Case Report

The important role of family members in guiding medical professions to reach a diagnosis: Case report of under-noticed bilateral apraxia of eyelid closure

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Abstract

Apraxia of eyelid closure (AEC) is a rare disorder characterized by the inability to close the eyes on command with the preservation of the motor and sensory systems, coordination, comprehension, and cooperation. The prevalence of AEC is extremely small and the exact pathophysiological mechanisms underlying this condition remain unknown. It is, however, associated with extrapyramidal disorders. Very few cases of bilateral AEC have been reported. We report a case of an 81-year-old male patient having multiple comorbidities including neurological, respiratory, and abnormalities complicated by COVID-19 infection, who developed AEC that was noticed by the caregiver. We illustrate the clinical course leading to the diagnosis of bilateral AEC and highlight the important role of the caregiver in reporting subtle signs such as AEC.

Keywords: Apraxia of eyelid closure, eyelid disorders, neurodegeneration, Parkinson's disease, progressive supranuclear palsy

Introduction

Apraxia of eyelid closure (AEC) is a rare disorder characterized by the inability to close the eyes on command with the ability to close the eyes during other times (sleep) and with the preservation of spontaneous blinking and reflex shutting of the eyes.[1,2] The term "apraxia" applies when the execution of a motor action fails despite the patient's understanding of the command and obtaining a physical desire to perform the action and with the preservation of the motor and sensory systems, coordination, and comprehension.[3,4] The prevalence of AEC is extremely less common than a similar condition, namely, apraxia of eyelid opening (AEO), which is also seen in progressive supranuclear palsy (PSP)[5] and Parkinson's disease (PD).[6] The exact pathophysiological mechanisms underlying this condition remain unknown.[7-14] No long-term follow-ups have been carried out and treatment is still variable. Apraxia is a subtle sign that may be missed by non-neurologist or thought of as a normal aging process. Recognition of apraxia by the primary care physician is important during a routine clinical exam or when caregivers raise this concern. Referring these patients to a neurologist can help in diagnosing underlying neurological diseases such as PD or other neurodegenerative diseases. Therefore, delivering appropriate treatment and counseling is needed.

Case History

Following the consensus-based clinical case reporting (CARE) guideline, we present a case of an 81-year-old Saudi male patient with multiple comorbidities including a history of hypertension, old ischemic stroke, and advanced PD for the last 6 years

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on carbidopa/levodopa multiple times a day in addition to pramipexole. He presented to the emergency department with symptoms of COVID-19 pneumonia and required intubation and intensive care unit (ICU) admission due to respiratory failure. On the next day following extubation, during the routine morning round, the patient's son reported that his father was not blinking since he first noticed his eyes early in the morning. He closed his eyes during sleep and maintained their closure with manual closure. The patient was assessed by the neurology team. The patient was conscious, alert, and oriented, with masked facial expression with normal vision acuity and visual field tests and no diplopia or restriction on eye movement, and no obvious saccadic abnormalities. The patient's eyes were open with preserved blinking reflex, and no facial weakness but he was unable to close his eyes after verbal request despite being able to follow other verbal instructions. He had bilateral resting tremors and bradykinesia but no rigidity. Primitive reflexes including glabellar tap and palmomental reflex were positive. The patient had brain magnetic resonance imaging (MRI), which revealed multiple bilateral deep white matter ischemic changes with no interval changes compared to the previous MRI. There were generalized atrophic changes more prominent in the frontal area [Figures 1 and 2]. The patient was diagnosed with bilateral AEC, which is related to the neurodegenerative disease he had, the diagnosis of PD was reviewed as this is more likely a progressive supranuclear palsy due to the presence of multiple atypical features including early loss of mobility, recurrent fall, and poor response to treatment in addition to the new finding of APC, which is more common with PSP. This patient continues to receive medical care, his PD treatment was adjusted gradually, refreshing tears were also prescribed, and he is currently under follow-up with a multidisciplinary team.

Discussion

AEC is a neurological abnormal condition that is not yet fully understood, with several theories presented for its pathophysiological mechanism. It is postulated that a lesion in the corona radiata, a brain area responsible for the AEC, is secondary to the disconnection between the primary and secondary motor cortices.[15] AEC is of importance as it is associated with some neurodegenerative disorders. Being aware of the presence of this sign will help the primary care physician to better understand the patient's complaint and make a differential diagnosis of possible neurological disease.[16] Table 1 presents a comparison between the clinical findings of the study and those reported in the literature.[2,10-12,17,18] In this study, however, we emphasized the ability to notice such uncommon features such as AECs by the care-providing team, family members, and those who take care of patients, which can be valuable information to reach a correct diagnosis and facilitate optimal treatment and early referral. PSP and PD diagnosis can be challenging especially in the early course of the disease. Although PSP patients commonly have restricted vertical eye movement, some patients never develop this sign.[19,20] Therefore, recognition of less common features such as AEC is important. Patients with PD may benefit from multiple therapeutic interventions; however, PSP requires more supportive care and symptomatic treatment only. Reaching the correct diagnosis in a patient with atypical parkinsonism such as PSP can save the patient from the side effect of unnecessary treatment. Raising the level of knowledge and awareness about this uncommon sign among non-neurologists such as family physicians can improve patient care and satisfaction by getting an early referral to a neurologist and accurate diagnosis. Management of this condition has been broadly addressing underlying etiology as no definitive guidelines have been established yet.

This case presented a PSP case, which was initially labelled as PD. The caregiver's recognition of AEC helped in making the correct diagnosis. The lack of common features of PD complicated by AEC makes it difficult to notice and address such a condition. Families with patients with neurological disease are an essential group when coming to assess patients with chronic neurodegenerative disease. Listing to the caregiver and taking time to assess subtle signs is important as abnormalities such as AEC can be missed in busy clinical practice or hospitalized patients with other comorbidities.

Key points

- AEC is a rare and poorly understood disorder.
- AEC is characterized by loss of voluntary control to close the eye with preserved reflex blinking.

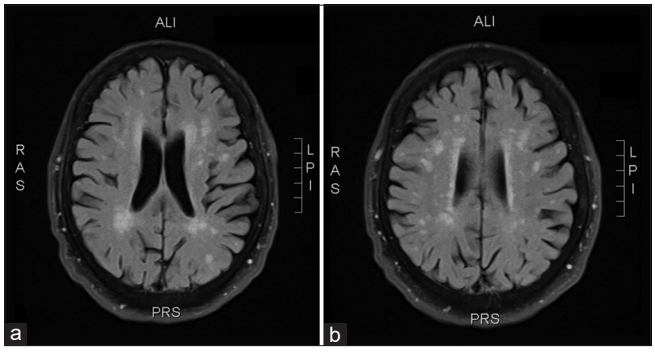

Figure 1: (a and b) Axial T2-weighted MRI flair images with deep white matter ischemic changes with multiple bilateral frontoparietal tiny ischemic foci

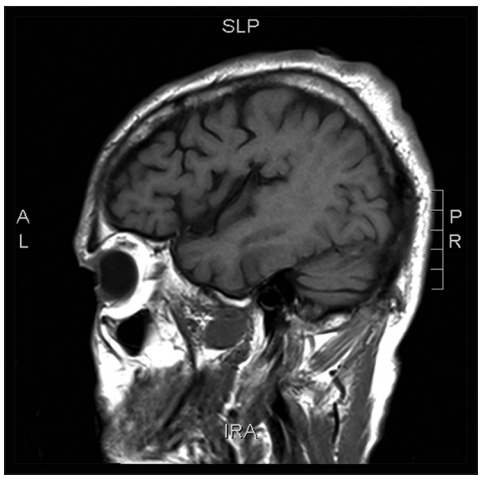

Figure 2: Sagittal T1-weighted MRI images with age-related brain involutional changes

Table 1: A summary of literature findings for cases with AEC

Birth details and dysmorphic features	Our case	Nicoletti *et al.*[17]	Almallouhi and Dale[21]	Thon *et al.*[18]	Favaretto *et al.*[12]	Fukushima *et al.*[10]	Pérez *et al.*[2]	Bonelli and Niederwieser[11]
Year of publication	2022	2021	2018	2017	2011	2007	2007	2002
Sex	Male	Female	Male	Male	Male	Female	Male	Female
Age	81 years	89 years	90 years	56 years	71 years	57 years	67 years	59 years
Comorbidity	Parkinson disease	Stroke	Bilateral subcortical frontal infarcts	Cerebral infarction	Progressive supranuclear palsy	Amyotrophic lateral sclerosis	Stroke	Huntington disease
Site	Bilateral	Bilateral	Bilateral	Bilateral	Bilateral	Bilateral	Unilateral	Bilateral
Hypertension	+	NM	NM	NM	+	NM	NM	NM
Brain								
Facial droop	-	NM	+	NM	NM	NM	NM	NM
Hemiparesis	-	NM	+	+ (Lt	NM	NM	NM	NM+NM
Frequent falls	+	NM	+ (Lt side)	side)	+	NM	NM	+
Gait instability	-	NM	-	NM	+	NM	NM	
Sleeping with closed eyes	+	+	+	+	+	NM	+	
Eyes								
Abnormal Ocular movements	-	-	-	NM	+	NM	NM	NM
Mouth								
Dysphagia	+	+	NM	NM	+	NM	NM	NM
Dysarthria	+	+ (mild)	NM	NM	+	NM	NM	NM
Cardiac abnormalities	-	NM	NM	NM	+	NM	NM	NM
Failure to thrive	-	NM	+	NM	NM	NM	NM	NM
Infections	+ (COVID-19)	NM	+	NM	-	NM	NM	NM

- AEC may be an under-recognized sign in hospitalized patients.
- AEC can be secondary to neurodegenerative disease or stroke.
- Recognition of an uncommon sign such as apraxia of eyelid closure can help in making the correct diagnosis.

List of Abbreviations

- AEC = Apraxia of eyelid closure
- AEO = Apraxia of eyelid opening
- ICU = Intensive care unit
- PD = Parkinson's disease
- PSP Progressive supranuclear palsy

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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