

Bilateral perisylvian infarct: a rare cause and a rare occurrence

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ABSTRACT

Foix-Chavany-Marie opercular syndrome is a severe form of pseudobulbar palsy occurring due to bilateral anterior opercular lesions. We report a case of a 51-year-old man with sudden onset of inability to speak and dysphagia, and a history of synovial sarcoma of the right hand. Detailed language evaluation was normal. The patient had right upper motor neuron facial paresis and absent gag reflex bilaterally. Magnetic resonance (MR) imaging revealed acute and subacute infarcts involving the bilateral insular cortex. Two-dimensional echocardiography and cardiac MR imaging showed a mobile mass in the left atrium attached to the interatrial septum, which was likely a myxoma. Chest radiograph and computed tomography imaging of the chest revealed multiple cannonball shadows that were suggestive of secondaries in the lung. The probable cause of the cerebral lesions was the mass lesion in the heart or metastatic lesions from the synovial sarcoma. The cardiac surgeon and surgical oncologist recommended palliative care.

Keywords: cardioembolic stroke, perisylvian infarct, synovial sarcoma

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INTRODUCTION

The opercular syndrome, first described by Magnus in 1837, also referred to as Foix-Chavany-Marie syndrome (FCMS), is named after French authors who reported the syndrome in 1926.^(1,2) Alternatively, it is known as facio-labio-pharyngo-glosso-laryngo-brachial paralysis or the cortical type of pseudobulbar paralysis. The opercular syndrome is characterised by a loss of voluntary control of facial, lingual, pharyngeal and masticatory muscles in the presence of preserved reflexive and automatic functions of the same muscles.^(3,4) It may be congenital or acquired, persistent or intermittent. In acquired cases, vascular insults, such as ischaemic infarcts in the perisylvian region involving the insular cortex or corticobulbar tracts, constitute a major cause.⁽⁴⁾ Previous studies have highlighted cases of sequential infarcts

occurring bilaterally in the perisylvian area. This is the first report of a patient having bilateral near-simultaneous perisylvian infarcts.

CASE REPORT

A 51-year-old right-handed man with a history of synovial sarcoma of the right hand was operated in October 2002 for disarticulation of the medial three fingers. He presented to the outpatient clinic in April 2008 with a sudden onset of inability to speak and dysphagia of solids and liquids for a duration of four days. The patient had no history of focal deficits, loss of consciousness, seizures or any vascular risk factors. On examination, his vital signs were within normal limits. He was in significant distress and occasionally cried when questions were asked. Language testing showed normal comprehension and writing but no word output and repetition. Examination of the central nervous system (CNS) revealed normal fundus and extraocular movements. Facial sensations and muscles of mastication were normal. There was evidence of right upper motor neuron (UMN) facial palsy, with absence of a gag reflex bilaterally. The patient's spontaneous emotional movements were present and tongue movement was normal, but his voluntary control was lost. The rest of the neurological examination was non-contributory.

Routine haematological work-up was negative. The patient was subjected to advanced magnetic resonance (MR) imaging of the brain, which showed an acute infarct involving the left insular cortex extending to the centrum semiovale and corona radiata, with restriction on diffusion-weighted imaging (DWI) and no enhancement on contrast. An early subacute infarct involving the right insular cortex and extending to the corona radiata, with mild restriction on DWI, was seen upon enhancement and contrast, which was suggestive of bilateral perisylvian infarcts (Fig. 1).

In view of the multiple infarcts, the patient was subjected to two-dimensional echocardiography, which confirmed the presence of a 4.0 cm × 2.5 cm mobile mass in the left atrium attached to the interatrial septum. A radiograph and computed tomography imaging of the chest revealed multiple large, round lesions in the lung bilaterally (Figs. 2a & b). With the diagnostic possibility

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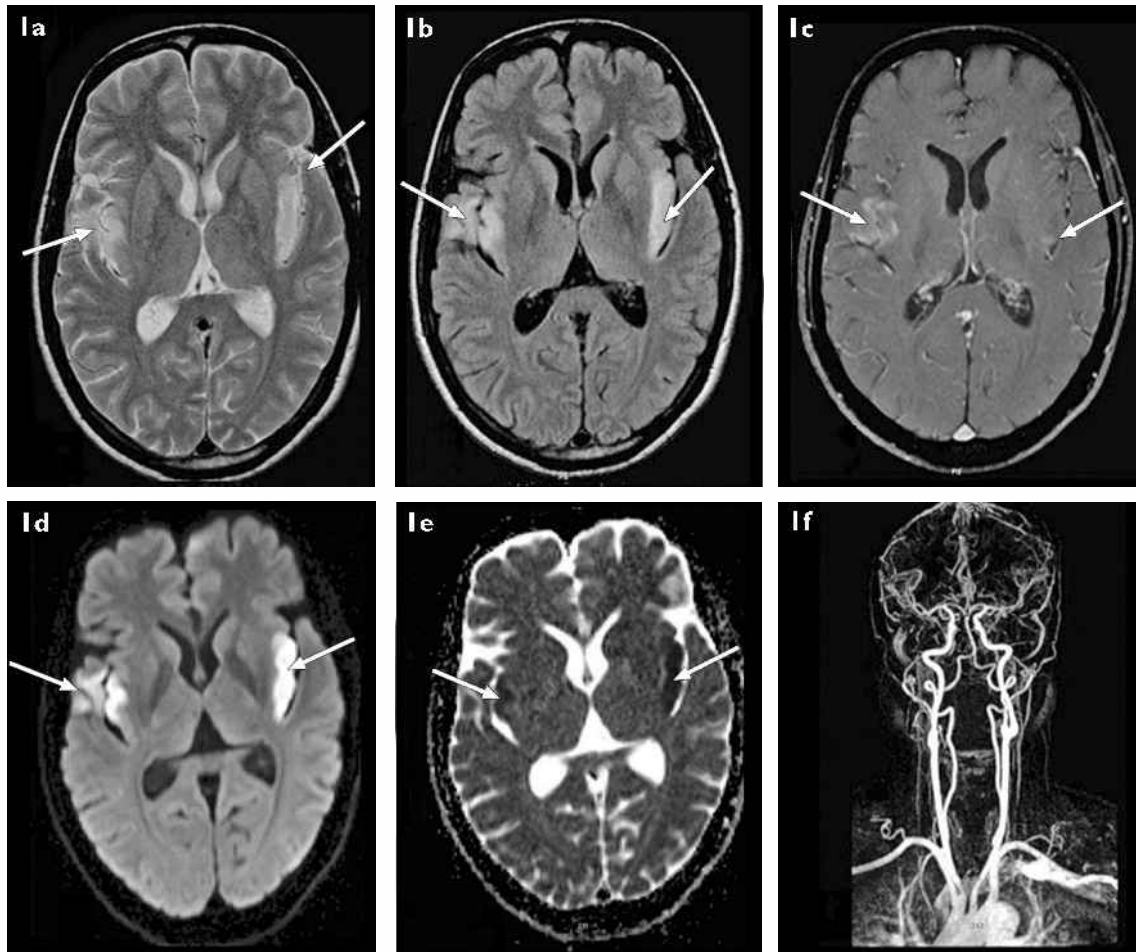


Fig. 1 Axial T2-W MR images (a) show bilateral insular cortex hyperintensity (arrows); with (b) the lesion appearing hyperintense in FLAIR sequence (arrows). (c) Post-contrast fat suppressed T1-W image shows enhancement in the right-sided lesion (arrows), with no enhancement in the left insular lesion. (d) Diffusion-weighted MR image and (e) apparent diffusion coefficient map show diffusion restriction within the lesion (arrows), with the MR image suggestive of bilateral insular infarcts. (f) MR angiography was normal.

of a metastatic left atrial mass or a left atrial myxoma, cardiac MR imaging (Figs. 2c & d) was planned. The results revealed a well-defined sessile lobulated lesion measuring 4.8 cm × 3.1 cm, which appeared to be mildly hyperintense on the T1-weighted image, with a brilliant contrast enhancement of the mass. On cine MR imaging, the mass was minimally protruding into the left ventricle without causing any obstruction. In view of the extensive metastasis, the possibility of cardiac metastatic sarcoma was considered. However, since the mass was sessile and attached to the interatrial septum, the possibility of a left atrial myxoma could not be ruled out.

The opinions of the cardiac surgeon and surgical oncologist were sought, and in view of the presence of extensive metastasis in the lungs, palliative care was advised. The diagnosis of the atrial mass could not be confirmed, as no surgical intervention was possible. Meanwhile, the patient was started on aspirin 150 mg once daily and atorvastatin 10 mg at bedtime. He was also evaluated and treated by a speech therapist. With

the above measures implemented, our patient showed significant improvement over the next five days. By Day 7 of admission, his nasogastric tube was removed. At the time of discharge, the patient was ambulatory and able to communicate. He still had residual facial paralysis, and his gag reflex remained impaired bilaterally. He was able to swallow food but had difficulty swallowing liquids.

DISCUSSION

Since the original case report of the opercular syndrome presented by Magnus in 1837, cases of FCMS to date have been described mostly by French authors.^(1,2) A literature review of 62 FCMS into cases by Weller⁽³⁾ classified FCMS into five clinical types: (a) the classical and most common form associated with cerebrovascular disease; (b) a subacute form caused by CNS infections; (c) a developmental form most often related to neuronal migration disorders; (d) a reversible form in children with epilepsy; and (e) a rare type associated with neurodegenerative disorders.

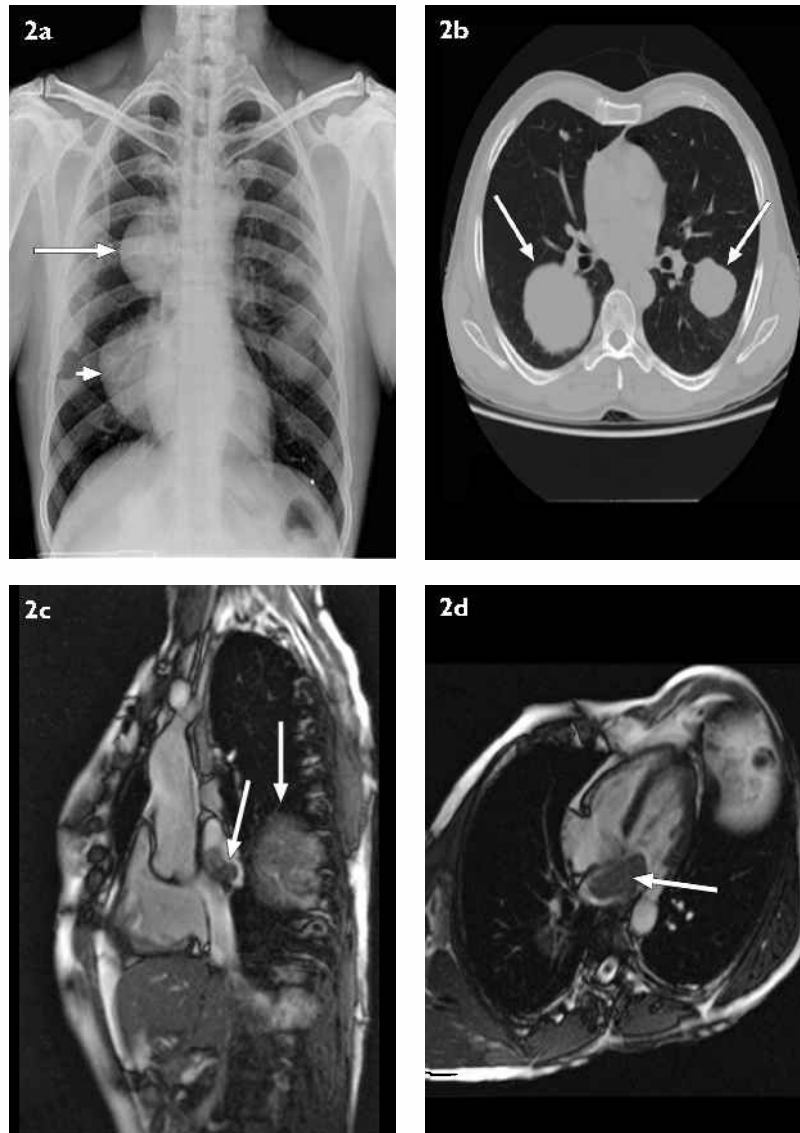


Fig. 2 (a) Radiograph of the chest (posteroanterior view) shows large round opacities (cannonball shadows) in the right paracardiac region (arrow) and left mid zone (arrowhead). (b) CT image of the chest shows round opacities (arrows) in both lung fields. (c & d) Cardiac MR images show a well-defined sessile lobulated lesion arising from the interatrial septum (arrows).

Our patient had the classical and the most common form of FCMS. He had acute onset right UMN facial palsy, weakness of the bulbar muscles and anarthria with normal comprehension. Emotional movements were present, but there was a loss of voluntary control of the facial and bulbar muscles. Emotional lability was present in the form of frequent crying spells, which improved rapidly over a period of two months. MR imaging of the patient's brain showed bilateral infarcts in the perisylvian areas, most of which were concentrated in the grey matter.

The occurrence of bilateral perisylvian infarcts is well documented in various case reports,^(5,6) but they have all described sequential infarcts. One report discussed a case of crossed aphasia after a right corona radiata infarct

and an old left hemispheric infarct.⁽⁷⁾ The present report is the first case of near-simultaneous infarcts in the perisylvian area. Considering our patient's past history of synovial sarcoma and the occurrence of simultaneous acute infarcts in both hemispheres, the possibility of a proximal source of the embolism was considered. His echocardiogram showed a mobile mass in the left atrium, suggesting a possible left atrial myxoma or a metastatic atrial sarcoma.

In the case of an intracardiac mass, the differential diagnosis of myxoma primarily encompasses other benign and malignant primary heart tumours, metastatic tumours and thrombi. Metastatic tumours to the heart are 20–40 times more common than primary tumours.⁽⁸⁾ Atrial myxoma, the most common benign cardiac tumour,

is found more commonly in young adults with stroke or transient ischaemic attack (1 in 250) than in older patients with these problems (1 in 750).⁽⁹⁾ The annual incidence is 0.5 per million population,⁽¹⁰⁾ with 75% of cases occurring in the left atrium. There is a 2:1 female preponderance,⁽¹¹⁾ and the age at onset is typically 30–60 years. Embolism occurs in 30%–40% of patients with myxomas,⁽¹²⁾ most are located in the left atrium, systemic embolism being particularly frequent.

The chest radiograph of our patient showed multiple cannonball shadows, which were suggestive of secondaries in the lung, while cardiac MR imaging was suggestive of atrial myxoma. The findings were unlike those of a primary cardiac sarcoma, but its possibility could not be ruled out, as a biopsy confirmation could not be done. Hence, the aetiology was most likely primary synovial sarcoma of the hand, resulting in a secondary endocardial lesion in the heart and secondaries in the lung. We could not confirm the exact cause of bilateral perisylvian syndrome in our patient; it could be attributed to the secondaries in the lung that had metastasised to the CNS or primarily the atrial myxoma. Due to the stage and extent of the disease, a biopsy was not done. In view of the patient's condition, we recommended palliative care instead.

The occurrence of bilateral perisylvian infarcts, although well described in the literature, is a rare entity and requires a high index of suspicion for diagnosis. A thorough work-up and advanced imaging, such as DWI or apparent diffusion coefficient, would facilitate the diagnosis of the condition.

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