Deafferentiated neurons would be stimulable in this case by hypertension in the cerebral blood vessels corresponding to these cortical areas; with administration of a hypotensive treatment and monitoring of the hypertensive crisis, stimuli and trigger factors disappeared, the episode resolved, and the deafferentiated cortex stabilised.

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Non-motor neurological symptoms in patients with amyotrophic lateral sclerosis☆

Síntomas neurológicos extra-motores en pacientes con esclerosis lateral amiotrófica

Dear Editor:

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease traditionally defined as clinical symptoms involving the upper motor neuron, in the motor homunculus, and the lower motor neuron, in the spinal cord.[1,2] However, recent publications have described non-motor signs and symptoms in patients with definite ALS, including impaired higher cerebral functions, signs of dysautonomia, metabolic disorders,[3,4] and cognitive alterations linked to frontotemporal dementia.[5] Post mortem histopathological studies of patients with definite sporadic ALS have identified protein inclusions of transactive response DNA binding protein 43 kDa (TDP-43) in non-motor areas of the central nervous system (CNS), including the nigrostriatal system, cerebellum, forebrain, hypothalamus, and neocortical and allocortical areas.[6,7] ALS and frontotemporal dementia are currently considered to be TDP-43 proteinopathies.

We performed a retrospective study of the medical records of 112 patients diagnosed with definite sporadic ALS according to the El Escorial clinical and neurophysiological criteria.[8] We analysed the non-motor neurological signs and symptoms reported at the time of diagnosis and those manifesting in the first year of clinical follow-up. All patients with diagnosis of definite ALS were assessed with the revised ALS functional rating scale (ALSFRS-r), the Mini-Mental State Examination, a neuropsychological test, a genetic test, and the Hamilton Anxiety and Depression Scale during the baseline consultation, and underwent at least one additional evaluation during the one-year follow-up period. In our sample, we identified 25 non-motor symptoms, with the most prevalent being: depression (in 47% of patients), pain (26%), fatigue (24%), anxiety (18%), pseudobulbar symptoms (13%), and paraesthesias (13%) (Table 1). No diagnosis of frontotemporal dementia was established in any of our patients; curiously, all cases with pseudobul-

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Table 1 Non-motor symptoms of ALS, classified by domain, frequency, and ratio.

Symptom	Frequency[a]	Ratio[b]
Cardiovascular		
Oedema	5	4
Orthostatic hypotension	1	1
Somnolence/fatigue		
Fatigue	27	24
Restless legs	1	1
Insomnia	1	1
Daytime somnolence	1	1
Mood/cognition		
Depression	53	47
Anxiety	20	18
Anhedonia	1	1
Cognitive impairment	1	1
Perceptual alterations/hallucinations		
Pseudobulbar symptoms	14	13
Blurred vision	1	1
Gastrointestinal tract		
Nausea	2	2
Vomiting	1	1
Bowel incontinence	1	1
Constipation	8	7
Urinary		
Urinary problems	2	2
Miscellaneous		
Pain	29	26
Paraesthesias	14	13
Diaphoresis	5	4
Seborrhoea	1	1
Weight gain	1	1

[a] Frequency of symptom in the ALS patient total.
[b] Ratio: percentage of symptom in patient total.

bar symptoms, depression, or anxiety were negative for the chromosome 9 hexanucleotide repeat expansion normally associated with pseudobulbar symptoms (*C9Orf72* gene). We identified an average of 4.2 ± 2.03 non-motor symptoms per patient. Patients with bulbar ALS presented a higher number of non-motor symptoms than patients with spinal ALS (5.41 ± 1.61 and 3.85 ± 2.01 non-motor symptoms per patient, respectively). Furthermore, we observed correlations between symptom pairs which may have similar pathophysiological mechanisms, including: anxiety and depression (χ^2; $P = .025$), bladder incontinence and constipation (χ^2; $P = .017$), and pain and paraesthesias (χ^2; $P = .027$).

A recent study by Cykowski et al.[7] detected TDP-43 inclusions in the forebrain and hypothalamus, reporting a significant correlation between this histopathological finding and predominantly bulbar ALS. In our sample, patients with predominantly bulbar ALS presented more non-motor symptoms than patients with spinal ALS.

Clinical heterogeneity of ALS is characteristic[9]; the presence of pathological inclusions in non-motor areas of the CNS in cases of definite sporadic ALS may explain the presence of atypical symptoms, which are frequently omitted and therefore are not approached or treated adequately, limiting quality of life. In our sample, the non-motor symptoms observed in patients with ALS may be incidental manifestations due to disability, or clinical manifestations due to the progression of the neurodegenerative process. Timely identification of non-motor manifestations in patients with classic motor symptoms may facilitate medical treatment and improve the clinical condition of these patients. The wide variety of clinical phenotypes, the recent histopathological findings in the CNS, and the presence of non-motor symptoms suggest that ALS is a multisystemic disease which is not restricted to the motor system.

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Subdural intracranial and spinal haematoma secondary to neuraxial anaesthesia☆

Hematoma subdural intracraneal y espinal secundario a anestesia neuroaxial

Dear Editor,

The actual incidence of complications secondary to neuraxial anaesthesia (NA) is unknown, with postdural puncture headache being the most frequent example.[1,2] Intracranial subdural haematoma (SDH) is estimated to affect one in 500 000 to 1 000 000 patients undergoing lumbar puncture[3] and spinal haematoma is estimated to affect one in 190 000 undergoing epidural anaesthesia and one in 320 000 undergoing spinal anaesthesia.[1] Both complications have been associated with considerable levels of morbidity and mortality[3–6]; every effort should therefore be made to ensure early diagnosis and treatment.)

Clinical case

Our patient was a 56-year-old man with no relevant clinical history. Twenty-four hours after undergoing prostate surgery under spinal anaesthesia, he presented intense occipital headache radiating to the neck, triggered by standing. Examination revealed no focal neurological signs. The patient was diagnosed with intracranial hypotension and started on intravenous corticosteroids and nonsteroidal anti-inflammatory drugs. One month after onset, the patient attended our department due to the persistence of symptoms. For 15 days he had experienced constant, pulsatile pain; the intensity had decreased, but pain now also occurred in the supine position, increasing with Valsalva manoeuvres; he had also begun experiencing intense neck pain. A brain MRI scan was performed, revealing an extra-axial collection in the left frontoparietal region, with a maximum thickness of 1 cm. It appeared hyperintense on proton density-weighted sequences (Fig. 1 and isointense on T2-weighted sequences. A neck and chest MRI scan showed

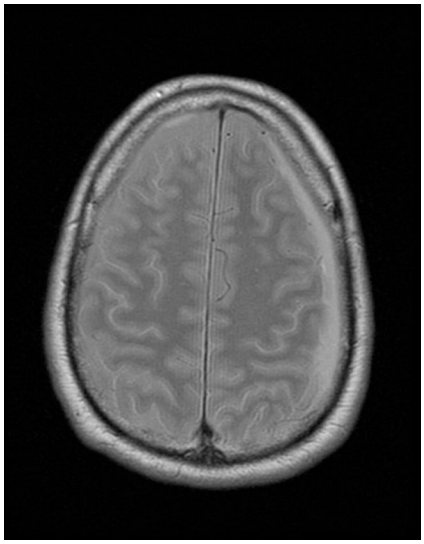

Figure 1 Axial proton diffusion-weighted brain MR image displaying a hyperintense left frontoparietal extra-axial collection with a maximum thickness of 1 cm.

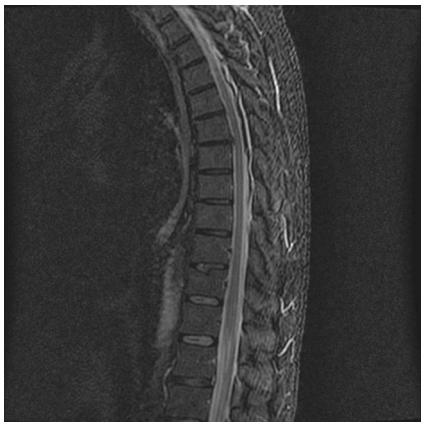

Figure 2 Sagittal cervicothoracic STIR sequence displaying a posterior subdural space-occupying lesion from C7 to T10-T11, of heterogeneous intensity.

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