venting this risk factor; the use of maps of high-pollution areas and tools for calculating vascular risk will enable clinicians to make individualised recommendations to reduce the risk of cerebrovascular disease associated with exposure to air pollution.[13]

References

1. Feigin VL, Roth GA, Naghavi M, Parmar P, Krishnamurthi R, Chugh S, et al. Global burden of stroke and risk factors in 188 countries, during 1990—2013: a systematic analysis for the Global Burden of Disease Study 2013. Lancet Neurol. 2016;15:913—24.
2. Maheswaran R. Air pollution and stroke — an overview of the evidence base. Spat Spatiotemporal Epidemiol. 2016;18:74—81.
3. Vivanco-Hidalgo RM, Wellenius G, Basagaña X, Cirach M, González AG, de Ceballos P, et al. Short-term exposure to traffic-related air pollution and ischemic stroke onset in Barcelona, Spain. Environ Res. 2018;162:160—5.
4. Shah ASV, Lee KK, McAllister D, Hunter A, Nair H, Whiteley W, et al. Short term exposure to air pollution and stroke: systematic review and meta-analysis. BMJ. 2015;350:h1295.
5. Yuan S, Wang J, Jiang Q, He Z, Huang Y, Li Z, et al. Long-term exposure to PM2.5 and stroke: a systematic review and meta-analysis of cohort studies. Environ Res. 2019;177:108587.
6. Mustafić H, Jabre P, Caussin C, Murad MH, Escolano S, Tafflet M, et al. Main air pollutants and myocardial infarction: a systematic review and meta-analysis. JAMA - J Am Med Assoc. 2012;307:713—21.
7. Shah ASV, Langrish JP, Nair H, McAllister DA, Hunter AL, Donaldson K, et al. Global association of air pollution and heart failure: a systematic review and meta-analysis. Lancet. 2013;382:1039—48.
8. Brook RD, Franklin B, Cascio W, Hong Y, Howard G, Lipsett M, et al. Air pollution and cardiovascular disease. Circulation. 2004;109:2655—71.
9. Fihn SD, Gardin JM, Abrams J, Berra K, Blankenship JC, Dallas AP, et al. 2012 ACCF/AHA/ACP/AATS/PCNA/SCAI/STS guideline for the diagnosis and management of patients with stable ischemic heart diseass. Circulation. 2012;126.
10. Piepoli MF, Hoes AW, Agewall S, Albus C, Brotons C, Catapano AL, et al. 2016 European Guidelines on cardiovascular disease prevention in clinical practice. Eur Heart J. 2016;37:2315—81.
11. Knuuti J, Wijns W, Achenbach S, Agewall S, Barbato E, Bax JJ, et al. 2019 ESC guidelines for the diagnosis and management of chronic coronary syndromes. Eur Heart J. 2020;41:407—77.
12. García Pastor A, López-Cancio Martínez E, Rodríguez-Yáñez M, Alonso de Leciñana M, Amaro S, Arenillas JF, et al. Comité ad hoc del Grupo de Estudio de Enfermedades Cerebrovasculares de la Sociedad Española de Neurología. Recomendaciones de la Sociedad Española de Neurología para la prevención del ictus. Actuación sobre los hábitos de vida y la contaminación atmosférica. Neurol. 2020, http://dx.doi.org/10.1016/j.nrl.2020.05.018 (article in press).
13. Hadley MB, Baumgartner J, Vedanthan R. Developing a clinical approach to air pollution and cardiovascular health. Circulation. 2018;137:725—42.

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The Foix-Chavany-Marie syndrome due to herpes symplex virus encephalitis type 2☆

Síndrome de Foix-Chavany-Marie secundario a encefalitis por virus herpes simple tipo 2

Dear Editor:

The Foix-Chavany-Marie syndrome (FCMS) or anterior opercular syndrome is characterised by loss of voluntary movement of the orofacial and pharyngeal muscles, with preserved automatic and reflex movements,[1] secondary to bilateral lesions to the anterior opercular cortex; however, cases of unilateral involvement or bilateral subcortical lesions have also been reported.[2] The most frequent aetiology is vascular,[3] but the syndrome may also be of infectious, demyelinating, traumatic, neoplastic, epileptic, or neurodegenerative origin.[4] We describe a case secondary to herpes simplex virus type 2 (HSV-2) encephalitis.

Our patient was a 27-year-old, right-handed woman with history of untreated autoimmune thyroiditis, who presented a 2-month history of progressive bilateral facial weakness predominantly affecting the left side, inability to speak due to anarthria and severe hypophonia, and swallowing difficulties together with severe mixed dysphagia and sialorrhoea. In the previous 24 hours, she had presented 3 episodes suggestive of focal seizures, of 5 min duration each: the first episode consisted of tonic rigidity of the upper limbs preceded by snoring during sleep; in the second episode, she presented jaw rigidity and globus sensation; the third episode consisted of left facial clonic movements.

The general examination revealed no relevant data. The neurological examination revealed a loss of voluntary movement in the muscles of the face, tongue, and pharynx, with

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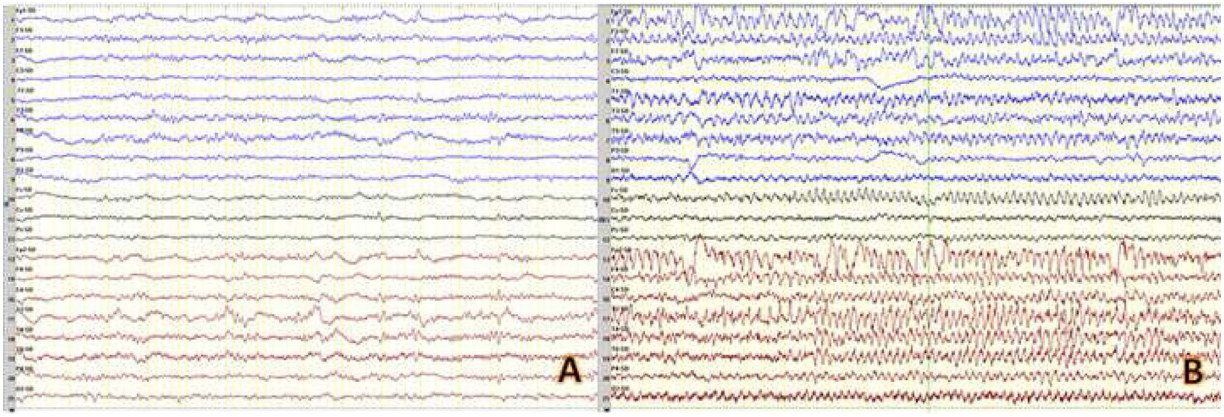

Figure 1 Video EEG. A) Interictal EEG showing right frontotemporal focal abnormalities consisting of periodic complex waves with a frequency below 1 Hz and variable propagation to other areas. B) Ictal EEG recording showing a generalised spike-and-wave pattern that was preceded by recruiting rhythms in the right temporal region, with fast propagation.

preservation of the reflex blink to visual threat, automatic facial mimicry, and gag and coughing reflexes; these findings are compatible with FCMS.

A complete blood analysis, including serology tests for HIV and a lymphocyte population study, yielded normal results. Serial electroencephalography (EEG) studies showed epileptiform activity in right temporal regions, together with signs of focal slowing. During a video EEG study, the patient presented a generalised tonic-clonic seizure with a generalised spike-and-wave pattern preceded by right temporal recruiting rhythm (Fig. 1). Brain magnetic resonance imaging (MRI) revealed T2-FLAIR hyperintensities without contrast uptake in both insular cortices, superior temporal gyri, cingulate, and pre- and postcentral gyri (Fig. 2). A cerebrospinal fluid (CSF) analysis revealed a high protein level (0.81 g/dL), with 121 cells (94% lymphocytes); a PCR test for HSV-2 yielded positive results. Results were negative in the other microbiological studies, including the PCR tests for the remaining viruses of the herpes, cytomegalovirus, human parechovirus, and enterovirus families; PCR test and antigen detection for Cryptococcus; Gram staining and bacterial cultures; and PCR test for *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Listeria monocytogenes*; as well as testing for anti-NMDA receptor antibodies.

We started treatment with intravenous aciclovir for 21 days and antiepileptic treatment with lacosamide; a follow-up CSF analysis returned negative PCR results for HSV-2. A follow-up brain MRI performed at 20 days showed cystic necrosis in bilateral frontal regions (Fig. 2).

The patient showed improvements in facial mobility and was able to perform voluntary movements with the face, mouth, and tongue; however, due to persistent dysarthria, dysphonia, and dysphagia, she required nutrition via nasogastric tube for 3 months, which was subsequently removed. No further epileptic seizures were observed.

Herpesvirus encephalitis is the most frequent cause of sporadic encephalitis worldwide, with HSV-1 infection being more frequent in adults and HSV-2 in newborns.[5] HSV-2 encephalitis in adults is exceptional, and has been described in association with ischaemic and/or haemorrhagic stroke,[6] or with symptoms similar to those of HSV-1 encephalitis.[5]

The most frequent cause of FCMS in adults is vascular lesions due to simultaneous or sequential involvement of both opercular cortices or their subcortical connections.[2,3] It has rarely been associated with a unilateral opercular lesion.[7] HSV-1 encephalitis is an important infectious cause of FCMS,[8,9] due to its high prevalence and the predominant involvement of the frontal lobes.[5] The most typical presentation consists of encephalitis with fever, impaired level of consciousness, and epileptic seizures (mainly focal motor seizures) whose clinical symptoms include signs of opercular syndrome; however, cases have also been reported of FCMS in isolation or associated with focal motor seizures only.[10] In these cases, the appearance of recurrent focal seizures, even in the absence of fever, headache, or other data suggesting meningoencephalitis, should move us to rule out infection of the central nervous system, especially in patients presenting no vascular risk factors. Other microbial causes (cytomegalovirus, Epstein-Barr virus, toxoplasma, tuberculosis, or JC virus) are extremely rare.[11–14]

Other causes described include head trauma, demyelinating lesions, brain tumours, neurodevelopmental disorders in childhood, neurodegenerative diseases, and status epilepticus.[1,15]

Our patient's symptoms initially consisted of focal seizures, with no language impairment or altered level of consciousness between seizures, and without headache, fever, or increased levels of acute-phase reactants. She subsequently developed anarthria, dysphagia, and bilateral facial paresis with preserved automatic and emotional responses (smiling, crying, yawning), which is typical of

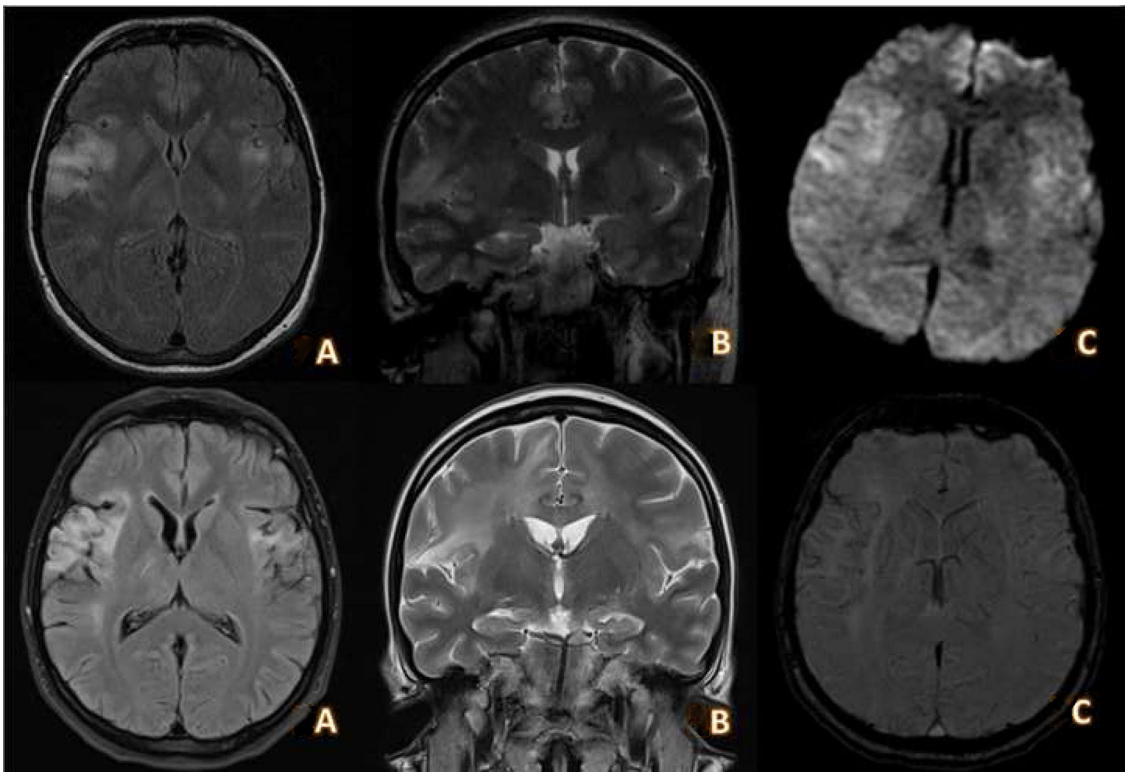

Figure 2 Brain MRI study performed during the acute phase (top) and at 20 days after completion of antiviral treatment (bottom). Top, A and B) Axial and coronal slices from a T2-FLAIR sequence showing hyperintense lesions extending through the insular cortex and superior temporal gyri towards the pre- and postcentral gyri and predominantly right cingulate gyrus. C) DWI sequence showing moderately restricted diffusion in the lesions described. Bottom, A and B) Axial and coronal slices from a T2-FLAIR sequence showing areas of gliosis with cystic necrosis in the insular, opercular, and bilateral frontotemporal regions, predominantly affecting the right side. C) DWI sequence showing haemosiderin deposition predominantly in the right insular area, compatible with superficial siderosis secondary to encephalitis.

bilateral anterior opercular involvement. This automatic-voluntary dissociation is explained by preservation of the indirect corticobulbar pathway.[1]

In short, HSV-2 encephalitis can cause FCMS in immunocompetent adults; this has not previously been described in the literature.

References

1. Weller M. Anterior opercular cortex lesions cause dissociated lower cranial nerve palsies and anarthria but no aphasia: Foix-Chavany-Marie syndrome and ''automatic voluntary dissociation'' revisited. J Neurol. 1993;240:199—208.
2. Moragas Garrido M, Cardona Portela P, Martínez-Yélamos S, Rubio Borrego F. Heterogeneidad topográfica del síndrome de Foix-Chavany-Marie. Neurología. 2007;22:333—6.
3. Milanlioglu A, Aydn MN, Gökgül A, Hamamc M, Erkuzu MA, Tombul T. Ischemic bilateral opercular syndrome. Case Rep Med. 2013;2013:3—6.
4. Bakar M, Kirshner HS, Niaz F. The opercular-subopercular syndrome: Four cases with review of the literature. Behav Neurol. 1998;11:97—103.
5. Kennedy PGE, Steiner I. Recent issues in herpes simplex encephalitis. J Neurovirol. 2013;19:346—50.
6. Zis P, Stritsou P, Angelidakis P, Tavernarakis A. Herpes Simplex Virus Type 2 Encephalitis as a Cause of Ischemic Stroke: Case Report and Systematic Review of the Literature. J Stroke Cerebrovasc Dis. 2016;25:335—9.
7. Ohtomo R, Iwata A, Tsuji S. Unilateral opercular infarction presenting with foix-chavany-marie syndrome. J Stroke Cerebrovasc Dis [Internet]. 2014;23:179—81.
8. Wolf RW, Schultze D, Fretz C, Weissert M, Waibel P. Atypical herpes simplex encephalitis presenting as operculum syndrome. Pediatr Radiol. 1999;29:191—3.
9. McGrath NM, Anderson NE, Hope JKA, Croxson MC, Powell KF. Anterior opercular syndrome, caused by herpes simplex encephalitis. Neurology. 1997;49:494—7.
10. Almekhlafi MA, Couillard PL, Patry DG, Jetté N. Herpes encephalitis presenting with an opercular syndrome and epilepsia partialis continua. Neurologist. 2010;16:208—10.
11. Conforti R, Capasso R, Capaldo G, Dato C, Saracino D, Di Iorio G, et al. Foix-chavany-marie syndrome in a 17-year-old female with congenital cytomegalovirus infection. Neuropsychiatr Dis Treat. 2014;10:2249—52.
12. Matsushima T, Nishioka K, Tanaka R, Yokoyama K, Hattori N. Anterior opercular syndrome induced by Epstein—Barr virus encephalitis. Neurocase. 2016;22:103—8.
13. Moodley M, Bamber S. The Operculum Syndrome: an Unusual Complication of Tuberculous Meningitis. Dev Med Child Neurol. 1990;32:919—22.
14. Dijkstra F, Guldolf K, Schotsmans K, Marechal E, Hernalsteen D, Crols R. Foix-chavany-marie syndrome as the presenting sign of HIV-related PML. Neurol Clin Pract. 2018;8:537—40.

15. Uttner I, Brettschneider J, Unrath A, Riecker A. Slowly progressive Foix-Chavany-Marie syndrome as a precursor of a primary progressive aphasia. J Clin Neurosci. 2012;19:765—7.

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