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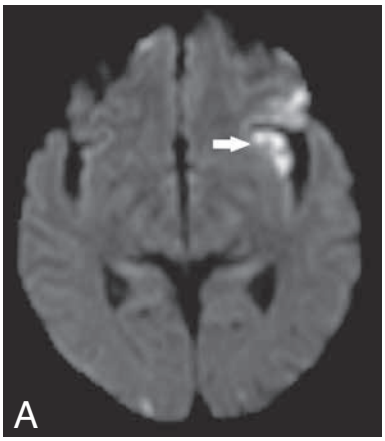
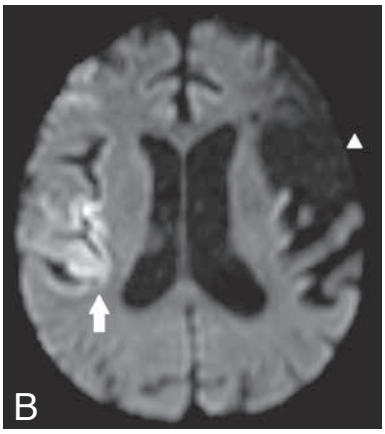
IMAGES IN CLINICAL RADIOLOGY





Foix-Chavany-Marie or opercular syndrome

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A 69-year-old male was admitted to the emergency department with suspicion of a recurrent cerebrovascular accident (CVA). Medical history included a prior CVA in the vascular supply area of the left middle cerebral artery in 2010, due to a patent foramen ovale, causing cardiogenic emboli (Fig. A). The patient was respectively treated with anticoagulation and percutaneous closure of the foramen; a significant motoric aphasia persisted. At a neurologic clinical investigation, besides the known aphasia, apraxia of the tongue, dysarthria and a limited left hemisyndrome were noted with predominant paresis of the left hand and impairment of the left visual field. Imaging was requested, with CT only showing age-related atrophic changes and sequelae of left middle cerebral artery infarction. MRI revealed additional diffusion restriction in the right opercular region, suggestive of recent stroke (Fig. B).

Holter monitoring revealed paroxysmal atrial flutter, confirming the suspicion of cerebral infarction as the basis of cardiogenic emboli. Following the recent ischaemia, the speech-language pathologist noticed the patient was not able to eat, but when food was deposited in his mouth, no eating difficulties or swallowing impairment were observed. Patient was treated with anticoagulant regimen, percutaneous entero-gastrostomy, anti-epileptic drugs and neurologic revalidation.

Comment

Facio-labio-pharyngo-glosso-masticator paralysis, also known as Foix-Chavany-Marie or opercular syndrome, consists of dissociation between automatic and voluntary movements. Bilateral lesions in the operculum cause a loss of volitional control of lingual, pharyngeal, facial and masticatory musculature, with preserved automatic movement. Typical manifestations include difficulties in mouth opening, tongue protrusion, chewing and swallowing food as well as speech impairments. Opercular syndrome may be congenital or acquired (as is the case in our patient), and intermittent or persistent. Possible causes of opercular syndrome include cerebrovascular disease (acute setting), CNS infections (subacute form), neuronal migration disorders (developmental), neurodegenerative disorders or epilepsy (reversible). Most reported cases, like our case, can be attributed to thrombo-embolism of middle cerebral artery branches, which irrigate the operculum.

The operculum represents the cortex surrounding the insula, and can be divided into three parts: the frontal operculum, the frontoparietal operculum and the temporal operculum; with variable involvement of the subcortical white matter. Bilateral lesions in these areas clinically mimic a pseudobulbar palsy in the distribution of the 5th, 7th, 9th, 10th and 12th cranial nerves. The so-called "automatic-voluntary dissociation" can be elucidated by analysis of functional neuro-anatomy. Volitional control of the facial, oral and pharyngeal musculature demands intact motor cortices and pyramidal pathways; pathology in these areas leads to a selective palsy of voluntary use of these muscle groups. Emotional or spontaneous use of these muscle groups however require intact extrapyramidal pathways as well as parts of the hypothalamus and thalamus, thus explaining the dissociation between automatic and voluntary movements.

Some reports have also been made of patients with bilateral opercular syndrome who presented with only a unilateral lesion; it is believed that an occult underlying lesion on the contralateral side would account for these manifestations.

Imaging thus plays an important role in detection of opercular lesions, most frequently by detection of recent stroke (by way of diffusion weighted imaging), allowing adequate management.

Reference

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