A. García-Caballero
J. González-Hermida
I. García-Lado
M. J. Recimil

Impaired facial emotion recognition in a case of right frontotemporal dementia

Psychiatry Service
Complexo Hospitalario de Ourense
SERGAS
Orense (Spain)

Introduction. After the description of its involvement in amydalin lesions, there has been growing interest in the last decade on the neuropsychological examination of impaired emotional recognition in different diseases. This study aims to demonstrate the existence of emotional recognition impairment in a case of frontotemporal dementia affecting right temporal lobe structures with an experimental battery.

Clinical case. The case of 7 year long frontotemporal dementia with right temporal predominance, clinically characterized by behavior disorders such as loss of hygiene habits, eating food in bad condition, approach to marginal groups and other psychiatric disorders (megalomanic delusional ideation) is presented. The psychiatric, neurological, neuropsychological and neuroimaging examination are described.

Methods. Facial recognition impairments were assessed with a modification of Ekman and Friesen Task (1976). The results were compared with those obtained in three controls matched by age, and educational level.

Results. The case we report showed marked impairment in discrimination, matching, selection and naming of negative facial emotions (anger, fear, sadness and disgust). The impairment was more striking in the selection and naming paradigms. Anger was the most affected emotion. It was hypothesized if the impairment of emotional recognition could be in the base of certain behavior disturbances of the patient such as approach to marginal groups.

Key words:
Frontotemporal dementia. Recognition. Emotion. Facial expression.

Actas Esp Psiquiatr 2006;34(6):416-419

Alteración en el reconocimiento de emociones faciales en un caso de demencia frontotemporal de predominio derecho

Introducción. Tras la descripción de su compromiso en lesiones amigdalinas, en la última década se ha producido un creciente interés por la exploración neuropsicológica de las alteraciones del reconocimiento emocional en diferentes patologías. El propósito de nuestro trabajo ha sido demostrar por medio de una batería experimental la existencia de alteraciones en el reconocimiento emocional en un caso de demencia frontotemporal de predominio temporal derecho.

Caso clínico. Se presenta un caso de demencia frontotemporal de predominio temporal derecho de 7 años de evolución, clínicamente caracterizado por alteraciones conductuales, como pérdida de hábitos higiénicos, consumo de alimentos en mal estado, aproximación a grupos marginales y otras alteraciones psiquiátricas (ideación delirante megalomaníaca). Se realiza la descripción de la exploración psiquiátrica, neurológica, neuropsicológica y de neuroimagen.

Métodos. Las alteraciones del reconocimiento de emociones faciales son evaluadas por medio de una batería experimental derivada de la prueba de Ekman y Friesen (1976), comparando los resultados con los obtenidos en tres controles pareados por edad y nivel educativo.

Resultados. El caso que presentamos presenta alteraciones en la discriminación, emparejamiento, selección y denominación de emociones faciales especialmente negativas (ira, miedo, tristeza y asco). El compromiso fue más llamativo en los paradigmas de selección y denominación. La emoción más afectada fue la ira. Se hipotetiza si la alteración del reconocimiento emocional podría estar en la base de determinadas alteraciones conductuales del paciente como el acercamiento a grupos marginales.

Palabras clave:
Demencia frontotemporal. Reconocimiento. Emoción. Expresión facial.

INTRODUCTION

Frontotemporal dementia is a neurodegenerative disease that predominantly affects the frontal lobe and anterior

Correspondence:
Alejandro García-Caballero
Unidade de Saúde Mental A Ponte
Xral. Aranda, 8
32001 Ourense (Spain)
E-mail: Alejandro.Garcia.Caballero@sergas.es

portion of the temporal lobe. On the contrary to that which occurs in Alzheimer's dementia, characterized by memory loss, frontotemporal dementia occurs with early behavior changes such as apathy, disinibition, and emotional impairments[1].

Frontotemporal dementias make up a group of diseases with differentiated locations, symptoms and anatomicopathological characteristics[1]. We present the case of frontotemporal dementia with right anterior temporal predominance.

The anterior pole of the temporal lobe (especially amygdala) is considered essential in the processing of emotions[2,3]. The existence of laterality in regards to the subtype of emotions processed is debated in the literature[4,5]. The purpose of this study has been to characterize facial emotion recognition impairments that a patient with extensive atrophy of the anterior pole of the right temporal lobe had.

CLINICAL CASE

Patient F. C. is a 61 year old man, right-handed, without family background of neurological or psychiatric disease. He studied technical engineering in the chemical branch, although his professional career was mainly dedicated to the real estate business. He is a widower who lives alone, since behavior disorder appeared after the death of his wife that led to the fact that his two minor children are being cared for by a sister in another city. The patient maintains contact with them and goes to visit them frequently. The family reports that the behavior disorders of the patient began when his wife became ill seven years ago, appearing a florid delusional picture having megalomanic overtones in which the patient states he has discovered a technique to cure cancer. The behavior problems he has had are hygiene and dressing disorders, irritability, psychomotor restlessness, he does not accept advice or recommendations from his family and he eats food in bad condition. They also state that he has begun to have relationships with persons having a marginal life, which makes them fear that he may be the object of economic abuses.

The patient was admitted to the Psychiatry Unit involuntarily three years after the death of his wife (four years after the onset of the symptoms) due to deterioration of his hygiene habit and bizarre behaviors. A cranial CT scan was performed during this admission that showed hemispheric asymmetry with mild atrophy having right predominance, but whose entity was not considered sufficiently relevant to justify the clinical picture. A cognitive study was done but did not supply conclusive data (CME 29/30). The patient was transferred on request of his family to another hospital with the diagnosis of delusional disorder and cognitive deterioration to be studied.

Two years after his admission, he returned again involuntarily, observing megalomanic overtones with significant and stable neuropsychological disorders after discharge.

Neuropsychological examination

Partial disorientation in time. Serious impairment in fixation memory. No attentional impairment was seen. CME: 28/30. Addenbrooke´s Cognitive Examination (ACE)[6]: 63/100 (Cut-off for his educational level positive for dementia ≤ 74 points). Verbal phonetic fluency FAS: 3-3-2 severe deficit. Verbal semantic fluency (animals): 8. Motor sequences of Luria: altered. Stroop: normal. Boston naming test: severe abnormality. Normal calculation. Normal visual-construction. No prosopagnosis is observed; the patient correctly recognizes famous persons of the experimental test - Famous Face Test (FFT) (Regional Government of Galicia: PGIDIT 03SAN 92302) between three photographs of confusion matched by age and gender (50 trials). However, he has serious problems to remember proper names, including those of family whom he recognizes.

Neurological examination: normal reactive isocoric pupils, normal pairs, conserved reflexes and sensitivity, no frontal release reflexes are seen.

Complementary examinations: complete blood count and biochemistry normal, thyroid hormones normal; B1, B12, and folic acid normal; leutic serology, HIV and hepatitis negative. MRI: asymmetric cortical atrophy of right predominance with predominant involvement in anterior pole of right temporal lobe. Dilation of lateral ventricles and III ventricle (fig. 1).

Figure 1 *MRI coronal cut. Observe the extensive atrophy of the right temporal anterior pole and asymmetry regarding left hemisphere.*

MATERIAL AND METHODS

According to Rosen et al.[7], the following functions must be studied separately for the study of facial emotion recognition impairments.

- *Identity discrimination.* Confusion photographs used in the FFT, creating a Power Point presentation with two photographs in black and white of 8×10 cm per slide were used. The photographs were matched by age and gender. Twenty trials were made.
- *Emotion discrimination.* Two photographs selected for this from the *Emotion Recognition Task* of Ekman and Friesen[8] were modified, constructing a presentation of 20 slides, each one with two 8 × 10 cm photographs. In each test, 12 slides with different emotions and 6 (one for each basic emotion) presenting the same emotion, always matched by gender, were used. The resulting score was dichotomic (correct/incorrect).
- *Emotion naming.* We used the Ekman battery[8], scanning the originals and having transformed them into a portable presentation of Power Point. The 8×10 cm photographs include a legend that indicates the names of six fundamental emotions, the test consisting in a forced choice between these six emotions. The battery consists of 60 trials (10 for each emotion) plus six initial test trials.
- *Emotion selection.* On the basis of the Ekman battery[8] six photographs from the same individual showing the six basic emotions were selected in each slide. In the test, the patient is given a verbal order to indicate the different emotions sequentially. Five trials of six emotions each one is done.
- *Matching.* A battery consisting in six slides with five 8 × 10 cm photographs was made. The stimulus photograph is in the lower part and there are four photographs above with different emotions taken from the Ekman battery[8]. The patient should match the stimulus photograph with that presenting the same emotion. Six trials were done.

Selection of controls

The data corresponding to the case were compared with three controls without psychiatric or neurological background of interest and who were comparable with the patient in regards to age, gender and educational level.

Table 1 Results in discrimination (total correct) and matching (total correct)

	Descrimination (20)	Matching (6)
Patient	12	4
Controls (DS)	18.33 (1.15)	6 (0)

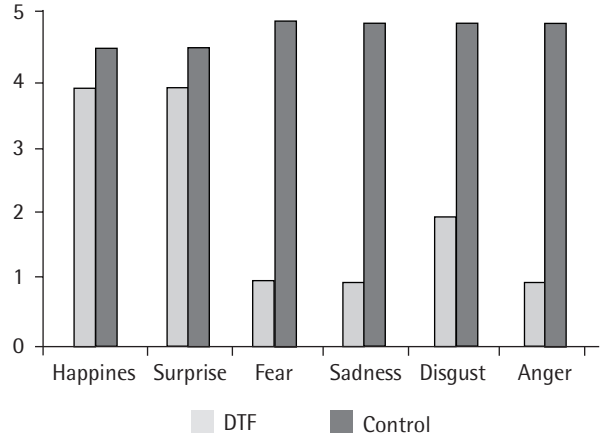

Figure 2 *Selection emotions.*
DTF: frontotemporal dementia.

RESULTS

No impairments in identity discrimination were observed, the results of the patient being comparable with those of the control group.

The results corresponding to the discrimination and matching tests are summarized in table 1.

The results corresponding to selection and identification are summarized in the next figures (figs. 2 and 3).

DISCUSSION

Frontotemporal dementias typically appear at the beginning of the senile age, it being characteristic that the

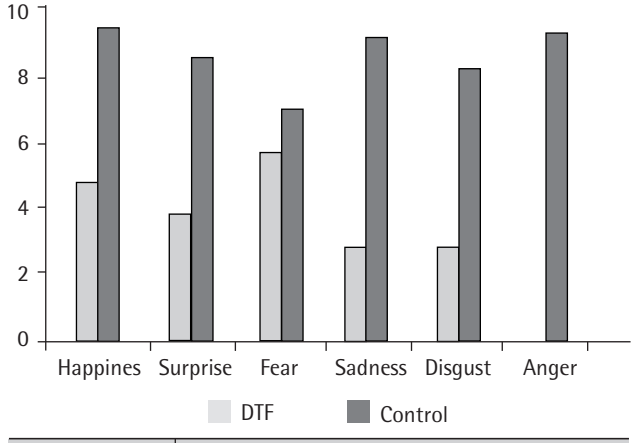

Figure 3 *Naming emotions.*
DTF: frontotemporal dementia.

symptoms are insidious and the diagnosis is reached after several years of evolution[9]. As we mentioned in the introduction, behavioral disorders and language impairments that depending on the region affected cause fluent aphasias (semantic dementia)[10] or not fluent aphasia (primary progressive aphasia)[11] are typical. When the dementia causes predominant atrophy in the right temporal lobe, the speech involvement is less prominent, although in our case, as well as in others described in the literature[1], there is a significant abnormality that does not improve with phonetic or semantic cues.

However, those that are most interesting in the case in question are emotional recognition impairments. The patient has impairments in all the tests performed (table 1 and figs. 2 and 3). With the selection paradigm, the patients correctly answered the positive emotions: happiness and surprise, failing in the negative ones, especially in those with greater valency (sadness, fear and anger). In the naming paradigm, the results are worse in all the emotions, include the positive ones. The existence of no correct answer in anger and a better than expected result in fear stands out.

These results agree with that described in the literature[2,4,5,7]. Our patient, with an extensive right anterior temporal lesion (fig. 1), is severely incapacitated to recognize negative facial expressions, not only fear, but also anger, sadness and disgust. In the case of anger, the result is very clear in the naming paradigm (fig. 3) and contradictory with the Anderson et al. study[3]. This difference could be because of the involvement of the left amygdala[12]. From the behavioral point of view, it is possible that the approach of the patient to marginal groups is related with this impairment of the temporal lobe emotional recognition mechanisms, that could affect «somatic marker» functioning[13]. This possible impairment that could easily be demonstrated with a dermal conductance study, has not been confirmed at present in other cases.

ACKNOWLEDGEMENTS

The famous faces test has been financed with a research grant by the Regional Government of Galicia: PGIDIT 03SAN 92302. We thank Dr. Purificación Sevilla, of the Psychiatry Service, and Dr. Guillermo Ozaita, of the Neurology Service, both from the Complexo Hospitalario of Orense, for their collaboration in the case study.

REFERENCES

1. Edwards-Lee T, Miller BL, Benson DF, Cummings JL, Russell GL, Boone K, Mena I. The temporal variant of frontotemporal dementia. Brain 1997;120:1027-40.
2. Anderson AK, Phelps EA. Lesions of the human amygdala impair enhanced perception of emotionally salient events. Nature 2001; 411:305-9.
3. Anderson AK, Spencer DS, Fulbright RK, Phelps EA. Contribution of the anteromedial temporal lobes to the evaluation of facial emotion. Neuropsychology 2000;14:526-36.
4. Anderson AK, Phelps EA. Is the human amygdale critical for the subjective experience of emotion? Evidence of intact dispositional affect in patients with amygdala lesions. J Cogn Neurosc 2002;14:709-20.
5. Adolphs R, Tranel D, Denburg N. Impaired emotional declarative memory following unilateral amygdala damage. Learn Mem 2000; 7:180-6.
6. García-Caballero A, García-Lado I, González-Hermida J, Recimil MJ, Area R, Manes F, et al. Validation of the Spanish version of the Addenbrooke's Cognitive Examination in a rural community in Spain. Int J Geriatr Psychiatry 2006; 21:1-7.
7. Rosen HJ, Perry RJ, Murphy J, Kramer JH, Mychack P, Schuff N, et al. Emotion comprehension in the temporal variant of frontotemporal dementia. Brain 2002;125:2286-95.
8. Ekman P, Friesen WV. Pictures of facial affect. Palo Alto: Consulting Psychologists Press, 1976.
9. Hodges JR. Frontotemporal dementia (Pick's disease). Clinical features and assessment. Neurology 2001;56:S6-10.
10. Hodges JR, Patterson K, Oxbury S, Funnell E. Semantic dementia. Progressive fluent aphasia with temporal lobe atrophy. Brain 1992;115:1783-806.
11. Mesulam MM. Slowly progressive aphasia without dementia. Ann Neurol 1982;11:592-598.
12. Scott SK, Young AW, Calder AJ Hellawell DJ, Aggleton JP, Johnson M. Impaired auditory recognition of fear and anger following bilateral amygdala lesions. Nature 1997;385:254-7.
13. Damasio AR. The somatic marker hypothesis and the possible functions of the prefrontal cortex. Philos Trans R Soc Lond B Biol Sci 1996;351:1413-20.