

Case Report

Unilateral Opercular Infarction Presenting with Foix-Chavany-Marie Syndrome

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A 76-year-old man with a history of pontine, cerebellar infarction suddenly became speechless during the procedure of percutaneous coronary intervention. On examination, he was unable to close his mouth voluntarily, but spontaneous closing was preserved when smiling. He had anarthria and hypophonia, although his comprehension was preserved. He also had a severe dysphagia. Radiological studies revealed an acute stroke in the left anterior operculum, indicating Foix-Chavany-Marie Syndrome (FCMS) caused by a unilateral opercular lesion. Pathophysiology of the previous cases reported as unilateral FCMS remains controversial, but in our case, it could be delineated by the combination of the new lesion in the unilateral operculum and the old one in the contralateral pons. Since FCMS is not only related to biopercular lesions, we should consider thorough radiologic examination to clarify its anatomic basis. **Key Words:** Stroke—psuedobulbar palsy—automatic voluntary dissociation.

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Case Report

A right-handed 76-year-old man with a history of pontine and cerebellar infarction suddenly became speechless during a percutaneous coronary intervention and was referred to our department with suspected cerebral ischemia. On examination, his mouth was half open and he could not close it when requested, but could do so spontaneously when smiling. He had central facial paralysis on the right side. He had anarthria and hypophonia and could not say anything but “ah,” although his comprehension was preserved. He also had severe dysphagia. Laryngoscopy revealed intact vocal cords. He had slight

weakness in the extremities on the right side, which was noticeable in his hands.

Three hours after onset of symptoms, magnetic resonance imaging (MRI) with diffusion-weighted imaging (DWI) revealed mild hyperintensity in the left anterior operculum, indicating acute stroke (Fig 1A). T2-weighted MRI (T2WI) revealed old infarctions in the right ventral paramedian pons and left cerebellar hemisphere (Fig 1B). Along with aspirin and clopidogrel prescribed as a postcoronary stenting regimen, intravenous heparin was initiated immediately. DWI and T2WI performed again 5 days after onset clearly showed the new lesion (Fig 1C and D). ¹²³I-IMP single photon emission computed tomography (SPECT) and ¹⁸F-fluorodeoxy glucose positron emission tomography showed significantly decreased blood flow (Fig 1E and F) and glucose uptake (Fig 1G and H) in the left anterior operculum, consistent with the stroke lesion. No obvious abnormalities in the contralateral hemisphere were detected.

The patient's impaired jaw movement and muscle weakness resolved within 2 months. Dysphagia also

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Received July 10, 2012; revision received August 21, 2012; accepted August 23, 2012.

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1052-3057/\$ - see front matter

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<http://dx.doi.org/10.1016/j.jstrokecerebrovasdis.2012.08.015>

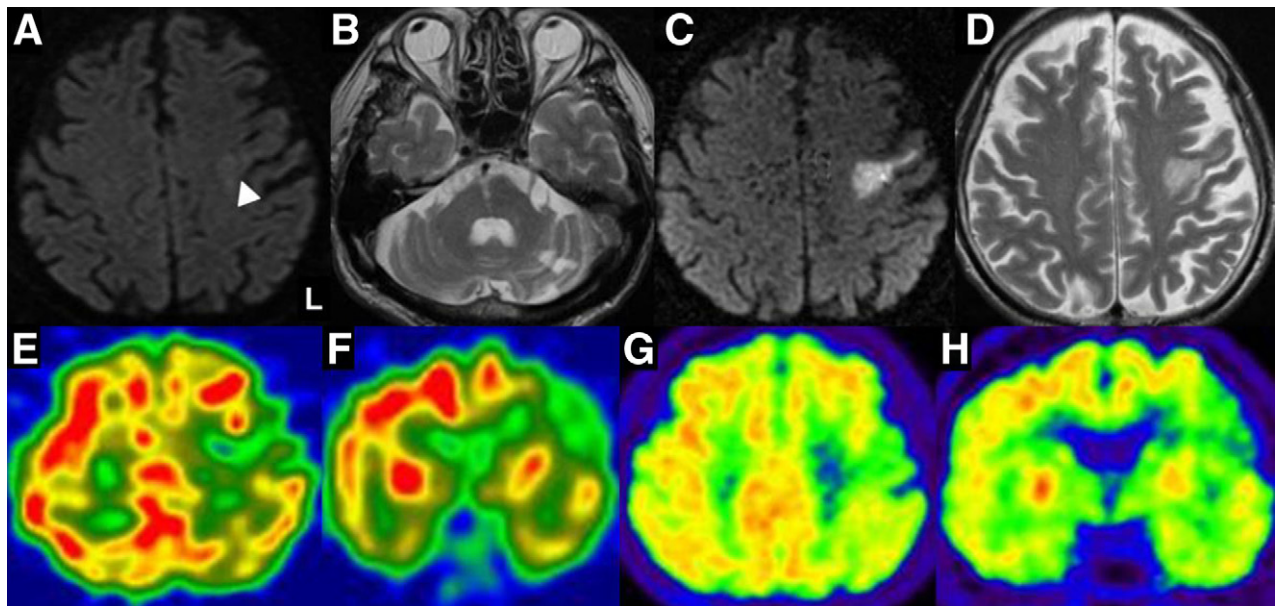


Figure 1. (A) DWI showing soft hyperintensity (arrowhead) in the left anterior operculum. (B) T2WI revealing old infarctions in the right ventral paramedian pons and the left cerebellar hemisphere. (C, D) DWI and T2WI taken 5 days after the onset indicating acute stroke in the left anterior operculum. (E–H) Axial and coronal SPECT and ¹⁸F-fluorodeoxy glucose positron emission tomography indicating decreased blood flow and glucose uptake within the new lesion, respectively. No significant changes are evident in the contralateral hemisphere.

improved after 5 months of rehabilitation, but loss of speech was not ameliorated.

Discussion

Foix-Chavany-Marie syndrome (FCMS), originally reported by Magnus¹ and later described in detail by Foix et al,² is a cortical type of pseudobulbar palsy characterized by loss of voluntary control of facial, pharyngeal, lingual, and masticatory muscles with preserved reflexive and automatic functions. FCMS is most commonly induced by sequential infarcts to bilateral anterior opercula or subcortical insular regions.^{3–5} Whereas neurons that lie adjacently in the operculum project supranuclear fibers to

the cranial nuclei for the voluntary movement of facial, pharyngeal, lingual, and masticatory muscles, emotional movement of these muscles is controlled by alternative pathways that run from the amygdala and lateral hypothalamus to the brainstem via the medial forebrain bundle and dorsal longitudinal fasciculus. This is usually considered an anatomic basis for the “automatic voluntary dissociation” seen in FCMS. Fibers from the amygdala do not project directly to the motor nuclei of the cranial nerves, but rather project to the lateral tegmentum of the pons, where interneurons for these nerves are located. Crosslinks between these fibers and neurons remain to be elucidated, obscuring the precise mechanism of FCMS.

Table 1. Previously reported cases of FCMS caused by a single lesion

Patient	Reference	Age, years/sex	Handedness	Examinations for localization	Lesion	Other symptoms
1	Magnus (1837) ¹	25/F	ND	Necropsy	Left anterior operculum	-
2	Starkenstein et al (1988) ⁸	55/M	Right	Computed tomography, necropsy	Right insula, frontotemporoparietal operculum	-
3	Kutluay et al (1996) ⁹	73/F	Right	MRI, SPECT	Left perisylvian region (MRI), right parietal cortex (SPECT)	Right hand weakness
4	Morgas et al (2007) ⁷	36/M	ND	MRI	Right operculum	Left upper extremity weakness
5	Morgas et al (2007) ⁷	61/M	ND	MRI	Left operculum	-

Abbreviation: ND, not documented.

Patients with FCMS caused by unilateral opercular stroke with contralateral white matter lesion and patients with bilateral subcortical infarcts have been reported.^{6,7} In these patients, pathophysiology was attributed to involvement of bilateral corticobulbar tract and the possible existence of a separate pathway for emotional expression apart from the lesion in the deep white matter. FCMS apparently resulting from a single stroke lesion has been reported as well (Table 1).^{1,7-9} However, these cases were reported before the advent of MRI or nuclear scanning,^{1,8} along with 1 patient with what initially appeared to be a unilateral lesion on MRI but that later proved to be associated with hypoperfusion of the bilateral opercula by conducting brain SPECT.⁹ As such, these reported cases lack rigorous radiologic assessment data and detailed descriptions of neurologic features, and thus the existence of unilateral FCMS remains controversial.

As long as pseudobulbar palsy (ie, bilateral involvement of the corticobulbar tract) is essential for the diagnosis of FCMS, attributing this patient's FCMS to a single lesion is problematic, although definitive abnormalities in the contralateral anterior operculum were not radiologically demonstrated in this patient. Rather, based on previous studies describing a volitional type of central facial palsy caused by stroke in the upper pons,^{10,11} we speculate that along with the new lesion, the old lesion in the pons involving the corticobulbar tract but sparing the lateral tegmentum resulted in pseudobulbar palsy causing anarthria, dysphagia, and a discrepancy between voluntary and automatic facial muscle movement. None of the previously reported cases presented in Table 1 mentioned brainstem lesions, but we suspect that some of them could be convincingly described by a combination of unilateral opercular and brainstem lesions.

When encountering stroke survivors with FCMS, we should keep in mind that this unique syndrome is related not only to biopercular lesions, and should consider

a thorough radiologic examination to delineate its neuro-anatomic basis.

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