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].

This was an 18-year-old female, veterinary student, who underwent tonsillectomy at age 16, without a family history of movement disorders, who referred a clinical case of abrupt onset, of 36 hours of evolution, characterised by involuntary movements (occasionally controllable for a short period) that began at the proximal level in arms, trunk and neck; these movements disappeared with sleep and increased with stress and lack of rest. A few days before these clinical signs commenced, she presented an isolated fever spike, without an apparent infectious focus. She denied consumption of toxic substances, oral contraceptives or other drugs.

The general physical examination was unremarkable (no fever). No skin nodules or erythematous lesions were observed. In the neurological examination, higher functions and language were normal. The cranial nerves were also normal and eye-tracking was good. Segmental force was preserved 5/5, with a myotatic reflexes symmetrically alive +++/++++, with Gordon's phenomenon present (Hung-Up Knee Jerks). Discrete generalized hypotonia was observed, and there were also some complex movements (with choreiform elements and motor tics) in the arms, trunk and neck. There were no data of motor impersistence. She did not present associated bradykinesia. Sensitivity, coordination and gait were preserved.

She underwent the following additional tests: complete blood count, basic biochemistry, polymerase chain reaction (PCR), variant surface glygoprotein (VSG), toxins in urine, alcohol in blood, iron study, lipid profile, ceruloplasmin, tumour markers, thyroid hormones, lead, copper, zinc, urine sediment, study of autoimmunity (antinuclear antibodies, anti-DNA, anti-SM, anti-RNP, anti-Ro, anti-La, anti-Jo1, antimitochondrial antibodies, anti-LKM, IgG antireticulin antibodies, anti-smooth muscle antibodies, parietal cells antibodies, anti-TPO (antimicrosomal) antibodies, anti-thyroglobulin (Tg) antibodies, ANCA antibodies, IgA and IgG anti-gliadin antibodies, IgG and IgM anticardiolipin antibodies), peripheral blood acanthocytes, B2-glycoprotein I, serology for HIV, HBV, HCV, syphilis, HSV-I and II, varicella zoster, CMV, EBV, Paul-Bunnell, VH6, VH8, rubella, measles, mumps, *Rickettsia conorii*, parvovirus, *Brucella*, *Borrelia*, HTLV-I and II, enterovirus, and leptospirosis, all within normal limits. A lumbar puncture was also carried out, with biochemistry, anatomical pathology and microbiology for neurotropic viruses (including PCR for EBV), culture and gram-negatives. An antistreptolysin O test (ASOT) was requested, with a titration of 439 IU/ml (below 200). The pharyngeal exudate was negative. Additionally, ECG, chest radiograph, cranial CT, brain MRI and transthoracic echocardiography were also conducted: all results were within normal limits.

While admitted, the patient was treated with 1 daily intravenous bolus of 1000 mg methylprednisolone for 3 consecutive days, and 1 intramuscular injection of 1,200,000 IU of benzathine penicillin G. The evolution during hospital stay was favourable. Home therapy with amantadine (every 24 h) and Zyprexa 2.5 mg (every 12 h) was instituted. In consultation one month after discharge, the ASOT titration had fallen to 401 IU/ml and the pharyngeal culture remained negative. At present, the patient has clearly improved from the initial symptoms, but they still remain at a more discreet level and there is evidence of a worsening during periods of stress.

As explained previously, the patient was diagnosed with the adult variant of PANDAS. Among the 5 diagnostic criteria for PANDAS are: the presence of OCD and/or tics, onset in paediatric age, episodic course of symptom severity, association with streptococcal infection (BHGAS) and association with neurological disorders, such as hyperactivity or associated, choreic-type involuntary movements. Several variants have been described, such as the dystonic, myoclonus, chronic and adult variant[1].

Until the nineties, SC was considered the only neurological sequelae associated with streptococcal infection[4]. Subsequently, the spectrum of neurological manifestations was expanded and it currently includes entities such as PANDAS, Tourette syndrome, attention deficit disorder with hyperactivity, dystonia, myoclonus, anorexia nervosa and others[5-9].

The term PANDAS, coined by Swedo[10], is an acronym that refers to a paediatric autoimmune neuropsychiatric disease associated with streptococcal infection[11]. With respect to its etiopathogenesis, it has been hypothesised that its symptoms are due to the antibodies produced as part of the immune response to suberic bishydroxamic acid (SBHA) surface proteins, which have antigenic similarities with proteins from the central nervous system[9]. We would thus face an autoimmune process affecting the basal ganglia, triggered by a streptococcal infection in genetically susceptible patients[12].

PANDAS is a mainly paediatric disease, predominant in males (2.6:1). Its onset age varies from 3 years until the beginning of puberty, although there are variants of this disorder in adulthood. The beginning of the condition is abrupt, but its resolution is usually slow and gradual over weeks, months or even longer. It may present remissions and new relapses due to new streptococcal infections once the autoimmune process has been activated by the prior infection. Neuropsychiatric symptoms include not only the tics and OCD, but others such as difficulty in reading, attention deficit disorder, hyperactivity, depression, anxiety, emotional lability, sleep disorders, eating disorders, autism, paroxysmal dyskinesias, Parkinsonism and even acute disseminated encephalomyelitis[8].

Among the treatment options, and given that we would be facing an autoimmune disease, immunosuppressive therapy is a more than reasonable option for symptom control (corticosteroids, immunoglobulin and plasmapheresis). Tonsillectomy (as in SC) has been proposed, although there have been cases like that of our patient, in which the intervention precedes the onset of neuropsychiatric symptoms[13]. Antibiotic therapy at the time of diagnosis has also been proposed, to eradicate streptococcal infection. In addition to the treatment of acute infection; although there are no controlled studies in this regard, it has also been postulated that there is a potential benefit resulting from the use of prophylactic penicillin to prevent recurrences. The therapy used to treat tics and OCD (such as selective inhibitors of serotonin reuptake, neuroleptics and cognitive-behavioural therapy[14,15]) have also been proposed as a first line of

treatment. In addition, the symptomatic efficacy of amantadine in other types of chorea[16] is also well known.

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References

1. Pavone P, Parano E, Rizzo R, Trifiletti RR. Autoimmune neuropsychiatric disorders assocated with streptococcal infection: Sydenham chorea, PANDAS, and PANDAS variants. J Child Neurol. 2006;21:727-36.
2. Segarra AR, Murphy TK. Cardiac involvement in children with PANDAS. J Am Acad Child Adolesc Psychiatry. 2008;47:603-4.
3. World Health Organization Study group. Rheumatic fever and rheumatic heart disease. Technical report series No. 764. Geneva; 1988.
4. Van Toorn R, Weyers HH, Schoeman JF. Distinguishing PANDAS from Sydenham's chorea: case report and review of the literature. Eur J Paediatr Neurol. 2004;8:211-6.
5. Sokol MS. Infection-triggered anorexia nervosa in children: clinical description of four cases. J Child Adolesc Psychopharmacol. 2000;10:33-45.
6. Di Fazio MP, Morales J, Davids R. Acute myoclonus secondary to group A beta-hemolytic streptococcus infection: a PANDAS variant. J Child Neurol. 1998;13:516-8.
7. Dale RC, Church AJ, Surtees RA. Post-streptococcal autoimmune neuropsychiatric disease presenting as paroxysmal dystonia choreathetosis. Mov Disord. 2002;17:817-20.
8. Dale RC, Church AJ, Cardoso F, Goddard E, Cox TC, Chong WK, et al. Poststreptococcal acute disseminated encephalomyelitis with basal ganglia involvement and auto-reactive antibasal ganglia antibodies. Ann Neurol. 2001;50:588-95.
9. Wolf DS, Singer HS. Pediatric movement disorders: an update. Curr Opin Neurol. 2008;21:491-6.
10. Swedo SE, Leonard HL, Garvey M, Mittleman B, Allen AJ, Perlmutter S, et al. Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections: clinical description on the first 50 cases. Am J Psychiatry. 1998;155:264-71.
11. De Oliveira SK. PANDAS: a new disease? J Pediatr (Rio J). 2007;83:201-8.
12. Lougee L, Perlmutter SJ, Nicolson R. Psychiatric disorders in fisrt-degree relatives of children with pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS). J Am Acad Child Psychiatry. 2000;39:1120-6.
13. Arostegui S, Aguero JA, Escar C. PANDAS following amygdalectomy. An Sist Sanit Navar. 2003;26:287-90.
14. Arnold PD, Richter MA. Is obsessive-complusive disorder an autoinmune disease? CMAJ. 2001;165:1353-8.
15. Snider LA, Swedo SE. Childhood-onset obsessive-compulsive disorder and tic disroders: case report and literature review. J Child Adoles Psychopharmacol. 2000;13 Suppl 1:S81-8.
16. Bonelli RM, Hofmann P. A systematic review of the treatment studies in Huntington's disease since 1990. Expert Opin Pharmacother. 2007;8:141-53.

Primary sciatic nerve lymphoma

Linfoma primario del nervio ciático

Dear Editor:

Neurolymphomatosis is a well-known complication of lymphoma that involves the infiltration of cranial nerves, spinal roots or peripheral nerves by lymphoma cells from a systemic or central nervous system (CNS) lymphoma[1]. However, a lymphoma arises in the peripheral nervous system (PNS) only infrequently. In a recent paper, Descamps et al.[2] described a patient with a primary lymphoma of the sciatic nerve and reviewed the literature. They found references of only eight patients with sciatic nerve lymphoma[3-9] and another five who had primary condition of other peripheral nerves. Since the publication of that article, we have found only one case of primary lymphoma of the sciatic nerve[10]. To these ten cases, we add a new patient who presented a mononeuropathy of the common sciatic nerve and was diagnosed with diffuse B cell lymphoma of the sciatic nerve.

This was an 89-year-old man with a history of arterial hypertension, hyperlipidemia, diabetes mellitus and a smoking habit. In March 2005, he began to feel pain in his left leg and a tingling sensation in his left toes. Subsequently, he noticed increasing difficulty in mobilising his left foot. He underwent an ultrasound of the calves and the popliteal bursa, which was normal, at another centre. Three months later, he was treated at our Neurology Clinic; the exploration showed distal paresis of the left lower extremity affecting foot dorsiflexion, 0/5; plantar flexion, 2/5; foot eversion, 0/5; foot inversion, 0/5, and extension of the first toe, 1/5. Knee flexion was preserved; ankle reflexes were abolished. Algesic and tactile sensitivities were decreased in the lateral side of the left leg and the dorsum of the homolateral foot. The left plantar cutaneous reflex was indifferent. A paresis of the left common sciatic nerve was suspected.

A blood test showed the following values: glucose, 171 mg/dl; variant surface glygoprotein (VSG), 86 mm first hour; lactate dehydrogenase (LDH), 282 IU/l. A neurophysiological study showed a disease of the internal and external popliteal nerves and signs of denervation in the muscles depending on the common sciatic nerve, with the exception of the long and short heads of the biceps femoris. A pelvic magnetic resonance imaging (MRI) was normal; an MRI of the left thigh showed a fusiform tumour of 16 x 3 cm, located in the posterior side of the thigh, depending on the sciatic nerve, hyperintense on T2 and short TI inversion recovery (STIR) sequences and that captured contrast homogeneously after gadolinium administration (Fig. 1).