Anterior Biopercular Syndrome Caused by Unilateral Infarction

Sindrome Biopercular Anterior Devido a Enfarte Unilateral

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ABSTRACT

The anterior biopercular syndrome is characterized by facio-pharyngo-glosso-masticatory diplegia, with automatic dissociation of movements. It generally translates bilateral opercular lesion, often of vascular etiology. There are very few cases described with unilateral lesions. We present the case of a patient with a bilateral anterior opercular syndrome caused by unilateral infarction.
Keywords: Deglutition Disorders; Cerebral Infarction.

RESUMO

A síndrome biopercular anterior caracteriza-se por diplegia facio-faringo-glosso-mastigatória, com dissociação dos movimentos automáticos. Traduz geralmente lesão opercular bilateral, frequentemente de etiologia vascular. Há casos raros descritos de lesão unilateral. Apresentamos o caso de uma doente com uma síndrome opercular anterior bilateral causada por um enfarte unilateral.
Palavras-chave: Disturbios da Deglutição; Enfarte Cerebral.

INTRODUCTION

Anterior opercular syndrome, or Foix-Chavany-Marie syndrome, was first reported by Magnus in 1837[1] and later detailed by Foix et al[2] in 1926. It is characterized by facio-pharyngo-glosso-masticatory diplegia, with preservation of reflex and automatic functions of these muscles.[2] The patient cannot open or close the mouth, protrude the tongue or close the eyes to orders, but can smile, cry, yell or yawn automatically.[3] Ischemia is the most frequent cause[3,4] and it is generally due to bilateral opercular lesions, but bilateral subcortical lesions have also been reported.[4,5] Much less commonly, unilateral lesions can cause a biopercular syndrome.[3,6,7]

We present the case of a patient with a bilateral anterior opercular syndrome caused by unilateral infarction.

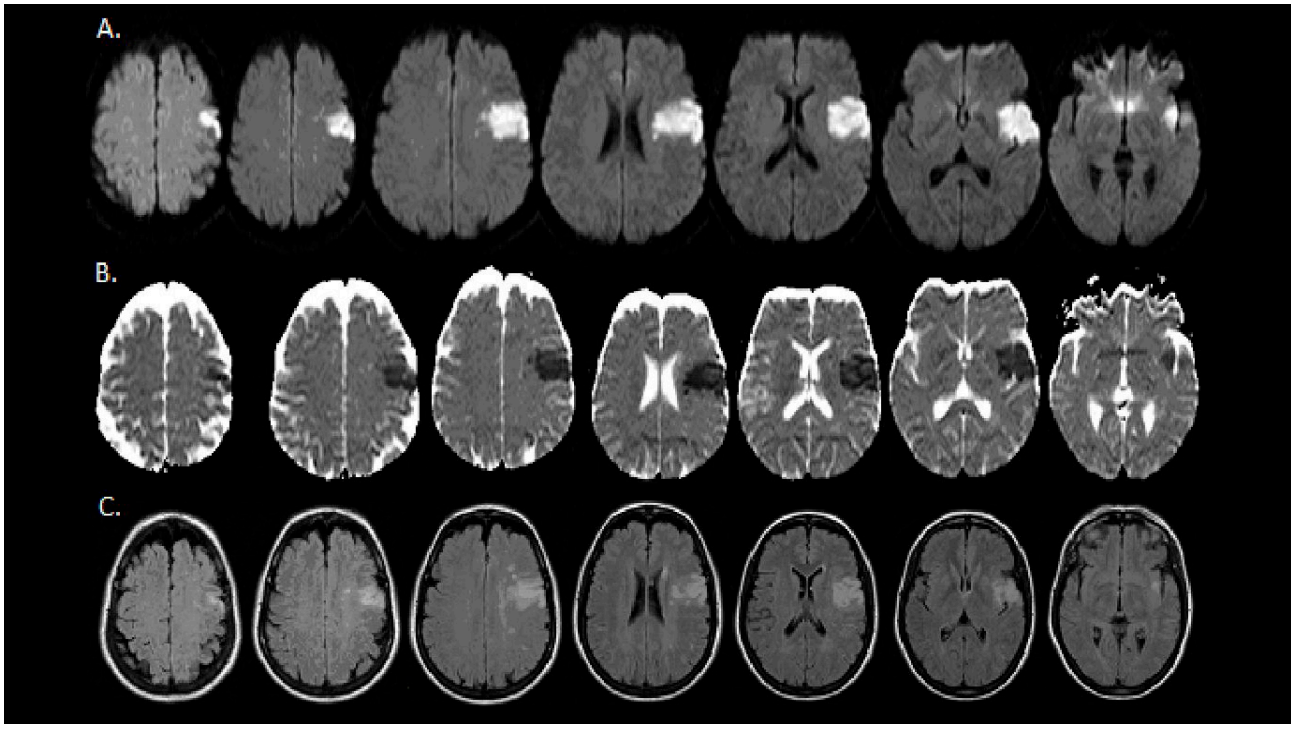

Figure 1 – Axial diffusion-weighted (A.), apparent diffusion coefficient (B.) and axial fluid attenuated inversion recovery (C.) magnetic resonance imaging showing acute, left anterior opercular infarct.

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CASE REPORT

A 47 years old female patient, right-handed, with a history of smoking and of two miscarriages, woke up one morning with speaking and swallowing difficulties. A week before she had felt paresthesia in the right hemiface and upper limb. On neurological examination the patient was awake, had good understanding of spoken and written language, but was unable to produce any sound. She had difficulty imitating simple gestures such as opening the mouth or protrude the tongue. Smile or yawn reflexes were preserved. She presented syntactic and praxis errors in her handwriting. She had a bilateral palate paralysis with anesthesia in this area and in the oropharynx, absence of palatal and gag reflexes, bilateral vocal cord palsy and dysphagia. She also had mild right hemiparesis, predominantly brachiofacial, with generalized osteotendinous hyperreflexia but without Babinski sign.

Magnetic resonance imaging (MRI) showed an acute left opercular infarction (Fig. 1). No other acute neither non-recent lesions were detected.

Routine blood tests, immunologic and pro-thrombotic studies, electrocardiogram and echocardiogram were normal. Doppler sonography of cervical arteries and the angio-MRI showed an occlusion of the proximal third of the left carotid artery and digital subtraction- angiography confirmed a carotid dissection.

An anterior biopercular syndrome was diagnosed in the dominant hemisphere secondary to cerebral infarction caused by carotid dissection.

The patient was hypocoagulated with acenocumarol and recovered significantly. Four years later she had only slight language changes, and cervical Doppler sonography showed recanalization of the left internal carotid artery.

DISCUSSION

In the anterior opercular syndrome the connections between the motor cortex and the V, VII, IX, XII cranial nuclei and brainstem are interrupted bilaterally. Emotional and spontaneous movements, dependent on extrapyramidal connections, thalamus and hypothalamus, are preserved (automatic-voluntary dissociation).[4]

In this case the syndrome was incomplete because the V and VII nerves were not bilaterally affected, and curiously the lesion was unilateral. Although the classical neuroanatomical basis of the opercular syndrome involves bilateral lesions[2], there are rare cases of unilateral lesions causing a bilateral palsy of the facial, chewing, tongue, pharynx and larynx muscles[3,6,7](Table 1). Several hypotheses have been suggested to explain this phenomenon; one possibility is the existence of an anatomical variant, so that the corticobulbar tract representation is predominantly unilateral.[3]

Regarding aetiology, cerebrovascular disease (mainly ischemic) is the most frequent cause.[3,4] However other disorders were also described: encephalitis, brain tumours, cortical dysplasia, vasculitis, epilepsy, degenerative diseases and traumatic brain injury.

Several authors identified poor prognosis in the recovery of swallowing and especially of verbal language.[4,6] In two reported cases of unilateral lesions with bilateral manifestations the prognosis was good[3,7] and the same occurred in our patient.

This case illustrates a peculiar bilateral anterior opercular syndrome caused by unilateral brain infarction. Unlike the cases due to bilateral damage, functional outcome was good.

CONFLICT OF INTERESTS

None stated.

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Table 1 - Previously reported cases of biopercular syndrome caused by unilateral lesion.

Reference	Age, years/sex	Handedness	Cerebral imaging	Lesion	Outcome
Starkenstein et al.[7] (1988)	55/M	Right	TC	Right operculum and insula	Marked improvement of jaw movement, muscle weakness and dysphagia; persistent anarthria
Moragas Garrido et al.[3] (2007)	61/M	ND	MRI	Left operculum	Absence of dysphagia and improvement of facial palsy and dysarthria
Moragas Garrido et al.[3] (2007)	36/M	ND	MRI	Right operculum	Improvement of dysarthria, dysphagia and brachial weakness; persistent facial diplegia
Giraldo-Chica et al.[6] (2010)	76/F	Right	MRI, SPECT	Right operculum	Improvement of dysphagia, persistent anarthria

REFERENCES

1. Magnus A. Fall von Aufhebung desWillenseinflusses auf einige Hirnnerven. Arch Anat Physiol Wiss Med. 1837;258-66.
2. Foix C, Chavany JA, Marie J. Diplègie faciolinguomasticatrice d'origine cortico souscorticale sans paralysie des membres. Rev Neurol. 1926;33:214-9.
3. Moragas Garrido M, Cardona Portela P, Martínez-Yélamos S, Rubio Borrego F. Heterogeneidade topográfica del síndrome de Foix-Chavany-Marie. Neurología. 2007;22:333-6.
4. Millán Pa, Montes MI, Uribe CS, Cabrera D, Arboleda A. Síndrome biopercular: presentación de los casos y revisión de la literatura. Biomédica. 2008;28:183-90.
5. Posterato L, Pezzoni F, Varalda E, Fugazzo G, Mazzucchi A. A case of unilateral opercular syndrome associated with a subcortical lesion. J Neurol. 1991;238:337-9.
6. Giraldo-Chica MM, Ramírez JD, Uribe C, Lopera F. Biopercular syndrome caused by unilateral ischemia. Report of one case. Rev Med Chil. 2010;138:341-5.
7. Starkstein SE, Berthier M, Leiguarda R. Bilateral opercular syndrome and crossed aphemia due a right insular lesion: A clinicopathological study. Brain Lang. 1998;34:253-61.