Akinetic mutism cases due to bilateral anterior cerebral artery infarct

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ABSTRACT
Akinetic mutism is a clinical status from the bilateral lesion of nucleus caudatus or anterior cingulate gyrus. Although voluntary motions, speech, and emotional responses are completely gone, the patient may be awake with eyes opened. A 54-year-old man was brought to the emergency clinic with complaints of meaningless stares and inability to speak and walk. On neurological examination, the patient was unresponsive, although he looked awake. The tonus was increased on all extremities, especially on the left arm and leg. On his follow-up, involuntary tiny motions around his mouth were observed. Bilateral anterior cerebral artery infarct was observed on diffusion-weighted magnetic resonance imaging. This case, which may be distinguished from psychiatric charts in clinical practice, is considered to be presented as akinetic mutism due to a rare bilateral ACA infarct.

Keywords: Akinetic mutism, bilateral anterior cerebral artery territory infarct, epileptic seizure

INTRODUCTION
Strokes of the anterior cerebral artery (ACA) constitute 0.5-3% of all ischemic strokes (1). Bilateral ischaemia of the ACA is an even rarer condition. The most common symptom is the contralateral hemiparesia or monoparesis, which is usually dominant in the lower extremity. In addition, the other rare symptoms include akinetic mutism, alien hand, grasp reflex, incontinence, increased tonus and transcortical aphasia (2). In this article, we will discuss a case of bilateral infarct of ACA that attracts attention with its different clinical aspects.

CASE REPORT
A fifty-four-year-old right-handed male patient was admitted to the emergency service with complaints of inability to speak and walk and blank stare for five hours. On neurological examination, the patient was unresponsive, although he looked awake. There was no sign of neck stiffness and meningeal irritation. The patient was observed to have difficulty in starting speech. The muscle strength examination of the patient showed remarkably increased tone in all four extremities, which was more pronounced in the left lower and upper left extremities. There was a flexed posture in the upper left extremity and an extension posture in the lower extremity. Deep tendon reflexes were hypoactive in all four limbs. In addition, the plantar response was bilaterally extensor. Furthermore, the bilateral grasping reflex was observed to be positive. On the second day of hospitalization, we noticed retraction-like rhythmic movements around the mouth, especially on the right edge of the upper lip. The neurological examination parameters were found to be normal.

Diffusion-weighted magnetic resonance imaging (MRI) revealed an appearance compatible with acute infarct with gyral pattern in both the anterior cerebral artery supply area in the parasagittal region. Pathological opacification was observed in the midline and falx cerebri in contrast-enhanced series. Lacunar infarct areas were also observed near the cortex on the left side, suggesting a haemodynamic infarct (Figure 1, 2). The results of colour Doppler ultrasonography of carotid and vertebral artery were normal. Magnetic resonance angiography (MRA) showed a thickened right anterior cerebral artery, whereas it showed a non-specific appearance of the distal branches of left anterior cerebral artery. Antiaggregant and antispasmodic treatment (lioresal, Novartis, İstanbul, Turkey and rivotril; F. Hoffmann-La Roche Ltd., Basel, Switzerland) was initiated.

Invasive bilateral selective carotid angiography conducted after obtaining the informed consent form from the patient’s relatives about information sharing showed a filling failure in the left anterior cerebral artery after injection into the left internal carotid artery (Figure 3). The right anterior cerebral artery had an increase in calibration in the intracranial segment and was divided into the branches (azygos anterior cerebral artery) in both frontal lobes. There was also a 70% stenosis in the right ACA.
Transthoracic echocardiography (ECG) of the patient revealed left ventricular segmental dyskinesia with left ventricular systolic dysfunction. The ejection fraction (EF) was calculated as 48%.

Electroencephalography (EEG) was performed after attributing involuntary motions around the mouth to seizure activity. In EEG, epileptiform paroxysms were seen in the left hemisphere, especially in the left parietotemporal region (Figure 4). In the light of clinical and EEG findings, the patient was diagnosed with focal epileptic seizure and antiepileptic treatment (Tegretol, Novartis, Istanbul, Turkey (800 mg/g)) was started. The frequency of seizures of the patient has decreased considerably, albeit not decreased to zero.

Further studies revealed a homozygous mutation in the methylenetetrahydrofolate a 1298 c (MTHFR A 1298c) gene. Anticoagulation therapy was planned following the acute
DISCUSSION

The anterior cerebral artery feeds the medial surface of the cerebral hemispheres. The ACA is divided into three segments called A1, A2 and A3. The most common variation is unilateral A1 segment hypoplasia (2). The A2 segment has 3 types of variations called as accessory ACA, bihemispheric ACA, and azyzgos (unpaired) ACA (3). The reason for the rare incidence of bilateral ACA ischemia is the establishment of collateral circulation with anterior communicating arteries and thus the prevention of ischemia (1).

Bogousslavsky and Regli (4) reported in a very large series that the incidence of ACA infarct was 1.8% and that 63% of them were due to cardiogenic causes or arterial embolism. Echocardiographic examination revealed no thrombus formation, whereas the presence of left ventricular segmental dyskinesia, an ejection fraction of 48% and left ventricular systolic dysfunction poses a risk for cardiac embolism. On the other hand, MTHFR homozygous mutation may also be involved in the etiology.

Akinetic mutism was first described in 1941 by Cairns et al. (5) as a neuropsychiatric syndrome characterized by the disturbance of voluntary movement and speech with the relative conservation of attention to sound and images in an awake patient. Sudden onset is often confused with conversion disorder in patients with a history of depression and delirium in elderly patients (6). In our case, acute onset akinetic mutism could initially be considered a psychiatric condition, but the presence of accompanied perioral involuntary movements and a tonus increase (lateralization) which was more pronounced on the left side, has suggested intracranial organic pathologies. Focal epileptic seizures were attributed to a cortical infarct with the help of EEG localization.

Levodopa and dopa-agonists are usually used for treatment of akinetic mutism due to the consideration of the presence of damage in the neuronal dopaminergic pathway. In order to explain this association in patients with akinetic mutism, Yang et al. (7) used single photon emission tomography (SPECT) to view presynaptic dopamine receptors and showed that dopamine receptors are damaged in symptomatic patients, whereas improved in asymptomatic patients. Since dopaminergic therapy has not been applied in our case, it is not possible to provide information on this issue.

Nagaratnam et al. (6) classified akinetic mutism into two subtypes according to the localization of the lesion. The most common subtypes are apical akinetic mutism, where the mesencephalic and diencephalic areas are affected, and hyperpetic akinetic mutism, where adjacent frontal lobes and anterior cingulate gyrus are affected. Our case had hyperpetic akinetic mutism, where adjacent frontal lobes were affected.

CONCLUSION

Clinical diagnosis of bilateral ACA infarcts may be difficult, where the first finding may be unconsciousness. In the literature, there are cases in which patients remain comatose for up to 4 weeks (Lipschutz et al. 1991) to 1 month (Oomman and Madhusudhanan 1999) after bilateral MCA infarction (8). In such cases, diagnosis is only possible with neurological examination findings and imaging studies. Although neurologists also include cranial infarction in the differential diagnosis for a patient admitted to the emergency department with the complaints of inability to speak and blank stare, psychiatric or metabolic causes can be considered primarily due to overcrowding of emergency departments. Therefore, bilateral ACA infarcts are a condition that can be overlooked, but must be known by emergency department staff and young neurologists.

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REFERENCES


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