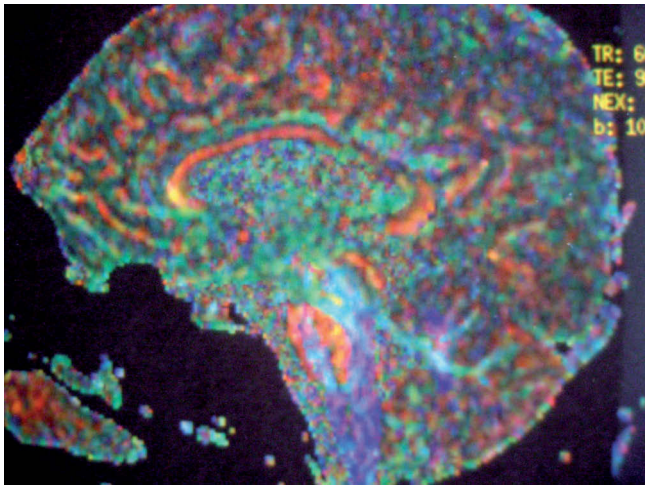

Figure 2 Diffusion tensor tractography image with absence of crossed fibres in the central portion of the corpus callosum.

(fig. 2). The genetic study revealed homozygosis in the G388A allele of gene eIF2B5 and heterozygosis in his parents. The patient has made good progress, recovering his deficits almost completely and is now able to live independently.

Vanishing white matter disease, also referred to in the literature as childhood ataxia with central hypomyelinization,[3] is one of the most prevalent hereditary alterations of the white matter in childhood.[4] It normally debuts between 2 and 6 years of age and the classic phenotype is characterized by progressive cerebellous ataxia, spasticity and mild mental deficiency. They may also present epileptic crises and optical atrophy. The symptoms characteristically worsen after mild trauma or infections with fever. There are other variants, as in this patient, with onset at later ages, even in adults, with mutations in gene eIF2B and usually with a less severe course.[4,5]

The diagnosis is confirmed through a genetic study as between 60% and 70% of patients present a mutation in gene eIF2B5.[2] The most frequent mutation is Arg113His, which is associated with the late onset of this pathology, but homozygosis for allele G388A of the gene presents a similar phenotype.[5]

Cerebral MRI is a fundamental complementary examination for diagnosis because of its characteristic findings, with alteration of virtually all the white matter, with conservation of U fibres, as in our observation. Over time, it progresses and cystic degenerations appear, as was also seen in our case. A limitation of MR arises in young children when the brains are still immature and the white matter is not fully developed with a high water content and little myelin.[4]

Diffusion tensor tractography is currently the only *in vivo* technique allowing the tracts of white matter to be analyzed. Its physical basis is anisotropic diffusion and it allows two-dimensional display and a reconstruction of the fibres in the central nervous system. Its clinical applications are varied and, in the field of demyelinizing diseases, allows quantification of plates and detection of sub-clinical lesions at early stages.[6]

For these reasons, we consider that it may be useful when applied in cases where conventional MR is not conclusive or genetics has not confirmed the diagnosis.

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Elderly patient with acquired long QT syndrome secondary to Levetiracetam

Paciente anciano con síndrome de QT largo adquirido secundario a levetiracetam

Dear Editor,

Acquired long QT syndrome (ALQTS) is an alteration of ventricular repolarization characterized by a prolonged QT interval corrected for heart rate on the electrocardiogram, that is, ≥ 460 milliseconds in women and ≥ 450 milliseconds in men.[1] ALQTS is associated with high risk, life-threatening ventricular arrhythmias, such as polymorphic ventricular tachycardia (torsade de pointes).[2] The most common causes of ALQTS are hydroelectrical alterations, anti-arrhythmia medication, antibiotics, prokinetics, psychoactive drugs and anti-histamines.[2]

We report the case of an 88-year old woman with a personal history of high blood pressure and a surgically treated fronto-temporal meningioma, currently on

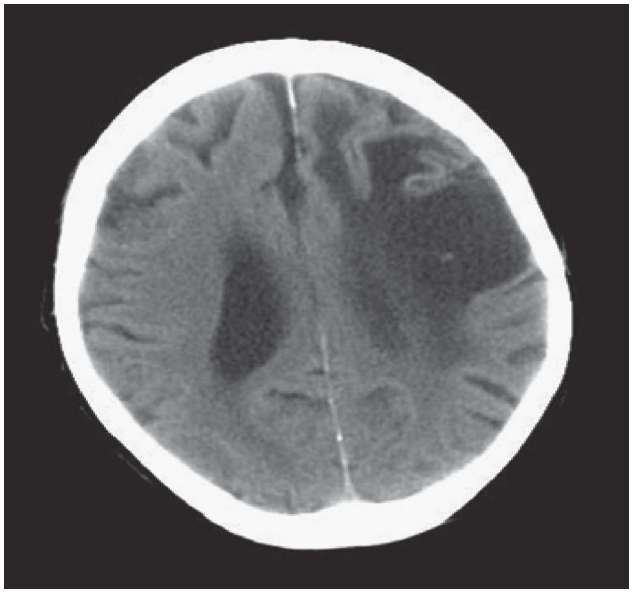

Figure 1 Cranial computerized tomography documenting the area of left fronto-temporal malacia.

treatment with acetylsalicylic acid and hydrochlorothiazide (Hidrosaluretil R), who came to the Emergency Room due to a switch-off episode accompanied by sucking mouth and facial movements lasting for one minute, with full recovery following the post-ictal status. Her vital signs and the clinical examination were normal and of note on the complementary testing were the haemogram and plasma biochemical analyses which included glycaemia, kidney, liver, and ion (calcium, sodium, magnesium, and potassium) profiles all within the normal range; and electrocardiogram (ECG) with sinus rhythm and a cranial computerized tomography (CT) with an area of left fronto-temporal malacia[3] (fig. 1). During her stay in the Emergency Room, she presented another switch-off episode with automatisms, followed by tonic-clonic movements. With a diagnosis of complex partial seizures evolving into generalized seizures,[4] treatment with levetiracetam was initiated at a dose of 500 mg IV every 12 hours. The patient was aysmptomatic 24 hours later, although the ECG documented sinus bradycardia with a heart rate of 55 bpm and a corrected QT interval (Bazett's formula) of 480 msec. An echocardiogram was ordered revealing mild tricuspid insufficiency and a 24 hour Holter documented the presence of sinus bradycardia and short spells of atrial fibrillation. In the light of these findings and without any evidence as to the cause of the QT prolongation, levetiracetam was substituted by valproic acid and the ventricular repolarization alterations were corrected within 48 hours (fig. 2).

Levetiracetam is a derivative of pyrrolidone indicated for the treatment of partial seizures with or without secondarily generalized seizures.[5] It is characterized as being efficacious and having a good safety profile, given that it is not metabolized via cytochrome P450, thereby reducing the risk of drug-drug interactions.[5,6] A previous clinical trial carried out on healthy adults reported that levetiracetam is not associated with alterations in the QT interval after a single dose.[7]

To the best of our knowledge, this case reveals the first ALQTS possibly related to levetiracetam. It is certainly true that the patient had many risk factors associated with drug-induced ALQTS, such as being elderly and female, having high blood pressure, paroxysmal atrial fibrillation, bradycardia, and prior treatment with diuretics.[8,9]

Moreover, the complexity involved in treating seizures in the elderly population is well-known, given the greater

QT interval QT interval

Figure 2 A: QT interval = 480 msec, recorded 24 hours after initiating treatment with levetiracetam. B: QT interval = 400 msec, recorded 48 hours after discontinuing treatment with levetiracetam.

likelihood of adverse events.[5] Levetiracetam is excreted largely by the kidneys and a positive correlation has been reported between creatinine clearance and drug clearance. In fact, the elderly have been shown to require a 40% dose reduction in order to achieve the same serum concentration and also to have a greater risk of side effects even at the same serum levels in comparison with young individuals.[6]

Consequently, with this case we have wanted to illustrate that before prescribing levetiracetam in elderly patients, kidney clearance must be calculated and the dose must be titrated up gradually in order to prevent adverse events;[6,10] in addition, electrocardiographic monitoring is also needed after initiating de novo treatment in patients at risk for developing QT prolongation.

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Balo's concentric sclerosis

Esclerosis concéntrica de Baló

Dear Editor,

Balo's concentric sclerosis is a demyelinating disease described for the first time by Jozsef Baló in 1928 as "periaxial concentric sclerosis".[1]

The classical clinical presentation is that of a sub-acute, fulminating, fatal encephalopathy, albeit in recent years cases have appeared in the literature with a fairly benign course, including full recovery from the disease.[2]

Historically speaking, the diagnosis has been made post mortem, by the characteristic pathological anatomy with lesions consisting of concentric rings of demyelination alternating with normal white matter. However, the most recent revisions support the role of magnetic resonance of the brain for early and definitive diagnosis, which has had a dramatic impact on the prognosis of the illness, improving the morbi-mortality it entails.

We present the case of a female patient with a sub-acute syndrome of neurological focality and a magnetic resonance image of the brain compatible with Balo's concentric sclerosis.

A twenty-seven-year-old female without any history of interest was admitted to our hospital due to a progressive course of weakness on the right side of the body, difficulty in co-ordinating the upper right limb and gait alteration for the last three months.

The general examination is normal and the neurological examination yields the following findings of interest: hemiparesis on the right side with signs of pyramidalism and spasticity of the lower limbs, accompanied by postural and kinetic tremor of the right arm. Three diffuse, rounded hypodense areas in the sub-cortical white matter with ring-shaped uptake of contrast are seen on the computerized tomography taken in the Emergency Room.

A complete magnetic resonance (MR) scan was performed of the brain and spine revealing several lesions in the supratentorial sub-cortical white matter (the largest measuring 4 cm) with an onion-like structure, displaying hypointense concentric rings in T1-weighted sequences (fig. 1) and hyperintense in T2 (fig. 2), alternating with isointense layers. After injecting paramagnetic contrast, they display ring enhancement (fig. 3). These images show no associated oedema surrounding the lesion or increased relative blood volume in the perfusion sequences. On spectroscopy, the choline spike is increased to 3.2 ppm and lactate is evidenced at 1.3 ppm (fig. 4). No infratentorial lesions are evident.

The cerebrospinal fluid (CSF) study rules out the presence of oligoclonal bands of IgG and malignant cells.

After the results of the complementary testing and with the diagnostic suspicion of Balo's concentric sclerosis-type