Letters published in earlier issues of this journal by Pato Pato et al. and Piñeiro et al. have aroused great interest and we agree with the possibility that this condition might be under-diagnosed. In this sense, we should like to describe a case with similar characteristics at our unit and we contribute our experience with diffusion tensor tractography, a new technique that might be useful in the diagnosis of this entity.

An 18-year-old male from Seville, without any history of consanguinity between his parents, suffered cranioencephalic trauma following a motorcycle accident without wearing a helmet and, in consequence, weakness in the right limbs, for which reason he was admitted to our department. His personal history of note included cranioencephalic trauma at 8 years of age which caused right hemiparesis and required admission to the intensive care unit for 10 days with total recovery on discharge. The neurological examination revealed proportional right hemiparesis (3/5) with pyramidalism and gait ataxia. The rest of the neurological and general examination was normal. The general analyses performed, including study of thyroids and lipids, were normal; cranial CT scan revealed a dilatation of the ventricular system and the magnetic resonance (MR) of the skull showed, in addition, extensive hyperintense areas in T2 of the white matter at the supratentorial level, affecting all lobes and the posterior arms of both internal capsules, compatible with demyelinization. Also noteworthy was generalized cortical-subcortical atrophy and marked thinning of the corpus callosum with cystic areas inside (fig. 1). Using diffusion tensor tractography, it was possible to observe the absence of crossed fibres in the central portion of the corpus callosum and an anomalous arrangement of the same in the genu, the cingulate gyrus and the splenium.

Figure 1 Magnetic resonance image with marked thinning of the corpus callosum with cystic areas inside (T2 sequence).

This paper was partially presented as a Poster at the 32nd Annual Meeting of the Andalusian Neurology Society.
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 hypomyelination, is one of the most prevalent hereditary alterations of the white matter in childhood. 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.

The diagnosis is confirmed through a genetic study as between 60% and 70% of patients present a mutation in gene eIF2B5. 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.

Cerebral MR 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.

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 demyelinating diseases, allows quantification of plates and detection of sub-clinical lesions at early stages.

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.

References


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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. ALQTS is associated with high risk, life-threatening ventricular arrhythmias, such as polymorphic ventricular tachycardia (torsade de pointes). The most common causes of ALQTS are hydroelectrical alterations, anti-arrhythmia medication, antibiotics, prokinetics, psychoactive drugs and anti-histamines.

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...