

external ophthalmoplegia is present. These disorders usually resolve in a significantly shorter period than is the case for ophthalmoplegia.<sup>6</sup> These findings show that the involvement of pupillomotor fibres is independent of any lesions in other subdivisions of the oculomotor nerve.

The patient will subsequently start to develop partial external ophthalmoplegia and mild gait ataxia with hyporeflexia.

As a general rule, anti-GQ1b IgG antibodies are present in more than 90% of MFS cases.<sup>7–9</sup> This is because oculomotor nerves and the optic nerve contain large quantities of GQ1b gangliosides.<sup>10</sup> Evaluating these antibodies and other gangliosides is helpful but not essential to the diagnostic process. In our case, no anti-GQ1b IgG antibodies were detected despite the fact that the patient presented external and internal ophthalmoplegia, but this does not rule out MFS. In this case, the diagnosis was determined based on clinical signs, albuminocytological dissociation in CSF, and having ruled out other processes.

Neurophysiological studies were not necessary in this case given that clinical progression and CSF analyses provided sufficient data. However, such studies can help determine early diagnosis in some specific cases, especially in the acute phase.<sup>11</sup>

The treatment of choice for MFS is IV immunoglobulins, or plasmapheresis in drug-resistant cases.<sup>12</sup> The patient started to improve on the fifth day after starting treatment with IV immunoglobulins, and her condition had resolved completely within 2 months.

In conclusion, we present a rare case of MFS with an exceptional form of onset consisting of isolated internal ophthalmoplegia and fronto-orbital headache. Furthermore, anti-GQ1b IgG antibodies are not present even though the patient presented internal and external ophthalmoplegia. The incidence rate of this condition has probably been underestimated since it may be overlooked in the initial diagnostic process when symptoms are so mild that they may spontaneously resolve. In our case, lack of concomitant neurological signs meant that onset could have been overlooked, but the condition progressed rapidly in this patient, resulting in manifestation of the classic triad. We must be aware of this type of condition since it progresses quickly and consequences for patients may be severe.

## References

1. Pou-Serradell A. Neuropatías disímunes adquiridas. Sintomatología clínica y clasificación. *Rev Neurol.* 2000;30:501–10.
2. Fisher M. An unusual variant of acute idiopathic polyneuritis (syndrome of ophthalmoplegia, ataxia and areflexia). *N Engl J Med.* 1956;255:57–65.
3. Sugita A, Yanagisawa T, Kamo T, Takahashi Y, Auki N. Internal ophthalmoplegia with anti-GQ1b IgG antibody. *J Neurol.* 2002;249:1475–6.
4. Chan YC, Wilder-Smith E, Chee MWL. Acute ophthalmoplegia with pupillary areflexia associated with anti-GQ1b antibody. *J Clin Neurosci.* 2004;11:658–60.
5. Bae JS, Kim JK, Kim SH, Kim OK. Bilateral internal ophthalmoplegia as an initial sole manifestation of Miller Fisher syndrome. *J Clin Neurosci.* 2009;16:963–44, <http://dx.doi.org/10.1016/j.jocn.2008.09.009> [Epub 13.03.09].
6. Mori M, Kuwabara S, Fukutake T, Yuki N, Hattori T. Clinical features and prognosis of Miller Fisher syndrome. *Neurology.* 2001;56:1104–6.
7. Snyder LA, Rismondo V, Millar NR. The Fisher variant of Guillain-Barre syndrome (Fisher syndrome). *J Neuroophthalmol.* 2009;29:312–24.
8. O'Hanlon GM, Plomp JJ, Chakrabarti M, Morrison I, Wagner ER, Goodyear CS, et al. Anti-GQ1b ganglioside antibodies mediate complement-dependent destruction of the motor nerve terminal. *Brain.* 2001;124:893–906.
9. Lo YL. Clinical and immunological spectrum of the Miller Fisher syndrome. *Muscle Nerve.* 2007;36:615–27.
10. Yuki N, Odaka M, Hirata K. Acute ophthalmoparesis (without ataxia) associated with anti-GQ1b antibody: clinical features. *Ophthalmology.* 2001;108:196–200.
11. Gabaldón Torres L, Badía Picazo C, Salas Felipe J. Papel del estudio neurofisiológico en el síndrome de Miller Fisher. *Neurología.* 2012, <http://dx.doi.org/10.1016/j.nrl.2012.04.014>.
12. Sederholm BH. Treatment of acute immune-mediated neuropathies: Guillain-Barré syndrome and clinical variants. *Semin Neurol.* 2010;30:365–72.

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## Transient ischaemic attack secondary to extracranial carotid artery aneurysm<sup>☆</sup>

### Accidente isquémico transitorio secundario a aneurisma carotídeo extracraneal

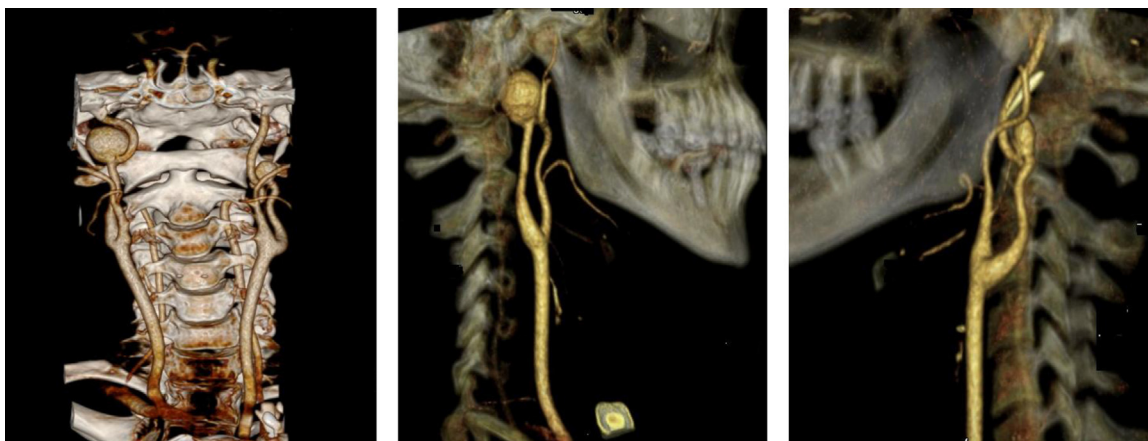
Dear Editor:

Extracranial carotid artery aneurysms present a low incidence (1.3%), making it more difficult to study

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their aetiology, natural course, and response to treatment.<sup>1–6</sup>

We present the case of a 32-year-old woman, a former smoker taking oral contraceptives. The patient presented a 15-minute self-limited episode of dysarthria and loss of strength in the right arm, followed by spontaneous full recovery. Results from the neurological examination were normal and no alterations were found in the blood test, electrocardiogram, chest radiography, or head CT. Clinical signs were compatible with transient ischaemic attack (TIA) in the left hemisphere. We requested tumour markers, serology tests, immunology tests, and a hypercoagulation study, which only revealed a homozygous *MTHFR* C667T mutation. We performed a transthoracic echocardiogram that showed no alterations. An echo Doppler study of the supra-aortic



**Figure 1** CT-angiography of SAT: patent saccular aneurysm in the distal portion of both ICs (19 mm on the RIC and 16 mm on the LIC).

trunks (SAT) and a CT-angiography of SAT, circle of Willis (CW) and thoracoabdominal aorta revealed a patent saccular aneurysm in the distal cervical portion of both internal carotid arteries (IC), with a maximum diameter of 19 mm on the RIC and 16 mm on the LIC (Fig. 1). After evaluating the case and ruling out surgical intervention because carotid artery aneurysms were inaccessible, we decided to perform endoluminal repair. Firstly, the symptomatic aneurysm on the LIC was repaired by placing 3 intraaneurysmal Matrix<sup>®</sup> coils and a Silk<sup>®</sup> stent (4 mm × 30 mm). Total thrombosis of the aneurysm, adequate permeability of the distal portion of the LIC and intracranial circulation were achieved (Fig. 2). Secondly, the asymptomatic aneurysm on the RIC was repaired by placing 3 intraaneurysmal GDC<sup>®</sup> coils and a Silk<sup>®</sup> stent (4 mm × 30 mm). Total thrombosis of the aneurysm, adequate permeability of the distal portion of DIC and intracranial circulation were achieved (Fig. 2). The patient was asymptomatic upon discharge and treated with dual anti-platelet therapy for 2 months, followed by single anti-platelet therapy to be continued for indefinitely. After 2 years of follow-up, she remains asymptomatic and the results of the endoluminal repair appear to be good.

Extracranial carotid artery aneurysms are usually located in the carotid bifurcation, followed by the IC and the external carotid artery as the next most frequent locations.<sup>1,2,4</sup> As in our case and according to some series, they may be bilateral, and/or associated with aneurysms in other locations.<sup>1</sup> They may be fusiform or saccular in shape<sup>1,2</sup> and their aetiologies vary.<sup>1-5</sup> Their main cause in the past was infection, but it is currently atherosclerosis (55%), followed by fibromuscular dysplasia of the arteries, trauma, dissection, and surgical procedures. Other less frequent causes are cystic medial necrosis, Marfan syndrome, Takayasu arteritis and idiopathic medial aortopathy.<sup>1-4</sup> Up to 60% of extracranial carotid artery aneurysms are symptomatic and their clinical manifestations vary according to their location, size, and aetiology. Patients may present symptoms compatible with TIA or stroke (40-45%)<sup>1-4</sup> as in our case, retro-orbital compression, pulsating headache, dysphagia, relapsing facial pain, deafness, hoarseness, tinnitus, Horner syndrome, and Raeder paratrigeminal syndrome. Physical examination usually reveals a pulsating mass on the neck or pharynx, often painful, and associated in some cases with deficits and/or focal neurological signs.<sup>1-3</sup> Differential diagnosis should examine carotid artery kinking or



**Figure 2** Embolisation of aneurysm on the RIC and LIC.

elongation, carotid body tumour, adenopathies, peritonsillar abscess, branchial cleft cyst, and cystic hygroma.<sup>1,2</sup> Echo Doppler and especially CT-angiography or MRI-angiography of SAT and CW are necessary to determine the diagnosis, but today's gold standard for assessing anatomical details and choosing the optimal treatment is arteriography of SAT and CW.<sup>1-3</sup> Treatment indications depend on the aneurysm's clinical manifestations, size, location, and aetiology, as well as the patient's surgical risk.<sup>1-3,5,6</sup> The aim of treatment is to prevent severe neurological complications and associated secondary mortality.<sup>1,2,5</sup> Extracranial carotid artery aneurysms show a mortality rate of 71% due to thrombosis, embolism, or rupture. In patients undergoing surgical or endoluminal repair, this rate decreases to 30%.<sup>1-4</sup> Today's treatment alternatives are surgical procedures involving aneurysm exclusion and arterial suture, or bypass graft (prosthetic or autologous). Techniques are associated with neurological morbidity (peripheral and central) ranging between 6% and 20%, depending on the series, and a mortality rate of about 2%.<sup>1-5</sup> Another alternative is endoluminal treatment with aneurysm embolisation and placement of endoprosthesis; this is useful when the aneurysms are surgically inaccessible, as in our case, or in patients with a high surgical risk. This last alternative is on the rise, but no randomised studies that analyse long-term results are available at present.<sup>1-3</sup> To conclude, we highlight that while this entity is infrequent, it should be considered among the possible causes of TIA or stroke.<sup>1</sup>

## References

1. Rutherford. *Cirugía vascular: aneurismas de la arteria carótida extracraneal*, vol. 2, 6.s ed. Madrid: Elsevier España S.A.; 2006. p. 2052–62.
2. SEACV *Tratado de las Enfermedades Vasculares: aneurismas de los troncos, supraaórticos*, vol. 2. Barcelona: Viguera Editores, S.L.; 2006. p. 777–85.
3. Attigah N, Külkens S, Zausig N, Hansmann J, Ringleb P, Hakimi M, et al. Surgical therapy of extracranial carotid artery aneurysms: long-term results over a 24-year period. *Eur J Vasc Endovasc Surg*. 2009;37:127–33.
4. Li Z, Chang G, Yao C, Guo L, Liu Y, Wang M, et al. Endovascular stenting of extracranial carotid artery aneurysm: a systematic review. *Eur J Vasc Endovasc Surg*. 2011;42:419–26.
5. Radak D, Davidovic L, Vukobratov V, Ilijevski N, Kostic D, Maksimovic Z, et al. Carotid artery aneurysms: Serbian multicentric study. *Ann Vasc Surg*. 2007;21:23–9.
6. Davidovic L, Kostic D, Markovic D, Vasic D, Markovic M, Duvnjak S, et al. Carotid artery aneurysms. *Vascular*. 2004;12:166–70.

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## Myasthenia gravis in association with extrathymic neoplasia<sup>☆</sup>

### Miastenia gravis y asociación con neoplasias extratímicas

*Dear Editor:*

Myasthenia gravis (MG) is considered a paraneoplastic phenomenon and it is associated with thymoma in 15% of all thymoma patients. However, its association with other extrathymic malignancies remains a matter of debate that has not been completely investigated.<sup>1,2</sup>

In an MG prevalence study carried out in our setting,<sup>3</sup> we found that 3 out of 29 patients (10%) had presented extrathymic malignancies previous to or at the time of diagnosis with MG. These 3 cases were a man with history of non-Hodgkin lymphoma, a man diagnosed with gastric adenocarcinoma when admitted due to MG symptom onset, and a woman with history of breast cancer.

The aim of our study is to review and discuss the association of MG with other extrathymic malignancies and present 3 new cases evaluated in our hospital.

*Case 1.* Male patient 62 years of age who had presented generalised adenopathy 15 years previously. Imaging tests showed bilateral axillary adenopathies, retroperitoneal adenopathies, and adenopathies on the right paratracheal lymph nodes. Results from the anatomical pathology study of adenopathies and of the spine were compatible with follicular mixed small-cleaved and large-cell lymphoma. Since the patient was clinically and radiologically stable, no treatment had been started at that date. Regarding neurological follow-up, the patient presented fluctuating diplopia in the past year that responded well to treatment with pyridostigmine (240 mg/day). A few months later, the patient's condition worsened and he presented total ptosis and difficulty chewing. Doctors then started treatment with low doses of prednisone (10 mg/day), with good tolerability and efficacy. Diagnosis of MG was based on the electrophysiological study, which revealed a pathological decrement of more than 10% to low-frequency repetitive facial nerve stimulation. No presence of thymoma has been confirmed by any of the imaging tests requested during follow-up. Although no antibodies were detected initially, subsequent measurement of anti-AChR antibodies showed high levels in blood, with a titre of 3.98 nmol/L (normal level < 0.20 nmol/L).

*Case 2.* A 72-year-old man was admitted to the emergency department due to progressive loss of limb and neck strength with no bulbar or ocular symptoms. He did not present wasting syndrome or pain. Treatment with IV immunoglobulins and pyridostigmine was started and

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