References

1. Goldstein DJ, Oz MC, Rose EA. Implantable left ventricular assist devices. N Engl J Med. 1998;339:1522—33.
2. Mancini D, Colombo PC. Left ventricular assist devices: a rapidly evolving alternative to transplant. J Am Coll Cardiol. 2015;65:2542—55.
3. Slaughter MS, Rogers JG, Milano CA, Russell SD, Conte JV, Feldman D, et al. Advanced heart failure treated with continuous-flow left ventricular assist device. N Engl J Med. 2009;361:2241—51.
4. Lazar RM, Shapiro PA, Jaski BE, Parides MK, Bourge RC, Watson JT, et al. Neurological events during long-term mechanical circulatory support for heart failure: The Randomized Evaluation of Mechanical Assistance for the Treatment of Congestive Heart Failure (REMATCH) experience. Circulation. 2004;109:2423—7.
5. Willey JZ, Demmer RT, Takayama H, Colombo PC, Lazar RM. Cerebrovascular disease in the era of left ventricular assist devices with continuous flow: risk factors, diagnosis, and treatment. J Heart Lung Transplant. 2014;33:878—87.
6. Alnaami I, Buchholz H, Ashforth R, Yeo T, Kotylak T, Alaklabi M, et al. Successful use of Solitaire FR for stroke in a pediatric ventricular assist device patient. Ann Thorac Surg. 2013;96:e65—7.
7. Rhee E, Hurst R, Pukenas B, Ichord R, Cahill AM, Rossano J, et al. Mechanical embolectomy for ischemic stroke in a pediatric ventricular assist device patient. Pediatr Transplant. 2014;18:E88—92.
8. From RP, Hasan D, Froehler MT, Goerbig-Campbell JL. Stroke and left ventricular assist device (LVAD). OJAnes. 2013;3:51—6.
9. Al-Mufti F, Bauerschmidt A, Claassen J, Meyers PM, Colombo PC, Willey JZ. Neuroendovascular interventions for acute ischemic strokes in patients supported with left ventricular assist devices: a single-center case series and review of the literature. World Neurosurg. 2016;88:199—204.
10. Nishijima Y, Akamatsu Y, Weinstein PR, Liu J. Collaterals: implications in cerebral ischemic diseases and therapeutic interventions. Brain Res. 2015;1623:18—29.
11. Rusanen H, Saarinen JT, Sillanpää N. The association of blood pressure and collateral circulation in hyperacute ischemic stroke patients treated with intravenous thrombolysis. Cerebrovasc Dis. 2015;39:130—7.
12. Jauch EC, Saver JL, Adams HP, Bruno A, Connors JJB, Demaerschalk BM, et al. Guidelines for the early management of patients with acute ischemic stroke: a guideline for healthcare professionals from the American Heart Association/American Stroke Association. Stroke. 2013;44:870—947.

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Muscle atrophy and fasciculations as a manifestation of sporadic Creutzfeldt-Jakob disease: a case report☆

Amiotrofia y fasciculaciones como forma de presentación de la enfermedad de Creutzfeldt-Jakob esporádica. A propósito de un caso

Dear Editor,

Sporadic Creutzfeldt-Jakob disease (CJD) is the most frequent form of presentation of prion diseases. Although the clinical characteristics of this condition are well known,[1,2] lower motor neuron involvement is a rare form of presentation, especially in the early stages of the disease.[3]

Our patient was a healthy 54-year-old man with a 4-month history of poor coordination and gait instability. The neurological examination detected fasciculations in the deltoid, biceps, triceps, and quadriceps muscles, associated with amyotrophy and mild weakness of the proximal muscles of his limbs, especially the quadriceps, deltoid, and periscapular muscles. The patient also displayed global areflexia with mild dysmetria in all limbs, predominantly affecting the left side; orthostatic tremor; and ataxic gait with inability to walk in tandem. He showed no signs of upper motor neuron involvement.

A brain MRI scan revealed cortical alterations on diffusion-weighted sequences only (Fig. 1). Laboratory tests included a complete blood count; a biochemical study; kidney, liver, and thyroid function tests; and vitamin B_{12} and B_9 determination. Results were normal. The tests for infection, paraneoplastic antibodies, tumour markers, and markers of autoimmunity yielded negative results. An electroencephalography revealed mild, diffuse disorganisation of background activity; electromyography showed extensive denervation in the form of fibrillations, positive waves, and fasciculations in all muscles explored. The nerve conduction study revealed no abnormalities. A neuropsychological study revealed mild visuospatial alterations, short-term memory deficits, and reduced verbal fluency; these findings are compatible with cortico-subcortical cognitive impairment. CSF analysis yielded positive results for 14-3-3 protein and

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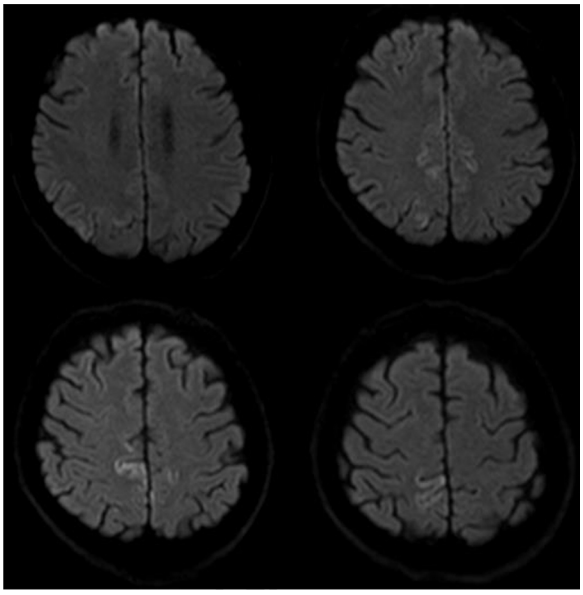

Figure 1 MRI scan at admission. Axial diffusion-weighted sequence showing abnormal cortical diffusion restriction in the precuneus bilaterally (particularly on the right) and in the right superior parietal lobule at the parasagittal level.

elevated tau protein levels. A brain MRI scan performed one month later revealed additional signal alterations in the cortex in diffusion-weighted sequences (Fig. 2). The patient's clinical status progressed to severe cognitive impairment and disabling ataxia; fasciculations were visible in distal muscles of the limbs, with no involvement of face or tongue muscles. In the final stages, he developed akinetic mutism, and died 15 months after diagnosis.

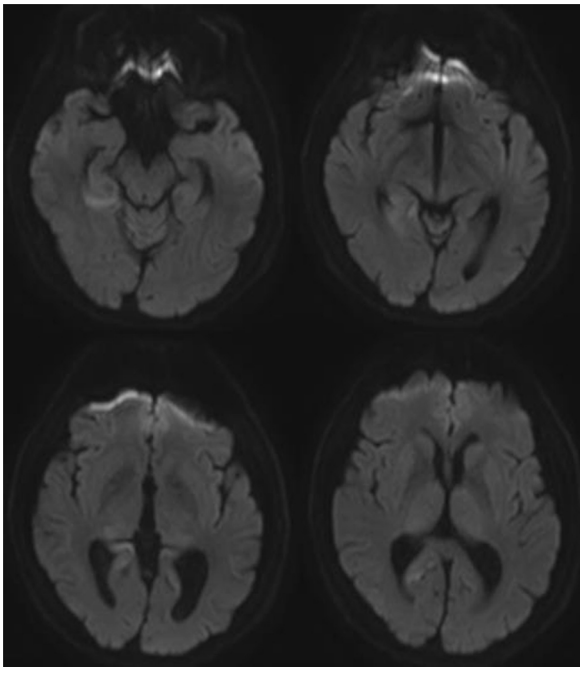

Figure 2 MRI scan at one month. Axial diffusion-weighted sequence showing new cortical hyperintensities, predominantly in the right occipital lobe.

Sporadic CJD is characterised by rapidly progressing dementia, ataxia, muscle tone alterations, and myoclonus.[3] However, patients may not develop all these symptoms, and they may present atypically. Our patient displayed amyotrophy, fasciculations, and arreflexia[4,5]; presentation of these symptoms at disease onset is extremely rare.[6,7] The pathophysiological mechanisms of this rare form of presentation are difficult to determine given that post mortem studies only examine the brain; neural loss in the anterior horn of the spinal cord secondary to spongiform degeneration has been proposed as the causal mechanism.[4,7,8] Amyotrophic CJD was once regarded as a specific form of the disease.[9] Lower motor neuron involvement has been described in familial and sporadic forms of the disease,[6] but has not been associated with a specific genetic alteration; the only case in the literature undergoing genotyping had the 129 Met/Met variant.[6]

Imaging studies reveal isolated cortical hyperintensity in up to one-third of patients with a final diagnosis of sporadic CJD.[10] As in our case, the available evidence suggests that diffusion-weighted sequences are the most sensitive for detecting spongiform changes in the brain secondary to CJD, particularly during the early stages of the disease.[11–13]

Although motor neuron involvement led us to broaden the differential diagnosis, neuroimaging findings and the presence of cerebellar involvement and mild cognitive impairment enabled early aetiological diagnosis. Copresence of amyotrophic lateral sclerosis is unlikely in view of the absence of signs of upper motor neuron involvement, the lack of bulbar symptoms, and the rapid progression of symptoms.

In conclusion, although lower motor neuron involvement is rare in patients with sporadic CJD, it should be included within the clinical spectrum of the condition; lower motor neuron involvement as an early manifestation of the disease may enable early diagnosis.

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References

1. Zerr I, Kallenberg K, Summers DM, Romero C, Taratuto A, Heinemann U, et al. Updated clinical diagnostic criteria for sporadic Creutzfeldt-Jakob disease. Brain. 2009;132:2659–68.
2. CDC's Diagnostic Criteria for Creutzfeldt-Jakob Disease (CJD), 2010. Available from: http://www.cdc.gov/ncidod/dvrd/cjd/diagnostic_criteria.html [accessed 10.03.16].
3. Zerr I, Schulz-Schaeffer WJ, Giese A, Bodemer M, Schröter A, Henkel K, et al. Current clinical diagnosis in CJD: identification of uncommon variants. Ann Neurol. 2000;48:323–9.
4. Esteban JCG, Atares B, Zarranz JJ, Velasco F, Lambarri I. Dementia, amyotrophy, and periodic complexes on electroencephalogram: a diagnostic challenge. Arch Neurol. 2001;58:1669–72.

5. Niewiadomska M, Kulczycki J, Wochnik-Dyjas D, Szpak GM, Rakowicz M, Łojkowska W, et al. Impairment of the peripheral nervous system in Creutzfeldt-Jakob disease. Arch Neurol. 2002;59:1430—6.
6. Panegyres PK, Armari E, Shelly R. A patient with Creutzfeldt-Jakob disease presenting with amyotrophy: a case report. J Med Case Rep. 2013;7:218.
7. Allen IV, Dermott JH, Connolly JH, Hurwitz LJ. A study of a patient with the amyotrophic form of Creutzfelt-Jakob disease. Brain. 1971;94:715—24.
8. Nowacki P, Kulczycki J, Narolewska A, Grzelec H. Amyotrophic form of Creutzfeldt-Jakob disease with rapid course in 82-year-old man. Folia Neuropathol. 2000;38:161—3.
9. Worrall BB, Rowland LP, Chin SM, Mastrianni JA, et al. Amyotrophy in prion diseases. Arch Neurol. 2000;57:33—8.
10. Meissner B, Kallenberg K, Sanchez-Juan P, Krasnianski A, Heinemann U, Varges D, et al. Isolated cortical signal increase on MR imaging as a frequent lesion pattern in sporadic Creutzfeldt-Jakob disease. Am J Neuroradiol. 2008;29:1519—24.
11. Shiga Y, Miyazawa K, Sato S, Fukushima R, Shibuya S, Sato Y, et al. Diffusion-weighted MRI abnormalities as an early diagnostic marker for Creutzfeldt-Jakob disease. Neurology. 2004;63:443—9.
12. Moreno F, Arriola L. Neuroimagen en el diagnóstico de las encefalopatías espongiformes transmisibles humanas. Neurologia. 2006;2:428—36.
13. Ortega-Cubero S, Luquin MR, Domínguez I, Arbizub J, Pagolaa I, Carmona-Abellán MM, et al. Neuroimagen estructural y funcional en las enfermedades priónicas humanas. Neurologia. 2011;27:27.

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Autoimmune necrotising myopathy: A case report☆

Miositis necrosante autoinmune: a propósito de un caso

Dear Editor,

We present the case of a 63-year-old woman with a history of obesity, arterial hypertension, dyslipidaemia, and diabetes mellitus who was being treated with losartan, hydrochlorothiazide, liraglutide, metformin, long-acting insulin, and rosuvastatin. She visited the neurology department due to a one-year history of slightly increased creatin kinase (CK) levels, which were initially symptomless, and were detected in routine blood tests. She subsequently presented progressive weakness in the proximal muscles of the lower limbs, displaying difficulties climbing stairs, and particularly standing up from the floor, which was associated with increased CK values (1932 IU/L). Both the patient's symptoms and the elevated CK levels persisted after discontinuation of statins. The neurological examination revealed normal cranial nerves; the upper limbs displayed preserved strength, slightly hypoactive stretch reflexes, and normal tactile perception; the lower limbs showed bilateral proximal loss of strength, especially in hip flexion (3/5) and adduction (4/5), and no deficit in knee flexion/extension, foot dorsiflexion, or plantar flexion bilaterally, positive Gowers sign, and markedly hypoactive stretch reflexes (1/4); the patient's gait presented mild myopathic features.

Analytical studies showed persistently increasing CK levels, with initial figures of 300-500 IU/L, raising to 1932 IU/L, and finally 2195 IU/L, with increased aldolase levels (22.9 IU/L) and proportionally elevated transaminase levels (AST 55 IU/L and ALT 115 IU/L). The remaining parameters, including blood count, erythrocyte sedimentation rate, and kidney function, displayed normal values. The electromyography study (EMG) showed diffuse moderate myopathic involvement. A quadriceps biopsy revealed focal myophagocytosis, and fibre necrosis with no inflammatory infiltrates, vasculitis, or amyloid deposition. The tests performed revealed no evidence of viral infection, connective tissue disease, or neoplasm. The immunology study yielded positive results for anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (HMGCR) antibodies, confirming the diagnosis of autoimmune necrotising myopathy (ANM) secondary to statin use. The patient started treatment with prednisone at 1 mg/kg/day and azathioprine at 50 mg/12 h, which improved her clinical condition at 10 months of follow-up; regarding motor symptoms, she presented slightly impaired hip flexion (4+/5), with no impaired strength in other muscle groups; CK levels had decreased to 72 IU/L.

ANM secondary to statin use is a rare, recently described clinical entity; the antibody involved in its pathogenesis was identified in 2010.[1] It is characterised by persistence of muscle weakness even after discontinuation of treatment with statins; cellular necrosis in the muscle biopsy; and presence of anti-HMGCR antibodies.[1] Incidence is estimated at 2-3 cases per 100 000 patients treated with statins.[2] The condition is slightly more frequent in women and more common in patients aged 50 years or older.[1] The class II HLA-DRB1*11:01 allele is associated with the development of anti-HMGCR antibodies.[3] In patients with a genetic predisposition, the use of statins causes overexpression of HMGCR through a

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