Prosopagnosia: A rare presenting manifestation of frontotemporal lobar degeneration

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Abstract

Frontotemporal dementia is an important neurodegenerative disorder accounting for a significant proportion of dementia cases with onset before 60 years of age. Apart from the well recognized behavioral changes the disease has many other distinctive features like predominant language involvement alone or associated features of motor neuron disease or parkinsonism etc. which at times may be the presenting manifestation itself. In the following article we describe a rare presenting manifestation: prosopagnosia, in the setting of frontotemporal degeneration.

Key words: Frontotemporal degeneration, prosopagnosia

Introduction

Frontotemporal dementia (FTD) is an important neurodegenerative disorder accounting for approximately 50% of dementia cases presenting before 60 years of age. The term FTD was introduced by workers in Lundh (Sweden) and Manchester (UK) to refer specifically to the progressive behavioral syndrome. Later researchers found that the syndrome of FTD was not confined to behavioral changes alone, but can also affect the language domains. Subsequently, in 1998, the Neary criteria were put forward which recognized three clinical syndromes:

1) FTD 2) Progressive nonfluent aphasia 3) Progressive fluent aphasia (semantic dementia). In addition to the three clinical syndromes recognized by Neary and colleagues there is increasing recognition of other clinical features that can occur in the setting of frontotemporal lobar degeneration (FTLD). These include aphasic syndromes that do not meet the Neary criteria, parkinsonism, motor neuron disease (MND), corticobasal degeneration, progressive supranuclear palsy, and prosopagnosia. We describe a rare presenting manifestation in the setting of FTLD.

Case Report

A 55-year-old male presented with an 18-month history of progressive difficulty in recognizing familiar faces. Initially he could identify them by their voices, but later he could not identify them by their voices or even after the individual’s name had been mentioned to him. He had a difficulty in identifying some common objects encountered in day to day life. His behavior and language was normal and he had no impairment of executive function, memory, praxis, and visuospatial problems. He did not have any weakness, parkinsonism, bulbar symptoms or alien limb phenomenon. There was no family history of any significant illness and there was no history of any trauma or psychiatric illness.

Examination showed a right-handed, conscious, cooperative and attentive person. Formal neuropsychological tests, namely Mini Mental Status Examination (MMSE) and a standard neuropsychological battery devised to explore memory, language, praxis, and executive functions, visual and space perception was normal except rare circumlocution in the language domain. The most marked abnormality was his difficulty in recognizing familiar faces. He could not match names
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to faces and he failed to give any details concerning the
named persons including his close friends and relatives.
He also had some difficulty in identifying common
objects and could not identify blue color.

His routine blood, urine examinations, blood sugar,
renal and liver function tests and a chest X-ray were
normal. A computed tomographic scan showed marked
atrophy of the nondominant frontotemporal lobe
[Figure 1]. Electroencephalogram was normal. A lumbar
puncture showed normal cerebrospinal fluid pressure,
<5 cells (all mononuclear cells), protein 65 mg/dl and
sugar 45 mg/dl. He was seen last at 21 months into his
illness when he still had the cross-modal agnosia subtle
behavioral changes of excessive anger, irritability and
rare wandering episodes.

Discussion

This patient presented with prosopagnosia. The term
prosopagnosia was coined by Bodamer in 1947. It
refers to the inability to recognize familiar faces. The
lesions that cause prosopagnosia are usually found
in the ventral occipitotemporal cortex involving the
fusiform and lingual gyrus[1] and are bilateral in most
cases although right unilateral lesions[2] can cause the
syndrome. The criteria for the prosopagnositic variant
of FTD defined by Neary[3] include

I. Core diagnostic features
   A. Insidious onset and gradual progression
   B. Perceptual disorder characterized by
      1. Prosopagnosia: impaired recognition of
         identity, of familiar faces and/or
      2. Associative agnosia: impaired recognition
         of object identity

II. Supportive diagnostic features
   A. Pressure of speech
   B. Idiosyncratic word usage

C. Surface dyslexia and dysgraphia

III. Brain imaging (structural and/or functional):
    Asymmetric abnormality predominantly affecting
    nondominant (right) anterior temporal lobe.

Our patient satisfies the core criteria and the imaging
features. But our patient has a cross-modality loss of
person recognition than simple prosopagnosia in that
he cannot recognize persons by their voices or even
after their name has been mentioned. A survey of the
literature showed that at least in some of the cases
reported as progressive prosopagnosia the recognition
disorder is not confined to face recognition but rather
represents a cross-model impairment of person
knowledge i.e. semantic knowledge.[4,5] In this context
the understanding of how the human brain perceives
and recognizes faces becomes important. Studies have
shown that faces activate the bilateral fusiform gyrus
although the most robust activity is seen on the right
side. This region has been referred to as a fusiform
face area (FFA) and is defined as a module of face
perception.[6] Similarly, voices have been found to
activate the bilateral superior temporal gyrus, more
on the right.[7] These two modalities may access the
semantic store independently or they may converge
to form a multimodal association area for person
recognition from which they may access the semantic
store. In our patient the initial inability to recognize
faces with preserved recognition by voices and names
suggests damage to the right FFA. Subsequent inability
to recognize persons by voices suggests damage to the
auditory processing area. The final stage where he is
unable to recognize a person by any modality suggests
damage to the semantic store itself.

The list of clinical features associated with atrophy of
frontal and temporal lobes is increasing. What is the link
between the varying clinical features and the common
denominator i.e. frontotemporal atrophy is not clear.
Future research may shed light upon this and thus open
up new therapeutic targets for this disabling illness.

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Figure 1: CT showing extensive atrophy of the right temporal lobe


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