

the dorsal pons. There was no apparent compromise of the abducens nucleus or the paramedian pontine reticular formation, as in the case we present. In our case, independent from impairment of the medial longitudinal fasciculus, the nucleus raphe interpositus ventral to that structure may be responsible for gaze paralysis.⁵

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Vocal cord paralysis as a manifestation of myasthenia gravis with anti-MuSK antibodies[☆]

Parálisis de cuerdas vocales como forma de presentación de miastenia gravis con anticuerpos anti-MuSK

Dear Editor:

Patients with myasthenia gravis with antibodies to muscle-specific tyrosine kinase (anti-MuSK antibodies) represent a subgroup whose characteristics differentiate them from patients with acetylcholine receptor antibodies. This disorder is predominant in women, and age of onset is typically around 40 years. Patients develop predominantly facial and bulbar symptoms and experience frequent episodes of respiratory failure. They also exhibit poor tolerance for or lack of response to cholinesterase inhibitors.¹ Many patients experience rapid decline at onset and plasmapheresis is the treatment of choice in some hospitals. However, long-term prognosis resembles that of patients presenting myasthenia gravis with acetylcholine receptor antibodies.²

We present the case of a 46-year-old woman with no relevant medical history. She was admitted to the emergency department due to a 3-week history of dyspnoea with moderate exertion, predominantly in the morning, associated with dysphonia and stridor. She presented no other bulbar or ocular symptoms and no limb weakness.

General examination yielded normal results. Neurological examination revealed dysphonic voice associated with stridor, which worsened with repeated efforts. It also revealed predominantly right-sided eyelid ptosis which also intensified with repetition. No ophthalmoparesis or any

other pathological signs were observed during the examination. Direct laryngoscopy revealed vocal fold abduction paresis that had reduced the abduction angle by 20°. A Tensilon® test yielded negative results.

Results from the blood test, including acetylcholine receptor antibodies and calcium channel antibodies, were negative. The patient tested positive for anti-MuSK antibodies: 9.0 nmol/L (normal <0.05). Chest radiography and CT were normal. Electromyography (EMG) showed increased jitter in the frontalis muscle and low-frequency repetitive nerve stimulation of the orbicularis oculi showed a significant decrementing response.

Treatment with pyridostigmine at doses up to 180 mg/day did not improve symptoms and was poorly tolerated, causing diarrhoea and nausea. We prescribed prednisone at a dose of 1 mg/kg/day, which lessened symptoms until the patient was asymptomatic. The dose was reduced to 20 mg/day during follow-up, as greater decreases can lead to stridor, eyelid ptosis, and diplopia.

Vocal cord paralysis can be due to multiple disorders, some of which are neurological. Paralysis causes an increased resistance to airflow, and therefore dysphonia and stridor, as a consequence of weakness of the posterior cricoarytenoid muscles. Although this is rare, vocal cord paralysis may indicate onset of myasthenia gravis. Despite the low frequency of anti-MuSK antibody myasthenia gravis, several cases with a similar effect have been reported^{3,4} because this type of myasthenia gravis mainly affects bulbar muscles. In the cases described to date, including ours, paralysis affects abduction and not adduction, and therefore the condition would preferentially affect posterior cricoarytenoid muscles.

Presence of dysphonia and stridor that intensify with fatigue should lead us to suspect bulbar myasthenia gravis. Lack of response to cholinesterase inhibitors and negative results from a test for acetylcholine receptor antibodies would not rule out this possibility. We have seen that these results are negative in cases of myasthenia gravis with anti-MuSK antibodies. Neurophysiological studies yield data typical of neuromuscular junction disease, including a decrementing response to low-frequency repetitive nerve stimulation and pathological jitter. Another entity which

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may cause bulbar symptoms with fatigue is Eaton-Lambert syndrome, but autonomic symptoms in these patients are usually pronounced (arterial hypertension, dry mouth, sexual dysfunction, etc.). Furthermore, during neurophysiological examination, high-frequency repetitive nerve stimulation shows facilitation, although the phenomenon in Eaton-Lambert syndrome can be indistinguishable from that of myasthenia gravis.

In myasthenia gravis with anti-MuSK antibodies, atrophy of facial and tongue muscles has been described in magnetic resonance studies.⁵ However, this finding is normally reported when the disease is at an advanced stage and its diagnostic utility remains uncertain.

Early diagnosis of myasthenia gravis with anti-MuSK antibodies eliminates unnecessary tests and prevents delays in treatment of what may be a life-threatening condition; a patient can die of respiratory failure if the vocal cords remain closed.

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Inflammatory amyloid angiopathy^{☆,☆☆}

Angiopatia amiloide inflamatoria

Dear Editor:

Cerebral amyloid angiopathy, that is, the deposition of β -amyloid, especially A β 40, on the walls of small cerebral and leptomeningeal arteries, is a very frequent finding both in patients with Alzheimer disease and in cognitively healthy elderly individuals.^{1–3} An anatomical pathology study provides the definitive diagnosis. However, finding typical abnormalities in a brain MRI, plus detecting cerebral amyloid disease by means of PiB-PET or by measuring the decrease in A β -40 and A β -42 in cerebrospinal fluid (CSF), eliminates the need for a histopathological study.³ One infrequent presentation known as inflammatory amyloid angiopathy (IAA) is characterised by the development of perivascular inflammatory infiltrates associated with amyloid angiopathy. Its clinical symptoms typically include rapidly progressive cognitive impairment, epileptic seizures, and headache. This entity was described in the 1980s as an isolated type of vasculitis of the central nervous system (CNS). However, with the appearance of the first cases of A β -induced meningoencephalitis, it began to be regarded as a different entity in which the β -amyloid deposited on the vascular walls acts as an antigen.^{4–6} This hypothesis has been reinforced by

the description of anti-A β -42 antibodies in the CSF and cerebral parenchyma as well as by new histopathology studies.^{7–9}

Here, we present a case of IAA and Alzheimer disease in which we concluded that biopsy was not necessary to assign a diagnosis.

An 82-year-old man was seen by the emergency department for head trauma caused by a fall the same day. Relevant medical history included multiple facial basal cell carcinomas that were surgically removed and treated with radiation in 2003 due to local recurrence. While in the emergency department, the patient displayed disorientation with no focal neurological signs. CT revealed haemorrhagic foci and extensive bilateral white-matter hypodensities that suggested underlying neoplasia. The patient was admitted to the neurosurgery department and treated with dexamethasone 4 mg/8 h.

Roughly 6 months before being admitted, the patient had begun to experience memory lapses; 3 months after that, he had developed apathy, hand tremor, and disorientation. His condition had deteriorated considerably by the week before he was admitted, and he needed supervision when showering.

Over the first few days of hospitalisation, his cognitive function grew worse and he experienced agitation episodes and night-time hallucinations. After a week of hospitalisation and treatment with dexamethasone, both his tremor and cognitive function began to improve significantly; his family found his condition to be better than it was before he was admitted. At that time, we performed a brain MRI which revealed large areas of vasogenic oedema in occipital-parietal and frontal regions of the right hemisphere, with more limited areas in the left hemisphere (Fig. 1a and b). Multiple millimetre-sized foci of hemosiderin deposition, predominantly cortical and diffuse, were also present

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^{☆☆} This case was presented at the case report contest held by the Valencian Society of Neurology.