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Importance of Rapid Clinical Recognition of the Anterior Opercular Syndrome (Foix-Chavany-Marie Syndrome): A Case Report

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Keywords
Acute anarthria · Anterior opercular syndrome · Foix-Chavany-Marie syndrome · Facio-pharyngo-glossal diplegia · Automatic-voluntary movement dissociation

Abstract
We have described a 55-year-old woman with the anterior opercular syndrome (Foix-Chavany-Marie syndrome). The clinical presentation included acute onset of bilateral facial palsy and anarthria. Immediate MRI of the brain revealed acute ischemia in the right opercular region and sequelae after a previous infarction involving the left opercular region. The patient was treated with intravenous thrombolysis resulting in full recovery. The anterior opercular syndrome is rare, and the most common reason is sequential stroke. We emphasize the importance of recognizing this syndrome early, and in all cases, consulting a revascularization center immediately.

Introduction

Anterior opercular syndrome (AOS), also known as Foix-Chavany-Marie syndrome is a rare type of pseudobulbar palsy, attributed to bilateral operculum lesions [1]. The first case was reported by Magnus in 1837 [2], who described a patient with mutism and bilateral loss of voluntary movement of the musculature innervated by the cranial nerves V, VII, IX, X, and

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XII, with preserved reflexes, automatic and emotional functions referred to as automatic-voluntary dissociation [2]. Affected individuals typically have anarthria and dysphagia, with preserved ability to close their eyes during sleep as well as involuntarily yawn, smile, laugh, and cry [1, 2], AOS has been reported in patients suffering from neuroinfection [3], neuroinflammatory or neurodegenerative diseases [4, 5], brain tumors [6], traumatic brain injury [7], normal pressure hydrocephalus [8], osmotic demyelination [9], but mostly due to acute stroke [10]. It is therefore extremely important to recognize this syndrome as it can have consequences for the further acute treatment, including early revascularization therapy. Here, we present a challenging case of AOS in a woman suffering an acute stroke, who rapidly recovered after intravenous (IV) thrombolysis.

Case Report

A 55-year-old woman was admitted to the thrombolysis section with an acute onset of anarthria and weakness of her left arm. She had a previous history of deep vein thrombosis and lung embolus 2 years prior, a left-sided middle cerebral artery M2 infarction with persistent discrete aphasia 9 months prior, and subsequent closure of patent foramen ovale 6 months prior and was taking clopidogrel 75 mg and atorvastatin 40 mg daily. The patient had undergone diagnostic laparoscopy $2^1/_2$ weeks prior on suspicion of appendicitis, which had revealed a tumor in the right ovary, requiring further evaluation for ovarian cancer (which was later confirmed). Thus, the patient had been on tinzaparin 4,500 IU daily up until 5 days prior to admission to the thrombolysis section.

On admission, the patient was dyspneic with stridor in the supine position. The neurological examination revealed anarthria, near-total facial diplegia with minimal voluntary movement of her forehead, bilateral palate paralysis, dysphagia, inability to protrude the tongue, a brisk jaw reflex, discrete flexion of the left elbow and pronation of the forearm, and bilaterally reduced fine motor function. The deep tendon reflexes were bilaterally hyper-reflexive in the upper extremities and brisk in the lower extremities. The patient could follow verbal commands directed at her limb movements and was able to gesticulate. She could write with no signs of aphasia. Moreover, she had no neglect and showed no signs of ophthalmoplegia. She smiled involuntarily, for instance when asked to whistle. She could move her tongue from side to side and had near-normal neck movement with lightly reduced strength on antero-flexion. There were no sensory deficits, no ataxia, and her gait was unremarkable.

On admission, the initial computed tomography (CT) scan of the brain showed substance loss from an old infarction in the left frontal operculum (Fig. 1a). CT angiography was normal, but the CT perfusion scan showed increased mean transit time with corresponding reduced cerebral blood flow and slightly increased cerebral blood volume in the right frontal operculum, indicating a penumbra region (Fig. 1b–d).

At this time, the patient and family did not accept IV thrombolysis due to the slightly increased risk of bleeding. Additional CT of the lungs due to dyspnea revealed a small peripheral lung embolus in the left lung. The patient was again offered IV thrombolysis which she accepted.

After 24 h, the patient significantly recovered, with only discrete dysarthria and bilateral palsy in the lower facial musculature, and CT showed no signs of a lesion in the right cerebral hemisphere or bleeding. She was discharged from the hospital after 1 week with a facial rehabilitation plan.

Discussion

The opercular regions are in the facio-pharyngo-glosso-masticatory area of the premotor- and primary motor cortex. Sequential supranuclear bilateral ischemic lesions involving the cortex or the corticobulbar projections from the opercular regions to the cranial nuclei (CN) in this patient accounted for her symptoms of discrete aphasia (left M2 infarction involving the Broca area), facial diplegia (CN. VII), anarthria, palatal paralysis, reduced tongue mobility (CN. IX, X, XII), and slightly reduced neck antero-flexion (CN. XI). The involuntary smiling when asked to whistle is a sign of automatic-voluntary dissociation, a hallmark of AOS.

The neuroanatomical basis of the patients' automatic-voluntary dissociation is thought to lie within preserved amygdala-latero-tegmental and lateral hypothalamic projections to the brainstem.

The most important non-stroke-related differential diagnostic considerations in this case include myasthenia gravis and Guillain-Barré syndrome (the patient was initially dyspneic), which requires considering the possibility of acute versus subacute onset of symptoms, bulbar palsy without automatic-voluntary dissociation, and brisk reflexes versus areflexia.

Certain findings help differentiating AOS from other stroke syndromes in our case:

She showed no signs of impaired language comprehension and was able to perform limb-specific tasks when prompted, as opposed to patients with global aphasia. She also demonstrated involuntary movement of her cranial musculature as opposed to patients with brainstem strokes.

Clinical improvement of AOS generally depends on the underlying pathogenesis.

To the author's knowledge, less than 40 cases of AOS caused by ischemic stroke have been reported in peer-reviewed papers, and the use of thrombolysis was only reported in 3 cases [11–13]. The authors speculate that the success of thrombolysis in this case is related to quick admission to the thrombolysis section from time of symptom onset, as no signs of demarcation were found on the non-contrast CT scan, compared to visible demarcation on CT and MRI in 2 out of 3 of the published cases.

None of the differential diagnoses typically require immediate action, except neuroinfection, which can be handled in all hospitals.

If clinicians fail to recognize AOS, the time from onset to correct diagnosis may be delayed beyond the $4^1/_2$ h thrombolysis window, especially in patients presenting with bilateral distribution of symptoms instead of the more common unilateral or crossed symptoms.

In conclusion, the AOS is a rare type of pseudobulbar palsy, which is mostly seen in patients suffering from sequential strokes in the frontal operculum. It is of extreme importance to recognize this syndrome and in all cases consult the revascularization center immediately.

Statements of Ethics

None of the authors have ethical conflicts to disclose.

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

F.M. Amin has received personal fee for lecturing/advisory board from Novartis, TEVA, and Eli Lilly. H.K. Iversen has received personal fee for lecturing/advisory board from Amgen, Boehringer Ingelheim, Bayer, BMS, and Pfizer.

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Author Contributions

Substantial contribution to the conception of the work: all authors. Acquisition, analysis, and interpretation of the data: all authors. Drafting: H.A. Saidane, F.M. Amin. Critical revision for important intellectual content: all authors. Final approval of the version to be published: all authors. Agreement to be accountable for all aspects of the work: H.A. Saidane on behalf of all the authors.

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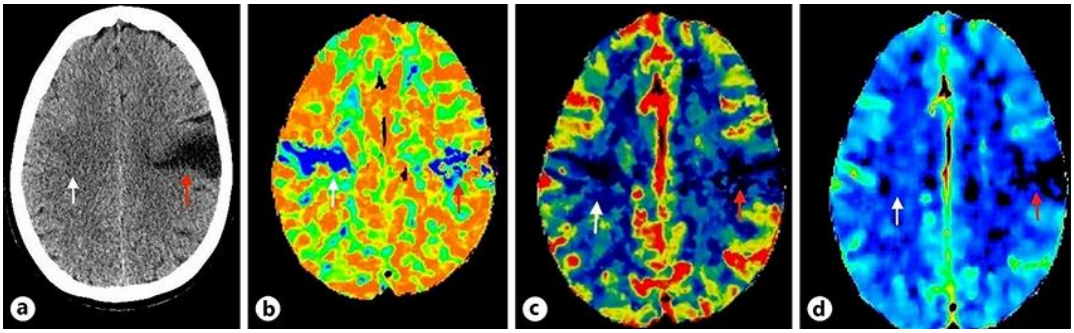

Fig. 1. **a** CT without contrast showing substance loss in the left frontal operculum due to a previous ischemic infarct (red arrow) with no visible infarct in the right opercular region (white arrow). **b** CT mean transit time showing elevated transit time in the right opercular region (white arrow) and in the left opercular region of the old infarction (red arrow). **c** CT cerebral blood flow (CBF) showing decreased cerebral flow in the right opercular region (white arrow), and in the left opercular region of the old infarction (red arrow). **d** CT CBF with slightly increased cerebral blood volume in the right frontal operculum (white arrow) compared to the left opercular region of the old infarction (red arrow).