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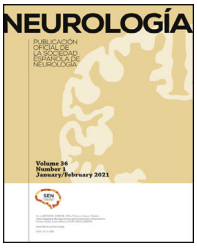

LETTER TO THE EDITOR

Clinical guide for the diagnosis and follow-up of myotonic dystrophy type 1, MD1 or Steinert's disease: Sleepiness and role of Epworth Sleepiness Scale☆

Guía clínica para el diagnóstico y seguimiento de la distrofia miotónica tipo 1, DM1 o enfermedad de Steinert: somnolencia y papel de la escala de somnolencia de Epworth

Dear Editor:

I read with great interest the study by Gutiérrez Gutiérrez et al.[1] "Clinical guide for the diagnosis and follow-up of myotonic dystrophy type 1, MD1 or Steinert's disease." The authors propose these clinical guidelines as a reference for healthcare professionals involved in the diagnosis and follow-up of MD1. Although this topic is difficult to analyse in depth, the authors underscore the presence of sleep disorders and somnolence, together with fatigue, in MD1. They assert that fatigue is associated not with muscle weakness but with sleep disorders and somnolence. Furthermore, these guidelines consider the Epworth Sleepiness Scale (ESS) "a good instrument for diagnosing" sleep disorders, citing the validation study by Johns.[2] In their Table 1, they also recommend the use of the ESS to diagnose somnolence and fatigue. These statements are only partially correct.[1] On the one hand, the third edition of the International Classification of Sleep Disorders[3] defines daytime sleepiness as "the inability to stay alert and awake during the major waking episodes of the day, resulting in unintended lapses into sleep." On the other hand, fatigue is defined as "reversible motor and cognitive impairment, with reduced motivation and desire to rest."[4] Although these 2 concepts are very different, the terms may be used interchangeably in everyday speech, as well as by patients and physicians. Fatigue and excessive daytime sleepiness (EDS) manifest in 62.5% and 30%-39% of the patients with MD1, respectively.[5] Despite the high prevalence of sleep apnoea syndrome (SAS),[5—8] several authors have been unable to show a correlation between EDS and SAS. From a methodological perspective, EDS in neurological diseases has been analysed using different measurement tools. The most frequent subjective scale is the ESS, a reliable questionnaire used in several surveys. Several factors have contributed to the extensive use of the ESS in assessing somnolence: the questionnaire is self-administered and easy to complete and score, making it suitable for quickly assessing EDS in outpatient clinics. It is currently thought to be the most widely used scale for assessing/screening for somnolence and has been translated and validated in several languages and countries. However, the ESS is not applicable to all clinical contexts. Somnolence and fatigue are strongly associated with MD1, and the ESS requires the performance of some activities that patients with MD1 do not frequently perform. Therefore, different tools have been proposed to assess somnolence and fatigue in MD1. Laberge et al.[9] validated the Daytime Sleepiness Scale (DSS), a self-administered scale more in line with the clinical characteristics observed in MD1 patients with somnolence (ie, naps, narcolepsy-like phenotype, and somnolence related to attention). Furthermore, the fatigue severity scale (FSS) has been considered the most important tool for assessing fatigue in MD1.[10] Very recently, Laberge et al.[11] assessed EDS and fatigue in a large, 9-year prospective study of MD1 using the DSS and FSS, and reported that high body mass index was a potential risk factor for both symptoms. In an exhaustive evaluation of both somnolence and fatigue in MD1, Hermans et al.[12] developed the Fatigue and Daytime Sleepiness Scale (FDSS) with the Rasch model, using the 22 items of the ESS, DSS, and FSS to obtain a scale of 12 items after rescoring and removing misfitting items. This scale showed good reliability and internal consistency and was able to distinguish between patients with MD1 and fatigue and those with EDS.[13] Therefore, we should underscore that the ESS does not represent a "good instrument for diagnosing" sleep disorders and is not the best subjective scale for assessing somnolence, with the DSS and FDSS being more appropriate tools for assessing the clinical status of patients with MD1, and more useful for distinguishing fatigue from EDS in this context.[14]

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Ischaemic stroke secondary to paradoxical embolism as a consequence of superior vena cava syndrome by a displacement of a ventriculoatrial shunt*

Infarto cerebral por embolismo paradójico secundario a síndrome de vena cava superior por malposición de un catéter de derivación ventriculoauricular

Dear Editor:

Superior vena cava syndrome (SVCS) is an infrequent condition characterised by a partial or total obstruction of blood flow through the superior vena cava due to extrinsic compression, infiltration, or thrombosis. Progression is variable and sometimes slow, and the condition can even be life-threatening; therefore, it requires a precise diagnosis and early treatment.[1,2]

We present the case of an 82-year-old woman, who was partially dependent and was using a colostomy bag due to perforated diverticulitis. In 2013, she underwent radiosurgery for a petroclival meningioma measuring 4 cm. She later developed non-communicating hydrocephalus due to external compression of the meningioma. As ventriculoperitoneal shunt was contraindicated due to the history of colostomy, she underwent ventriculoatrial shunt (VAS) implantation in December 2014 (Fig. 1).

In May 2019, the patient consulted our department due to global aphasia and right hemiparesis (muscle strength of 2/5), with onset upon waking. Given suspicion of stroke, we performed a multimodal CT scan, which revealed distal occlusion of the M1 segment of the left middle cerebral artery and favourable mismatch. The patient underwent mechanical thrombectomy, with angiography showing complete reperfusion. Despite this, neurological symptoms improved only slightly. A follow-up CT scan performed 24 hours after the procedure revealed an ischaemic lesion involving the left lentiform nucleus and insula.

During her stay at the stroke unit, the patient presented oedema in the right arm, hindering the insertion of peripheral venous catheters. On the fifth day after admission, we also observed oedema in the face and contralateral arm; examination of the upper limbs yielded normal results. Given the suspicion of SVCS, we performed a Doppler ultrasound of the supra-aortic trunks, which revealed thrombosis of the internal jugular veins. We also requested a non-contrast chest, abdomen, and pelvis CT scan, which initially ruled out extrinsic venous compression or tumour and

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