Problems of Face Recognition in Patients with Behavioral Variant Frontotemporal Dementia

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ABSTRACT

Introduction: Faces are very special as they are most essential for social cognition in humans. It is partly understood that face processing in its abstractness involves several extra striate areas. One of the most important causes for caregiver suffering in patients with anterior dementia is lack of empathy. This apart from being a behavioral disorder could be also due to failure to categorize the emotions of the people around them. Patients and Methods: Inclusion criteria: DSM IV for Bv FTD Tested for prosopagnosia - familiar faces, famous face, smiling face, crying face and reflected face using a simple picture card (figure 1). Exclusion Criteria: Advanced illness and mixed causes. Observations: 46 patients (15 females, 31 males) 24 had defective face recognition. (mean age 51.5), 10/15 females (70%) and 14/31 males (47). Familiar face recognition defect was found in 6/10 females and 6/14 males. Total- 40%(6/15) females and 19.35%(6/31) males with FTD had familiar face recognition. Famous Face: 9/10 females and 7/14 males. Total- 60% (9/15) females with FTD had famous face recognition defect as against 22.6%(7/31) males with FTD Smiling face defects in 8/10 female and no males. Total- 53.33% (8/15) females. Crying face recognition defect in 3/10 female and 2/14 males. Total- 20%(3/15) females and 6.5%(2/31) males. Reflected face recognition defect in 4 females. Results: Famous face recognition and positive emotion recognition defect in 80%, only 20% comprehend positive emotions, Face recognition defects are found in only 45% of males and more common in females. Conclusion: Face recognition is more affected in females with FTD There is differential involvement of different aspects of the face recognition could be one of the important factor underlying decline in the emotional and social behavior of these patients. Understanding these pathological processes will give more insight regarding patient behavior.

Key words: Behavior, dementia, extrastrate system, face recognition, social cognition

INTRODUCTION

Faces are very important for social interaction not only in humans but also in nonhuman primates and other animals. How people categorize faces and their emotions is very exciting. Although attractiveness is defined by different subjective parameters in

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How to cite this article: Chandra SR, Patwardhan K, Pai AR. Problems of face recognition in patients with behavioral variant frontotemporal dementia. Indian J Psychol Med 2017;39:653-8.
different cultures, there is uniformity in several aspects across races and regions.\[^{11}\] It is believed that frontal lobe, temporal lobe, and amygdala respond disproportionately to face stimuli though they are not visual areas. Judging identity, trustworthiness, etc., probably has a role in occipital face area and judging nonface objects from face happens in fusiform area and extrastriate areas. Higher-order face recognition involves functionally and structurally distinct regions of the brain. As per Bruce and Young model,\[^{2}\] expression independent of description takes place at visual system, superior temporal regions for gaze, lip movement, intraparietal sulcus for spatial attention, auditory cortex for prelexical speech perception, amygdala and insula for emotion recognition, anterior temporal for name, biography, and the whole cognitive system gets involved in the core system of face processing.

Self-faces are unique as none of us have seen our own face. Reflected faces, in addition, have a right left shift. Right medial temporal lobe structures participate in memory for faces at the time of visual experience but not in the subsequent recognition of recently stored memories. Right hippocampus and adjacent cortex are activated by unfamiliar faces. Face memory encoding and recognition are largely dissociable. While left prefrontal and left inferior temporal areas were associated with face memory encoding, right prefrontal and bilateral parietal and ventral occipital areas were associated with face recognition.\[^{3}\] It was very interesting for us when we encountered patients who could not identify their own reflected face as theirs and mistook them but could identify correctly their dress and ornaments and wondered why the person in the mirror is wearing their belongings.\[^{4,5}\] One of our patients even imagined the person in his mirror image as his bedroom cupboard as his nephew and was shocked when that image disappeared when the wife of the patient opened the cupboard to see to whom her husband was conversing. The patient was shocked at the disappearance of his nephew and went to police station to complain that his wife had murdered his nephew. Therefore, face recognition and its defects have far-reaching consequences in the life of patients who have problems in this skill. As lack of empathy is an important and common cause for caregiver burden in patients with dementia, we made an attempt to study the role of face recognition defect in patients with frontotemporal dementia (FTD).

Functional imaging techniques have indicated that right fusiform gyrus is strongly associated with identifying faces. However, there is controversy regarding the recognition of whose face it is, that is the ability to match with the name, the emotion, familiarity, voice, and famousness. Object recognition and face recognition probably involve different neural circuits.\[^{6}\] Information flow from modality-specific modules (voice and face) and semantic synthesis of information takes place at superior temporal gyrus (vision and face), at fusiform gyrus, and then transferred to retrosplenial space for emotional salience and episodic memory. This stored knowledge of personal identity is likely involved in famous face recognition involving temporoparietal region which involves no face processing but only retrieval from person identity system. Patients with anterior temporal cortex lesion can, however, identify the emotions in these cases and can also do familiarity recognition as well known or unknown. Hence, it is possible that different brain areas are involved in the multimodal processing of a person’s face. Studies indicate that negative emotion appreciation is more affected than positive one.\[^{7}\]

**Frontotemporal dementia**

FTD is the second most common type of degenerative dementia.\[^{6}\] It comprises a heterogeneous group with varied clinical and pathological presentation.\[^{8}\] It includes the frontal variant, semantic variant, nonfluent aphasia, and some less common combination syndromes.\[^{9}\]

Agnosias are disorders of recognition. These gnostic functions are likely to play an important role in the behavior of patients. Faces are highly salient, significant, visual stimuli critical for successful negotiations with the social world. Face recognition is quick and accurate and lasts for long in memory. This is seen consistently across all human populations. Faces are not simply a recapitulation of operation performed during the initial visual experience. Ventral occipito-temporal cortex is associated with face perception, hippocampus for encoding, and right prefrontal for recognition and retrieval.\[^{3}\] Prosopagnosia can occur without object agnosia. Fusiform gyrus has an important role in perceptual analysis of face. Some prosopagnosic patients are unimpaired in recognizing facial emotions.\[^{10}\] Emotional features activate amygdala, insula, and orbitofrontal cortex.\[^{10,11}\] The structural aspects are recognized at occipito-temporal areas. Hence, it is likely that FTD patients may recognize the structural features and not the emotion. This may explain the inappropriate use of social feedback such as anger, sadness, fear, and disgust by these patients.

![Figure 1: Pictures used for test containing famous face, crying face, and laughing face](image-url)
Prosopagnosia in Alzheimer’s disease is of visuospatial nature and hence can recognize the individual when named or spoken to, but in FTD, it is multimodal and hence cannot be recognized by any mode.

PATIENTS AND METHODS

A total of 46 patients who satisfied the Diagnostic and Statistical Manual of Mental Disorders-4 criteria for dementia of frontotemporal type were evaluated with Mini–Mental Status Examination (MMSE), Clinical Dementia Rating Scale, neuropsychological tests, and neuroimaging. Then they were questioned for prosopagnosia. It was considered present when close caregivers expressed that patient has difficulty in recognizing known people. They were then tested for familiar face recognition using the ability to recognize spouse or relative living with them for at least 1 year. Famous face was tested using the face of Mahatma Gandhi. Smiling face and crying face were tested using some pictures [Figure 1]. The patients were first asked to identify the pictures and then were told to inform the emotions seen in those pictures. A minimum of five trials were given, and if they were right in more than three trials, they were labeled normal. This is not a validated test but the available Western face recognition pictures were unsuited for our rural patients and therefore this modification was followed in this study. Reflected face recognition was tested by making the patients identify his/her own reflected image. Only patients with an MMSE score more than 20 were taken for the study. Patients with a history of head injury, tumors, surgery, radiation, and mixed dementia were excluded from the study.

RESULTS

There were 15 females and 31 males. Their age varied from 36 to 67 years, and the mean age was 51.5 years. Totally 24 patients had features of defective face recognition. Out of them, 10 (70%) females and 14 (47%) males had prosopagnosia. Familiar face recognition defect was found in six females and six males with prosopagnosia. Famous face recognition defect was found in nine females and seven males. Smiling face recognition defects were found in eight females and in no male. Crying face recognition defect was seen in three females and two males. Reflected face recognition defect was seen in four females and in no male [Figures 2-10]. One patient with reflected face recognition defect could identify the ornaments and dress correctly. Two patients considered the reflected face as a rival and one patient could not recognize her own reflected face but could recognize her daughter-in-law’s reflected face. Radiological features of all these patients are shown in tables. All had features of FTD with minor involvement of other areas [Tables 1-3].
This study shows that females are more prone to prosopagnosia that could be due to the innate gender-based differences in the human brain where apoptotic process is more in parietal region in females and frontal region in males. These in turn help us to understand the impaired social behavior, communication, and response to different kinds of emotions of caregiver and friends by these patients which leads to a lot of problems in interpersonal relationship.

Table 1: Radiological features in the patients evaluated

| Frontal and temporal atrophy                     | Parietal atrophy | White matter signal changes                      | Parietal lobe signal changes |
|-------------------------------------------------|------------------|--------------------------------------------------|----------------------------|
| Bifrontal and temporal                          | No               | Periventricular frontal lobes                    | No                         |
| Bifrontal and temporal                          | No               | Periventricular, subcortical bifrontal           | Left superior parietal lobe|
| Diffuse                                         | No               | Periventricular, microbleed left temporal        | No                         |
| Supratentorial                                   | Biparietal       | UBO                                              | Biparietal lobar white matter|
| Bitemporal and left frontal                      | No               | Bifrontal gyriform signal changes                | Biparietal lobar white matter|

UBO – Unknown bright objects

Table 2: Continuation of radiological features in the patients evaluated

| Frontal and temporal atrophy                     | Parietal atrophy | White matter signal changes                      | Parietal lobe signal changes |
|-------------------------------------------------|------------------|--------------------------------------------------|----------------------------|
| Subcortical and bifrontal                       | No               | UBO bifrontal and biparietal subcortical         | Subcortical and bifrontal  |
| Mild diffuse atrophy                            | No               | No                                               | Mild diffuse atrophy       |
| Diffuse                                         | Biparietal       | No                                               | Diffuse                    |
| Bifrontal and caudate atrophy                   | No               | No                                               | Bifrontal and caudate atrophy|
| Left hemiatrophy                                | No               | Left parieto-occipital region                    | Left hemiatrophy           |
| Diffuse                                         | No               | Diffuse lobar subcortical and periventricular   | Diffuse                    |
| Supratentorial                                   | No               | No                                               | Supratentorial             |
| Supratentorial                                   | Biparietal       | Right subcortical                                | Supratentorial             |

UBO – Unknown bright objects
In this small group of patients with FTD, males are double the number of females. However, prosopagnosia of all types was seen in females more than in males. There was no real difference between the patients with different severities of disease. Smiling face recognition was more defective than crying face recognition as against what is reported in literature. Crying face recognition and reflected face recognition defect were seen only in females in this study. Inanimate object recognition is preserved in most of these patients. Negative emotions were better appreciated than positive emotions.

**DISCUSSION**

This study involving 46 patients with behavioral variant FTD showed some unique features. Our patients were tested using Indian faces as against the standardised face testing cards, as most of our patients were not literate and had difficulty in identifying western faces. However, we used the same card for every one of our patients and we found the responses very encouraging as the normal bystanders of the patients could recognise all the cards including the novel faces and famous faces correctly. In our patients as against information available in literature positive emotions were not recognized by patients. This is cause of concern as patients do not share the happy moments of their loved ones resulting in feeling of dejection in caregivers.

**CONCLUSION**

Face recognition is significantly affected in patients with FTD. Females are more affected than males with this disability. There can be differential involvement of different aspects of the face recognition from patients to patients which could be one of the important factors underlying decline in the emotional and social behavior of these patients, even when memory is better preserved. Unlike the information in literature, our patients showed more difficulty in identifying positive emotion than the negative one; however, a larger number is needed to really know the pattern of face recognition defects in these patients. Understanding of these pathological processes will give us more insight in understanding these patients’ behavior.

**Acknowledgment**

The authors would like to thank the Department of Neurology, National Institute of Mental Health and Neurosciences, Bengaluru, Karnataka, South India.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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**Table 3: Continuation of radiological features in the patients evaluated**

| Frontal and temporal atrophy | Parietal atrophy | White matter signal changes | Parietal lobe signal changes |
|-----------------------------|-----------------|----------------------------|-----------------------------|
| Diffuse                     | Biparietal      | UBO bifrontal and biparietal subcortical | Diffuse                     |
| Supratentorial              | No              | Bifrontal and parietal signal changes | Supratentorial              |
| Bilateral perisylvian and right temporal | No | No | Bilateral perisylvian and right temporal |
| No                          | No              | Bifrontal and parietal | No                          |
| Bifrontotemporal            | No              | Periventricular parietal lobe | Bifrontotemporal            |
| No                          | No              | No                         | No                          |

UBO – Unknown bright objects
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