

CASE REPORT

Unilateral opercular infarction presenting with Foix-Chavany-Marie syndrome

Francisca Sá,^{1,2} Inês Menezes Cordeiro,^{1,2} Susana Mestre,^{1,3} Hipólito Nzwalo^{1,2}¹Stroke Unit, Centro Hospitalar do Algarve, Portugal²Neurology Department, Centro Hospitalar do Algarve, Portugal³Department of Physical Medicine and Rehabilitation, Centro Hospitalar do Algarve, Portugal**Correspondence to**

Dr Inês Menezes Cordeiro, ines.mcordeiro@gmail.com

Accepted 14 November 2014

SUMMARY

Foix-Chavany-Marie syndrome (FCMS) also known as bilateral anterior opercular syndrome is a form of suprabulbar palsy defined by the presence of bilateral voluntary facial, pharyngeal, lingual and masticatory paralysis with automatic–voluntary movement dissociation. We report an extremely rare case of FCMS in a patient with a unilateral left opercular lesion associated with a chronic asymptomatic contralateral cerebellar lesion. Despite intensive rehabilitation, little improvement was noticed at hospital discharge.

BACKGROUND

Foix-Chavany-Marie syndrome (FCMS), also known as bilateral anterior opercular syndrome, is an extremely rare suprabulbar palsy defined by the presence of bilateral voluntary facial, pharyngeal, lingual and masticatory paralysis with automatic–voluntary movement dissociation.^{1–4} The majority of cases are secondary to successive vascular insults bilaterally involving the anterior opercular regions.^{1–5} However, FCMS can also occur in association with central nervous system inflammatory conditions, neurodegenerative disorders, epileptic activity, cortical malformations and traumatic lesions.^{1–5} Unilateral lesions of the anterior opercular often coexisting with contralateral cortical–subcortical lesions in the primary motor cortex or its descending pathways can also cause FCMS.^{4–5} We present a case of FCMS following an acute unilateral opercular stroke in a patient with a chronic asymptomatic contralateral cerebellar lesion.

CASE PRESENTATION

A 76-year-old right-handed man with a medical history of type 2 diabetes mellitus and hyperuricaemia was submitted to biological aortic valve replacement. On the fourth day after surgery, he suddenly developed anarthria and voluntary bilateral facial, pharyngeal, lingual and masticatory paralysis. He was able to close his eyes during sleep, to yawn and to manifest emotional facial mimicry. Writing and comprehension were intact. There were no motor or sensory deficits. The initial acute brain tomography scan was unremarkable. Because the patient was hypocoagulated, local intra-arterial thrombolytic therapy was attempted without success. The brain MRI performed a week later disclosed the presence of acute infarction in the left opercular region (figure 1A, B). In addition to the presence of microvascular chronic disease, the brain MRI also disclosed the presence of a right cerebellar hemisphere chronic lesion (figure 1C, D).

Owing to the presence of severe dysphagia, a percutaneous endoscopic gastrostomy was placed.

INVESTIGATIONS

The complementary diagnostic work up, including ECG, Doppler of cervical vessels, carotid angiogram, thyroid hormones, haemogram and glycated haemoglobin yielded normal results.

OUTCOME AND FOLLOW-UP

Despite an intensive rehabilitation programme, no improvement was noticed at discharge from hospital after 1 month.

DISCUSSION

FCMS is a cortico-subcortical type of pseudobulbar palsy usually caused by bilateral lesions of the anterior operculum, which contains the motor fibres for cranial nerves V, VII, IX, X and XII.^{1–5} The dissociation between the presence of reflexive and automatic muscle action and voluntary palsy in FCMS is explained by the existence of alternative pathways connecting the amygdala and hypothalamus to the brainstem.^{1–5} FCMS is commonly secondary to sequential infarcts to bilateral anterior operculum or subcortical insular regions.^{3–4} However, unilateral anterior operculum lesions can cause FCMS.⁵ These cases can occur in patients with a single anterior opercular lesion or in patients with a single opercular lesion with a coexistent contralateral white matter lesion.^{2–5–6} It is hypothesised that the existence of contralateral subcortical lesions, whether or not visible by brain MRI, could interrupt the projections from the anterior opercular cortex causing FCMS.⁵ Unilateral representation of motor centres for the muscles involved in FCMS is also a possibility.^{2–5–7} However, Martino *et al* documented a case of transient FCMS secondary to intraoperative iatrogenic unilateral damage of the subcortical connections of the inferior frontal operculum gyrus with the frontal aslant tract and arcuate fasciculus. Functional MRI excluded the hypothesis of unilateral representation of the motor centres involved in FCMS.⁸ Tractography reconstruction of the damaged tracts (frontal aslant tract and arcuate fasciculus) allowed, for the first time, demonstration of a precise localisation of a structural substrate of FCMS.⁸ The role of the frontal aslant tract in particular as an important mediator of the connection between the supplementary motor area control of orofacial movement is increasingly being recognised. Motor initiation and speech disturbances are manifestations associated with lesions of the frontal aslant tract.⁹



CrossMark

To cite: Sá F, Menezes Cordeiro I, Mestre S, *et al*. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-206439

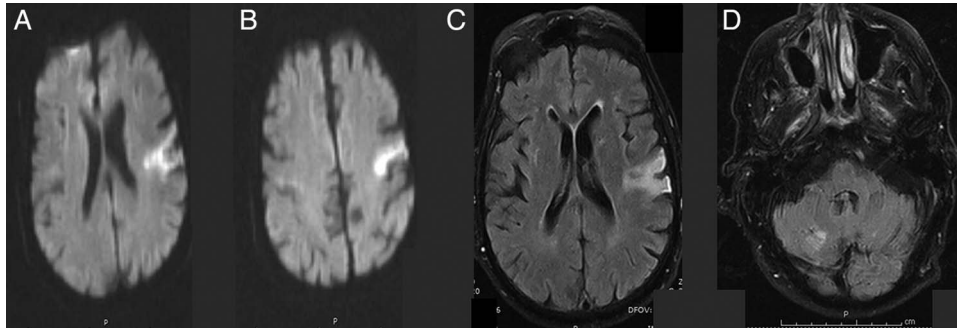


Figure 1 (A and B) Brain MRI showing acute infarction of the left opercular region on diffusion-weighted (A and B), T2 fluid-attenuated inversion recovery (FLAIR) (C) and chronic right cerebellar hemisphere on T2 FLAIR (D).

Torres-Perales *et al*¹⁰ presented a case of FCMS with a unilateral opercular lesion associated with a cerebellar chronic lesion. Considering the active contribution of the cerebellum in motor activity, the authors advanced that this association could not be a simple coincidence.¹⁰

Contrary to cases caused by unilateral opercular lesions, in FCMS with bilateral lesions, the prognosis tends to be poor, with the majority of patients having persistent anarthria and dysphagia.⁶ In our case, despite the presence of a unilateral opercular lesion and intensive rehabilitation, little recovery occurred.

In conclusion, in the presence of acute anarthria, and absence of voluntary facial and tongue movements with automatic–voluntary movement dissociation, the diagnosis of FCMS should be considered. Unilateral anterior operculum lesions can cause FCMS.

Contributors FS, SM and IMC directly cared for the patient. FS and IMC prepared the manuscript. SM and HN made the critical revision.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Foix C, Chavany JA, Marie J. Diplégie facioliinguomasticatrice d'origine cortico souscorticale sans paralysie des membres. *Rev Neurol* 1926;33:214–19.
- 2 Mao CC, Coull BM, Golper LAC, *et al*. Anterior operculum syndrome. *Neurology* 1989;39:1169–72.
- 3 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.
- 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 Ohtomo R, Iwata A, Tsuji S. Unilateral opercular infarction presenting with Foix-Chavany-Marie syndrome. *J Stroke Cerebrovasc Dis* 2014;23:179–81.
- 6 Brandão E, Ferreria A, Leal Loureiro J. Anterior biopercular syndrome caused by unilateral infarction. *Acta Med Port* 2013;26:177–9.
- 7 Moragas-Garrido M, Cardona-Portela P, Martínez-Yélamos S, *et al*. Heterogeneidad topográfica del síndrome de Foix-Chavany-Marie. *Neurología* 2007;22:333–6.
- 8 Martino J, de Lucas EM, Ibáñez-Plágaro FJ, *et al*. Foix-Chavany-Marie syndrome caused by a disconnection between the right pars opercularis of the inferior frontal gyrus and the supplementary motor area. *J Neurosurg* 2012;117:844–50.
- 9 Kinoshita M, de Champfleure NM, Deverdun J, *et al*. Role of fronto-striatal tract and frontal aslant tract in movement and speech: an axonal mapping study. *Brain Struct Funct* 2014.
- 10 Torres-Perales AM, Martínez-García FA, Andréu-Reinón ME, *et al*. Síndrome opercular producido por infarto en un solo opérculo cerebral en un paciente con antecedente de infarto cerebeloso. *Rev Neurol* 2013;56:495–6.

Learning points

- ▶ The presence of bilateral voluntary facial, pharyngeal, lingual and masticatory paralysis with automatic–voluntary movement dissociation defines Foix-Chavany-Marie syndrome (FCMS), also known as anterior opercular syndrome.
- ▶ The majority of cases of FCMS are caused by bilateral vascular lesions of the anterior operculum.
- ▶ Unilateral opercular lesions, often coexisting with contralateral white matter lesions, can very rarely cause FCMS.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow