The gold standard for diagnosis is brain MRI, which is reported to detect even asymptomatic cases, improving outcomes.[2] It should be noted that radiological findings may not be observed in the first week after symptom onset, and the MRI scan should be repeated when there is a high level of suspicion.[6]

Early treatment with dexamethasone after rapid correction of hyponatraemia has been tested in animals, with excellent clinical results and improved prognosis, due to the capacity of dexamethasone to regulate and prevent damage to the blood—brain barrier and to decrease microglial cytokine release.[7] However, no controlled trials have been performed in humans to date.

Our case is of special relevance due to several key observations: progression was subacute, isolated hyperglycaemia was the main trigger factor, and clinical symptoms fully resolved after achieving good metabolic control in a patient with cirrhosis as trigger factor.[2] Most of the reported cases of CPM occur in the context of hyperglycaemia with concomitant ketoacidosis, abnormal sodium levels, or after treatment for hyperosmolar hyperglycaemic state; less frequently,[8–10] onset of CPM is secondary to isolated hyperglycaemia.[8,11,12] Therefore, our case would support the hypothesis that fluctuations in osmolarity and even hyperglycaemia itself may act as a trigger factor in the aetiopathogenesis of osmotic demyelination syndrome.

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Report of a case of delayed posthypoxic leukoencephalopathy: a peculiar image☆

Descripción de un caso de leucoencefalopatía hipóxica tardía: una imagen característica

Dear Editor:

Posthypoxic leukoencephalopathy is an infrequent disease, and is especially underdiagnosed in paediatric patients. It is classified as acute or delayed, according to the time interval between the ischaemic insult and the onset of clinical symptoms. The delayed form was first described by Shillito[1] in 1936 and later in 1962 by Plum et al.[2] It is characterised by clinical worsening after a period of stability, between 2 and 40 days after the initial hypoxic event.[3] This form of the disease exclusively affects the white matter, sparing the U fibres. To date, only 2 cases of paediatric patients have been reported, with ours being the youngest.[4] The acute form, known as anoxic-ischaemic encephalopathy, occurs with no significant delay and may affect the U fibres and the grey matter.[5] In both cases, it is mainly the white matter that is affected, which is contrary to expectations in an ischaemic context, since the grey matter and basal ganglia are more vulnerable to anoxia. Recent studies suggest that the white matter is more sensitive to ischaemia than previously believed.[6]

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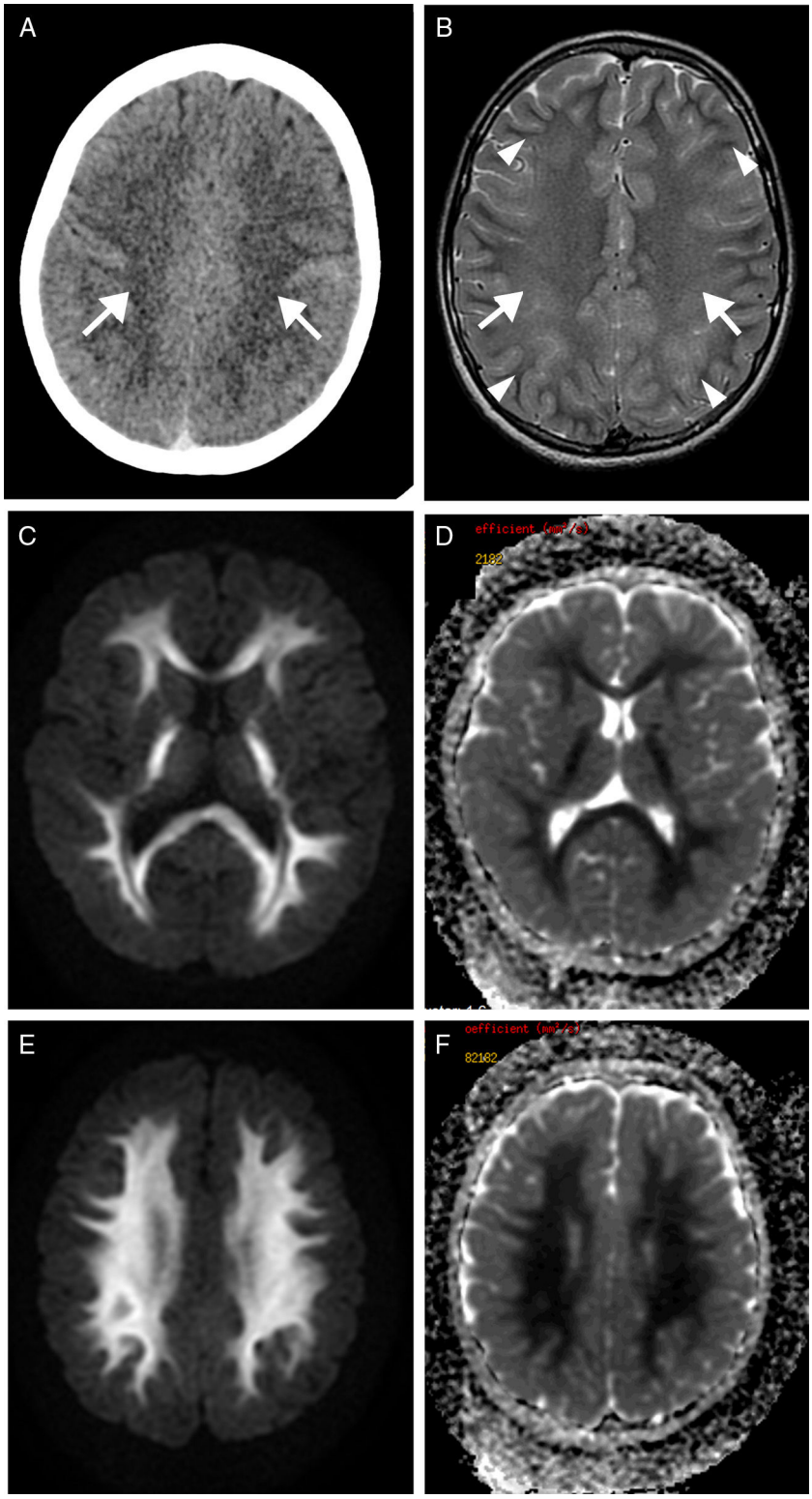

Figure 1 Transverse CT sequence (A) showing a faint hypodensity in the supratentorial white matter (arrows). Transverse T2-weighted MRI sequence (B) showing homogeneous hyperintensity of the supratentorial white matter, confluent with bilateral and symmetrical distribution (arrows), sparing the U fibres (arrowheads). Diffusion restriction is observed on the diffusion sequence (C and E) and corresponding apparent diffusion coefficient maps (D and F).

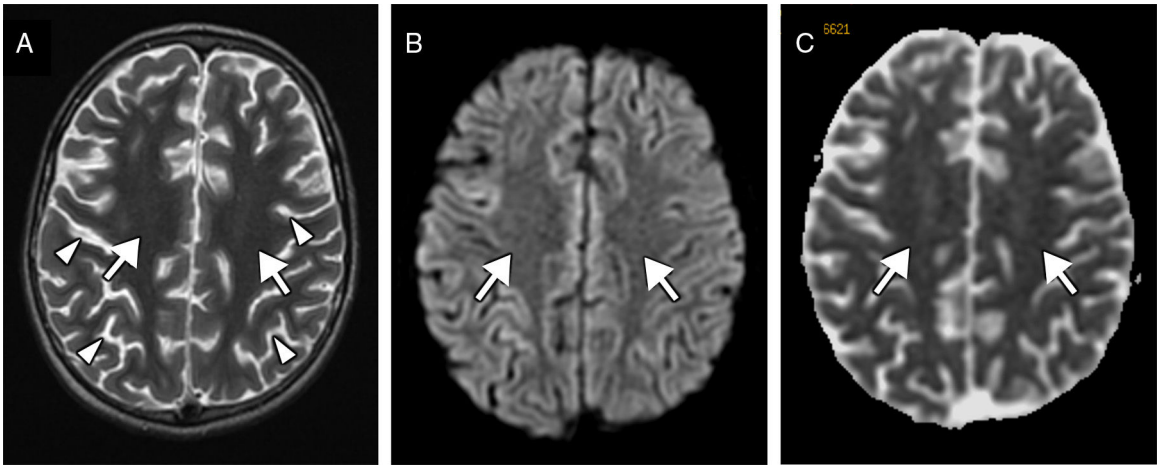

Figure 2 Follow-up MRI scan performed at 3 weeks. The transverse T2-weighted sequence (A) reveals normalised signal intensity in the white matter (arrows) and moderate loss of volume with more prominent sulci (arrowheads). Diffusion restriction (arrows) is no longer observed on the diffusion sequence (B) and corresponding apparent diffusion coefficient maps (C).

We present the case of a 9-year-old boy with a history of severe obstructive hypertrophic cardiomyopathy who was attended at the emergency department following a cardiorespiratory arrest lasting 5 minutes. Seven days after admission, he presented a new episode of haemodynamic instability due to ventricular fibrillation, with recovery after 4 minutes of resuscitation. Nine days after the initial event, the patient's level of consciousness decreased; a CT scan revealed a faint generalised hypodensity in the supratentorial white matter (Fig. 1A). An MRI study performed at 24 hours showed altered signal intensity in the supratentorial white matter, sparing the U fibres; the hyperintensity was symmetrical, extensive, confluent, and homogeneous on the T2-weighted sequence (Fig. 1B), displaying diffusion restriction (Fig. 1C and E) and no contrast enhancement. A follow-up MRI performed at 3 weeks revealed normal signal intensity (Fig. 2A) and no diffusion restriction (Fig. 2B and C) in the white matter, with a moderately reduced volume and more prominent sulci (Fig. 2A, arrowheads).

In this clinical context, the imaging findings are characteristic of delayed posthypoxic leukoencephalopathy. On the MRI study, the white matter lesion is supratentorial, sparing the short association fibres; the cortex and basal ganglia are unaffected. Diffusion is typically restricted due to intramyelinic oedema, and the lesion presents no contrast uptake. Differential diagnosis using MRI is very narrow due to the limited number of diseases exclusively affecting the white matter with diffusion restriction; these include: toxic leukoencephalopathy (due to such drugs of abuse as heroin or such medications as vigabatrin, methotrexate, and carmofur), metabolic leukoencephalopathy (phenylketonuria and extrapontine myelinolysis), inflammatory diseases (multiple sclerosis and acute disseminated encephalomyelitis), status epilepticus, post-traumatic diffuse axonal injury, and fat embolism. The clinical symptoms and characteristic distribution pattern of the injury help establish diagnosis in most cases.

The precise pathophysiological mechanism remains unknown, although 2 hypotheses have been proposed; the first is related to the replacement of some myelin-related proteins halting after ischaemia, which would explain the delay in disease onset but not the low incidence[3]; the second hypothesis is related to a decrease in arylsulphatase A levels,[7] although patients with normal levels have been reported.[8] In terms of histopathology, the lesion consists of diffuse demyelination with axonal integrity and presence of active macrophages and astrocytes. U fibres and the cortex are typically preserved.[2,7]

Symptoms tend to remit spontaneously. However, the natural progression remains unknown due to the condition's low incidence and the lack of follow-up in most of the published cases.[9]

In the appropriate clinical scenario, exclusive involvement of the supratentorial white matter, symmetrically and bilaterally with diffusion restriction, is highly suggestive of delayed posthypoxic leukoencephalopathy. Recognising its characteristic clinical progression and imaging pattern helps in diagnosis and enables ideal management.

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Temporal lobe encephalocele, a subtle structural lesion that can be associated with temporal lobe epilepsy☆

Encefalocele temporal, una lesión estructural sutil que puede asociarse con epilepsia temporal

Dear Editor,

Anterior temporal lobe encephalocele, or temporal pole encephalocele, has recently been associated with refractory temporal lobe epilepsy.[1,2] Temporal lobe encephalocele may be congenital or acquired and can occur in any supratentorial region of the cranium, although the anterior region of the middle cranial fossa is most frequently affected.[3] These are subtle lesions, and high-field MRI including specific sequences is essential for diagnosis.

We present the case of a 49-year-old man presenting episodes of disconnection from the environment, incoherent speech, and oral automatism, lasting 1-2 minutes. After the episodes, he remained disoriented for 30 minutes, with repetition of questions. In the neurological examination he presented a good level of consciousness and was oriented in time, space, and person. He showed no memory or language alterations. Examination of the cranial nerves only revealed mild facial droop. No other remarkable findings were observed in the neurological examination.

Results from a head CT scan were considered normal and 1.5T MRI revealed no alterations.

An electroencephalogram showed very mild abnormalities in the left temporal region, consisting of bursts of slow delta and theta waves that, although diffuse, presented increased amplitude in this region; and mild background slowing with hyperventilation, which was more evident in the left temporal region. No associated epileptiform anomalies were observed.

Treatment with carbamazepine was started, and seizures were controlled. At 5 months of follow-up, the patient's family reported memory failures that did not interfere with his daily activities.

Six years later, the patient presented an episode of disconnection, and we performed a 3T MRI scan with an epilepsy protocol including high-resolution T2-weighted coronal and axial sequences. The study revealed a small left temporal encephalocele with herniation of the brain parenchyma through a bone defect in the left temporal pole (Fig. 1).

Detecting structural alterations in the brain MRI study improves the chances of controlling seizures after surgery in patients with refractory focal epilepsy.[1,2] However, a significant percentage of patients do respond to pharmacological treatment,[1] as did our patient.

It should be noted that in 20%-30% of patients with normal MRI findings, subtle lesions may go undetected if analysis of the images is not guided by the clinical context.[1]

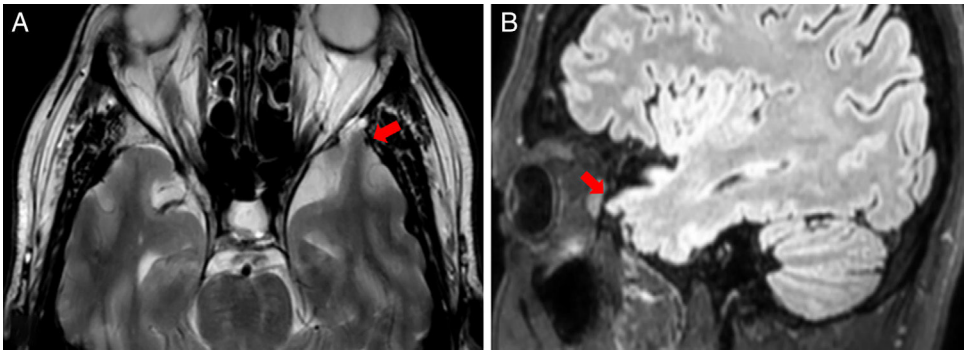

Figure 1 MRI scan: axial T2-weighted (A) and sagittal T2-weighted FLAIR sequences (B), showing a small herniation of the brain parenchyma in the anterior pole of the left temporal lobe through an anterior bone defect on the base of the cranium; this finding suggests encephalocele.

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