References

1. Foix C, Chavany JA, Marie J. Diplégie facio-linguo-masticatrice d'origene cortico sous-corticale sans paralyse des membres. Rev Neurol (Paris). 1926;33:214-9.
2. Bruyn GW, Gathier JC. The opercular síndrome. En: Vinken PJ, Bruyn GW, editores. Handbook of Clinical Neurology. Vol. 2. Localization in Clinical Neurology. Amsterdam: North-Holland Publishing; 1969. p. 776-83.
3. Krishnan E. Stroke subtypes among young patients with systemic lupus erythematosus. Am J Med. 2005;118:1415.
4. Mikdashi J, Handweger B, Langenberg P, Miller M, Kittner S. Baseline disease activity, hyperlipemia, and hypertension are predictive factors for ischemic stroke and stroke severity in systemic lupus erythematosus. Stroke. 2007;38:281-5.
5. Campello I, Velilla A, López-López A, Tapiador MJ, Marta E, Martín-Martínez J. Lesión biopercular con disociación inversa. Rev Neurol. 1995;23:1056-8.
6. Moragas Garrido M, Cardona Portala P, Martínez Yélamos S, Rubio Borrego F. Heterogeneidad topográfica del síndrome de Foix-Chavany-Marie. Neurologia. 2007;22:333-6.
7. Puertas I, García-Soldevilla MA, Jiménez-Jiménez FJ, Cabrera-Valdivia F, Jabbour T, García-Albea E. Mano distónica bilateral secundaria a síndrome biopercular o síndrome de Foix-Chavany-Marie. Rev Neurol. 2002;35:430-3.
8. Jiménez-Jiménez FJ, Puertas I, Alonso-Navarro H. Hemimasticatory spasm secundary to biopercular síndrome. Eur Neurol. 2008;59:276-9.
9. Konieczny PL, Eidelman BH, Freeman WD. Teaching video neuroimage: Foix Chavany Marie syndrome. Neurology. 2008; 70:88.
10. Szabo K, Gass A, Robmanith C, Hirsch JG, Hennerici MG. Difussion-and perfussion-weighted MRI demostrates synergistic lesions in acute ischemic Foix-Chavany-Marie syndrome. J Neurol. 2002;249:1735-7.
11. Kitagawa Y, Gotoh F, Koto A, Okayasu H. Stroke in systemic lupus erythematosus. Stroke. 1990;21:1533-9.
12. Futnell N, Millikan C. Frecuency, etiology, and prevention of stroke in patients with systemic lupus erythematosus. Stroke. 1989;20:583-91.

Frontotemporal dementia and motor neuron disease

Demencia frontotemporal y enfermedad de motoneurona

Dear Editor:

Degenerative diseases are the most common cause of dementia in our environment and, within these cases, 5-10% are frontotemporal dementias (FTD)[1,2]. This percentage increases when patients are younger than 65 years old, being the second leading cause of pre-senile dementia[3]. The major neurochemical alterations are deficits in serotonin and dopamine[2]. According to Diagnostic and Statistical Manual of Mental Disorders (4th ed.) (DSM-IV) criteria, the diagnosis of FTD is mainly clinical, consisting of behavioural alterations, psychotic disorders and language disorders, among others, aided by neuroimaging and neuropsychological tests[1].

An association with other neurodegenerative diseases has been noted, including motor neuron disease (MND), a scarcely frequent[3] association, which is clinically characterised by frontal and neurological signs[4]. FTD associated with MND is classified according to whether the condition is the first, second or both motor-neurons, and amyotrophic lateral sclerosis (ALS) is the most frequent form[4].

We present the case of a patient with FTD, with an evolution of 1 year, who developed MND.

A 59-year-old woman, right handed, with no family history of neurodegenerative disease or personal history of psychiatric illness. Only a mild dyslipidemia can be highlighted. The patient had suffered from alterations in personality, talkativeness, daytime sleepiness, easy laughter, great difficult in crying and psychotic disorders for 1 year: "the paintings in her house talked to her, and an impairment of speech had appeared in the last month".

Physical examination, with cardiopulmonary auscultation, was normal. Neurological examination showed that the patient was oriented in all three areas; she presented mild dysarthria, normal cranial nerves, except for a slight left hypoglossal paresis with some fasciculation in that territory; without myoclonus, ataxia and dysmetries; the strength, tone, sensation and reflexes were normal in the limbs and there were no fasciculations. Gait was also normal. In the neuropsychological study, the results were: MMSE, 29/30; clock test, 9/10; verbal fluency, 12; Trail Making A, 48 s; FAB, 13. Additional tests performed (biochemical, haemogram, serology (including HIV), thyroid hormones, vitamin B12 and folic acid) were normal.

Cranial computed tomography (CT): temporal atrophy. Cerebral and cervical magnetic resonance imaging (MRI): temporal atrophy and small meningioma of the right frontal sulcus; the rest was normal. Conduction studies of motor and sensory nerves were normal and no blockages were observed. The electromyographic study of muscles dependent on the cervical, lumbosacral and bulbar regions revealed a diffuse and asymmetric neurogenic pattern, with greater expression in the left side, with mild to moderate acute denervation activity. Thus, the patient met the DSM IV diagnostic criteria for frontotemporal dementia, associated in this case with motor neuron disease.

The great importance of the social impact of FTD, as of MND, its pre-senile onset, higher frequency in cases of family history (up to 50% in FTD), the burden on caretakers and the possibility of carrying out genetic advice and study[3] (up to 20% of patients with an association of these two diseases may have a history of ALS and up to 40% of FTD[5]), makes us give more importance to this association since, although rare, in recent years it is being noted that it is becoming more important, perhaps because it was previously underdiagnosed.

The FTD diagnosis is mainly clinical; we rely on the DSM-IV criteria, consisting of personality changes, alterations in behaviour and in language, among others. Greater relevance should be given to psychotic disorders such as paranoid delusions, which barely receive importance in most studies. However, one study has observed it in up to 30.3%of patients as a first complaint, and in up to 69.2%in the first visit[1].

We must remember that with the use of imaging techniques it is possible to find frontotemporal atrophy in 62% and even decreased glucose metabolism in frontal areas[3,6]. However, this is not always the case and we may not find atrophy or, as in this case, an atrophy of temporal areas in isolation, without a frontal condition.

The incidence of FTD in Spain is 14/100,000 inhabitants, and the incidence of ALSis 5-6/100,000 in Western European countries; only 2%do develop the ALS-dementia complex[7,8]. This indicates that it is highly unlikely for the two diseases to occur together, but there are studies showing that in up to 10% of FTD cases, where there was no MND clinic, a motor-neuron condition can be observed[4,9].

In our case, we noticed the speech alteration described by the patient's family, which led us to objectify the hypoglossal paresis and thus to search for MND, as she presented no other clinical manifestations that made us think of it.

Both diseases usually occur almost simultaneously and, when this happens, have a severe, rapid progression[6,10]. In this patient, contrary to what has been described, the MND appears (currently, subclinical) after 1 year of evolution of the FTD, no doubt of slow evolution, which confers her better prognosis than expected. Although at present she only presented affection of the lower motor neuron, she will most likely end up suffering first motor neuron lesions.

Theprognosisinthesecases,wherebothneurodegenerative diseases coexist, is very unfavourable; it can vary between 4 and 10 years[3]. For all these reasons, it is vitally important to search for such an association, to anticipate all the processes that may be necessary in the case of future complications, specific to the disease progression.

Ultimately, this case highlights that the occurrence of MND is not always simultaneous with that of FTD, nor must it be accompanied by a terrible evolution. We therefore consider that, in patients with FTD, adequate clinical monitoring and thorough neurological examination designed to accept that signs of motor-neuron disease might coexist are of great importance. This requires keeping it in mind in our differential diagnoses when faced with a cognitive impairment of the frontal type.

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References

1. Uterga JM, Rodríguez Iriarte ML, González García AI, Taramundi S, Alvaro LC, Cortina C. Demencia con atrofia frontal: estudio clínico de 18 pacientes con deterioro intelectual y degeneración de los lóbulos frontales según la neuroimagen. Rev Neurol. 2002;34:709-714.
2. AlonsoNavarro H, Jabbour Wadih T, Ayuso Peralta L, Jimenez Jimenez FJ. Neuroquímica y neurofarmacología de la demencia frontotemporal. Rev Neurol. 2006;42:556-61.
3. Peñe R, Campdelacreu J, Escrig A, Gascon Bayarri J, Hernandez Pardo M, Jauma S, et al. Degeneración lobular frontotemporal: estudio descriptivo de 42 pacientes. Neurología. 2008;23:511-7.
4. Toribio Diaz ME, Morera Guitart J. Clasificación clínica y biomolecular de las demencias frontotemporales. Revisión de la bibliografía. Rev Neurol. 2008;47:588-98.
5. Spina S, Murrell JR, Huey ED, et al. Clinicopathologic features of frontotemporal dementia with Progranulin sequence variation. Neurology. 2007;68:820-7.
6. Lomen Hoerth C, Anderson T, Miller B. The overlap of amyotrophic lateral sclerosis and frontotemporal dementia. Neurology. 2002;59:1077-99.
7. Lopez Pousa S, Garre Olmo J, Turon Estrada A, et al. Incidencia clínica de la demencia frontal. Rev Neurol. 2002;34:216-22.
8. Duque P, Paramo MD, Borges M, et al. Alteraciones neuropsicológicas en la esclerosis lateral amiotrófica. ¿No existen o no se detectan? Rev Neurol. 2003;36:3-8.
9. Jeong Y, Park KC, Cho SS, et al. Pattern of glucose hypometabolism in frontotemporal dementia with motor neuron disease. Neurology. 2005;64:734-6.
10. Mckhann GM, Albert MS, Grossman M, Miller B, Dickson D, Trojanowski JQ. Clinical and pathological diagnosis of frontotemporal dementia: report of the Work Group on Frontotemporal Dementia and Pick's Disease. Arch Neurol. 2001;58: 1803-9.

PANDAS: adult variant

PANDAS, variante del adulto

Dear. Editor:

Streptococcal infection in childhood is usually benign and with a self-limited presentation, although a small percentage of children may develop neurological and/or neuropsychiatric disorders[1]. Sydenham chorea (SC) is the best known of these entities, but PANDAS(PaediatricAutoimmuneNeuropsychiatric Disorder Associated with Streptococcal throat infection) is another well-defined syndrome in which tics and obsessive-compulsive disorder (OCD) may be caused by beta-haemolytic group A streptococci (BHGAS) infection. SC is the neurological manifestation of rheumatic fever (RF) and, although it is true that there may be overlap between this entity and PANDAS, the latter is well distinguished from SC by the absence of carditis (with equal prevalence in the general population) and chorea infection itself[2,3].