22] The symptoms related to visuospatial-perceptual function also observed in both group (65 % v/s 40%). Past studies showed relatively preservati on of visuospatial skills in bv FTD.[9,18] In PDD greater deficit assessed in range of 35-50%.[36,38]

Psychosis related features are present more pronouncedly in PDD than bvFTD group that is 80% v/s 50%. Delusions note d in (50%) of PDD while 10% of bvFTD patients give such history. Visual hallucinations observed in 70% PDD and 20% of bv FTD patients. Auditory illusions-hallucinations were numerically less common in FTD patients than in PDD. (10% v/s 15%). In PDD delusion and hallucination occurs frequently as 30% and 45%-65% respectively.[13] A comparative study showed delusion in around 40% and hallucination in 30% in PDD patients.[15] These studies also suggested that presence of visual hallucination in PD predict development of dementia. In compare to PDD in bvFTD psychotic behavioral reported unusually. In one study delusion reported in10-30% patients while hallucination reported in only 7-10% patient.[9] Psychotic symptoms such as delusions and hallucinations are surprisingly rare in study by Mario F et al.[18]

Our current research focuses on behaviours characters in a cross section. There is further scope for a prospective longitudinal study to confirmed or revaluate our findings. We also thought requirements of behavioral score which based on our findings to aid in diagnosis of various degenerative dementia e.g. AD, FTD, PDD, DLB and others.

5. Conflict of Interest

None

6. Source of Funding

None

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