

Figure 1 Liver biopsy. Central lobular confluent necrosis with inflammatory infiltration of histiocytes. No signs of fibrosis or chronic hepatocellular disease were seen. These findings were compatible with toxic hepatitis (haematoxylin and eosin $\times 20$).

first months of treatment, liver damage may also occur at later stages.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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Combined central and peripheral demyelination: A case description^{☆,☆☆}



Descripción de un caso de *Combined central and peripheral demyelination*

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Dear Editor:

The medical literature includes case reports and series of patients with chronic inflammatory demyelinating polyneuropathy (CIDP) plus central nervous system (CNS) demyelination as well as patients with multiple sclerosis (MS) associated with demyelinating neuropathy.^{1–3} Onset of central and peripheral nervous system symptoms is usually acute in paediatric patients.⁴ In recent years, the term “combined central and peripheral demyelination” (CCPD) has been proposed to describe the occurrence of demyelination in both these locations.⁵ However, there is still no formal definition for the condition, and its aetiopathogenic mechanisms are yet to be established.

Among the different phenotypes of CIDP, Lewis-Sumner syndrome, also known as multifocal acquired demyelinating

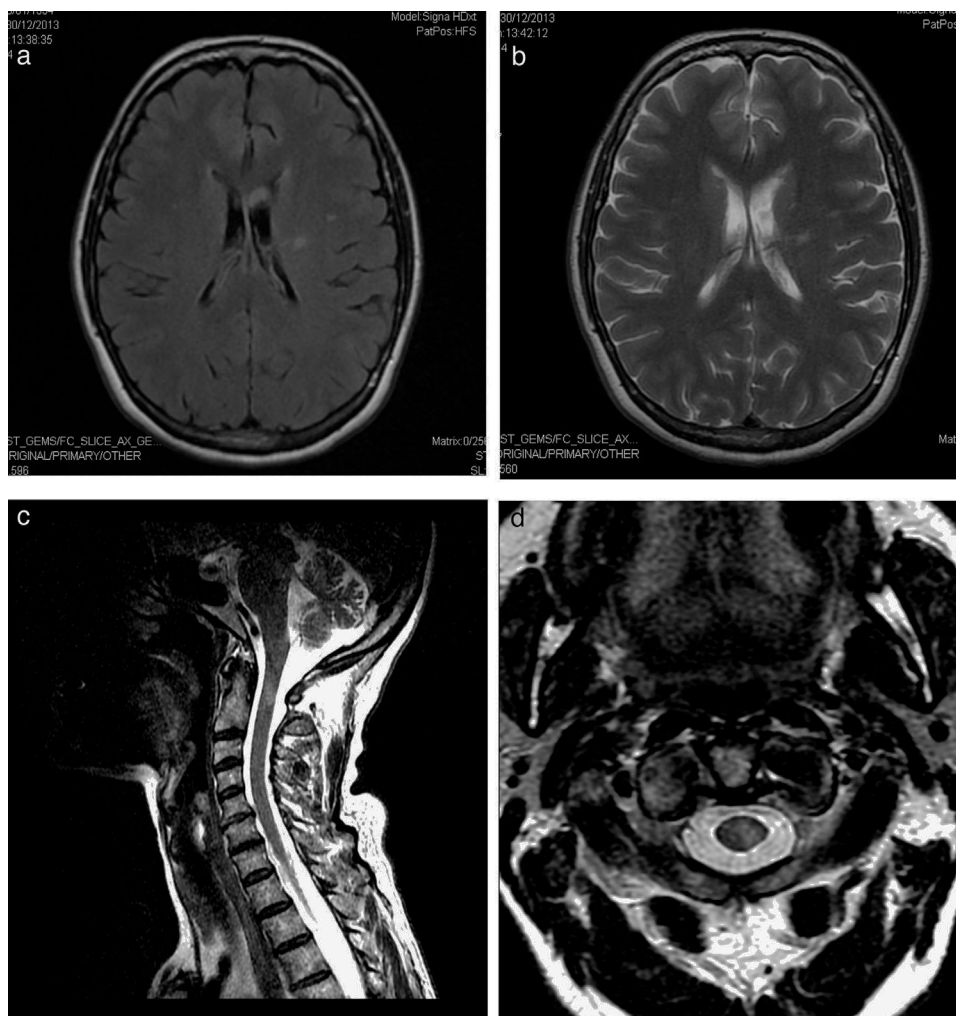


Figure 1 (a) FLAIR sequence showing signal hyperintensity in the left periventricular white matter extending towards the corona radiata and left centrum semiovale. (b) T2-weighted sequence displaying an area of probable malacia-gliosis. (c) Sagittal and (d) axial T2-weighted sequences showing a hyperintense lesion measuring 5 mm × 4 mm × 1.3 mm in the cervical spinal cord, at the level of the middle third of the odontoid process.

sensory and motor neuropathy (MADSAM), is characterised by asymmetry, predominantly affecting the upper limbs. We have found no association between this form and CNS demyelination. We describe the case of a patient diagnosed with a form of CIDP (Lewis-Sumner syndrome or MADSAM) and displaying CNS demyelination, a finding which conditions diagnosis and management.

Our patient was a 58-year-old woman with a history of Hashimoto thyroiditis, who was admitted to another hospital due to subacute onset of sensory alterations in her right hand and extending to the other limbs. A neurophysiological study revealed mixed neuropathy which was asymmetrical and predominantly motor. Our patient was diagnosed with multiple neuritis; symptoms improved with administration of immunoglobulins (IG). She came to our department due to a one-month history of dysaesthesia in her right hand, extending to the left hand, the distal region of her left foot, and the left side of her neck and rib cage. The examination revealed 4+/5 muscle balance in the right interossei muscles, 4/5 in the left interossei muscles and left extensor

digitorum muscle, 4+/5 in the left tibialis anterior and gastrocnemius muscles, 5–/5 in the right tibialis anterior and gastrocnemius muscles, and abolished stretch reflexes; no sensory alterations were found. Our patient had normal gait and displayed instability during the Romberg test. A complete blood count and serology and autoimmune tests revealed no abnormal findings except for elevated TPO-Ab levels (5631 IU/mL). A lumbar puncture disclosed no cells and a protein level of 97 mg/dL. A nerve conduction study revealed proximal and distal sensorimotor demyelinating polyneuropathy predominantly affecting the upper limbs, particularly the left upper limb (asymmetrical). A brain MRI scan displayed 2 hyperintense areas on T2-weighted and FLAIR sequences (Fig. 1a and b). Our patient improved after the first cycle of IG. During follow-up, she displayed distal weakness, ataxic gait, and tremor in her hands, especially in the left hand (Fig. 2). A T2-weighted MRI scan of the brachial and lumbosacral plexi revealed no abnormalities except for a hyperintense area in the spinal cord (Fig. 1C and D). Visual evoked potentials (VEP)

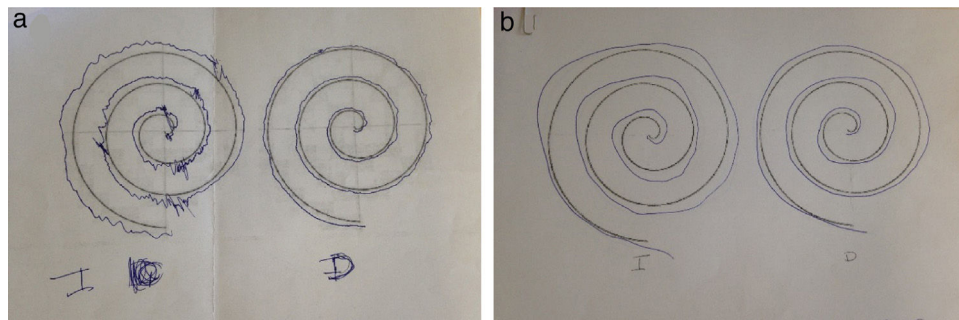


Figure 2 Action tremor, which was more pronounced in the left hand (a). Tremor decreased after treatment with IG and low doses of propranolol (b).

showed prolonged P100 latency in the left eye. Our patient tested negative for neurofascin-155 antibodies. Symptoms improved after several cycles of IG and propranolol (Fig. 2).

Our patient was diagnosed with Lewis-Sumner syndrome, initially manifesting with relapses. Given the published evidence of the association between MADSAM and brachial plexus signal hyperintensity and/or hypertrophy,⁵ we requested an MRI scan of the brachial and lumbosacral plexi; this revealed a hyperintense area in the spinal cord, which we interpreted as being inflammatory in nature. To our knowledge, the literature reports no association between this form of CIDP and CNS demyelinating lesions, but it does describe cases of similar cervical lesions in patients with extensive parenchymal lesions, such as ADEM,⁶ associated with demyelinating polyneuropathy in the form of CIDP or acute inflammatory demyelinating polyradiculoneuropathy.⁷

Co-presence of central and peripheral nervous system demyelination was first described by Amit et al.⁸ in 1992 in a paediatric patient. These authors used the term ‘‘CCPD’’, although several other designations have been proposed, including CIDP with CNS involvement, peripheral neuropathy with MS, and relapsing-remitting disease of the central and peripheral nervous systems.⁹ Kawamura et al.¹⁰ identified neurofascin antibodies in patients with CCPD and hypothesised that central and peripheral demyelination may have common pathophysiological mechanisms. Ogata et al.¹¹ published a study of 40 patients with CCPD, the largest series analysed to date. These authors described the demographic, laboratory, neuroimaging, and VEP features of this condition, and established 2 subtypes, depending on whether symptoms in these 2 locations appeared simultaneously (within 2 months of one another) or at different times throughout disease progression (>2 months). The first subtype (peripheral and central nervous system symptoms appear simultaneously) is associated with severe impairment, respiratory disorders, and more extensive cerebral and spinal cord lesions. On the other hand, the second subtype (symptoms manifest at different stages of the disease, even years apart) is usually associated with optic nerve involvement. Our patient met the criteria proposed by Ogata et al.¹¹ for the second subtype: asynchronous onset of central and peripheral nervous system symptoms and impaired visual evoked potentials. Tremor has been described in patients with CIDP,¹² but

to our knowledge no studies have linked it to CCPD or MADSAM.

In conclusion, the course of CCPD differs from that of demyelination affecting a single location. Therefore, correctly diagnosing CCPD may guide future immunomodulatory treatment for further relapses.

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Hospital stroke registers: Similarities and differences[☆]



Registros hospitalarios de ictus: similitudes y diferencias

Dear Editor:

In a recent article, Palomeras et al.¹ presented the results of the stroke registry of Mataró hospital in the province of Barcelona. We are pleased with the publication of a new hospital stroke registry, as analysing stroke registry data provides invaluable up-to-date information on the natural history of cerebrovascular diseases, particularly in relation to basic demographic, clinical, and developmental variables.

If we compare clinical data from the Mataró stroke registry ($n=2165$) with those from the 2 hospital stroke registries published previously in our setting (the 1984 registry from Hospital Sant Pau in Barcelona [$n=1044$]² and the 1993 registry from Hospital de la Alianza-Sagrat Cor in Barcelona [$n=1000$]³), we can observe that the most frequent cerebrovascular risk factors are the same in all 3 registries (arterial hypertension, cardiac arrhythmia, dyslipidaemia, and diabetes mellitus). However, mean age in the Mataró stroke registry was higher (73 years), and only the Mataró and Sagrat Cor registries describe and analyse the different subtypes of stroke (ischaemic and haemorrhagic). It is noteworthy that the Mataró stroke registry is the only one to include 3-month follow-up data,¹ while the data in the other 2 registries only covers the acute phase of stroke (data recorded during patients' hospital stay).^{2,3}

The higher mean age seen in the Mataró stroke registry is consistent with observations from daily clinical practice. This observation is further confirmed by the growing frequency of stroke in the very elderly population (aged 85 or older).⁴

Another of the most important novel features of the Mataró stroke registry is the inclusion of variables on follow-up after hospital discharge. The greater complexity and effort involved in data collection as a result of this

are compensated for by a substantial improvement in the scientific quality of registry data.

In summary, the 3 databases cited above display similarities in the frequency of cardiovascular risk factors. The differences observed between these registries unambiguously reflect a tendency towards greater prevalence of stroke at more advanced ages. Also heartening is the evidence the Mataró stroke registry provides of the clear improvements in the quality of data collection methods. This will result in better, more objective knowledge about certain basic clinical aspects of cerebrovascular diseases.⁵

Conflicts of interest

The authors have no conflicts of interest to declare.

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