CASE REPORT

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Ictal hypersalivation as a prominent symptom in a girl with insulo-opercular epilepsy

Ke Sun[1] | Dongju Yang[2] | Shuli Liang[1] | Liu Yuan[1] | Guojun Zhang[1]

[1]Functional Neurosurgery Department, Beijing Children's Hospital, Capital Medical University, National Center for Children's Health, Beijing, China
[2]Department of Neurology, Xuanwu Hospital, Capital Medical University, Beijing, China

Correspondence
Guojun Zhang, Functional Neurosurgery Department, Beijing Children's Hospital, Capital Medical University, National Center for Children's Health, Beijing 100045, China.
Email: zgjxwyy@126.com

Received: 5 March 2023
Accepted: 30 May 2023

ABSTRACT

Introduction: Hypersalivation has been associated with Rolandic epilepsy and other childhood epilepsy syndromes. However, pure salivatory seizures are a rare type of focal seizure in which ictal hypersalivation is the dominant feature throughout the seizures.

Case presentation: We present a case of pure salivatory seizures originating from the right post-central operculum cortex, confirmed by the favorable surgical outcome. We attempt to analyze the symptom from behavioral and neural network perspectives and propose a possible mechanism to generate ictal hypersalivation and pure salivatory seizures.

Conclusion: Based on previous reports in the literature and our case, we emphasize the importance of the operculum in patients with ictal hypersalivation, particularly in patients with pure salivatory seizures.

KEYWORDS

Ictal hypersalivation, Insulo-opercular epilepsy, Pure salivatory seizures, Semiology

INTRODUCTION

Hypersalivation, a well-known ictal semiology, has been associated with Rolandic epilepsy and other childhood epilepsy syndromes. The brain lesion of the Rolandic area, fronto-orbital cortex and cingulate gyrus, insula, operculum, and mesial temporal structures, or this area's dysfunction, is associated with ictal hypersalivation.[1] Generally, a variety of ictal automatisms, such as ictal vomiting, hypersalivation, spitting automatism, and oral-alimentary automatisms, are typically in combination with other epileptic manifestations. Seizures comprising exclusively autonomic symptoms, however, are rarely reported. Pure salivatory seizures are a rare type of focal seizure in which ictal hypersalivation is the dominant feature throughout the seizures.[2] Penfield and Jasper reported a case of pure salivatory seizures in 1954.[3] Subsequently, further cases of salivatory seizures in combination with varying degrees of oromotor dysfunction and other select clinical symptoms were reported.[1,4–6] Herein, we share a case of a patient with pure salivary seizures who presented with hypersalivation as a prominent manifestation, followed by the clonus jerks of the left face and forearm. After the resection of the right centro-operculum region, the patient was seizure-free. We discuss the possible mechanisms by which the brain cortex induces hypersalivation.

DOI: 10.1002/ped4.12390

CASE REPORT

A 14 -year old, right-handed girl who was referred for a presurgical evaluation, her focal seizures presented as an aura difficult to describe, followed by hypersalivation with impaired consciousness after tens of seconds, eyes deviation to the left, then the left forearm jerk, and hemifacial twitching. No facial flushing, no difficulty breathing, and no abnormal sensations. Seizures often occur when waking from sleep. She had a negative history of perinatal complications, febrile convulsions, and central nervous system infections or trauma. Her family history and examination results were unremarkable. Despite taking multiple medications (levetiracetam, lamotrigine, and lacosamide), the seizures remained medication-refractory.

Local signal enhancement in the right operculum cortex was found on the fluid-attenuated inversion recovery sequence of the brain magnetic resonance imaging (MRI). More precisely, the abnormal signal enhancement was mainly located in the post-central operculum and extended to the anterior long gyrus of the insula. Fluorodeoxyglucose positron emission tomography showed hypermetabolism in the right operculum cortex (Figure 1). Interictal video electroencephalography (EEG) revealed spikes in the left frontotemporal region. Hypersalivation was further depicted in six seizures recorded by video-EEG: awake, uncomfortable aura, sitting up, and ictal video-EEG showed hypersalivation with simultaneous fast activity in the right temporal regions. The seizure lasted 50–120 s, and thereafter, awareness returned quickly. The quantity of saliva collected was between 30 and 50 ml for each seizure.

The patient was discussed at a routine multidisciplinary presurgical evaluation conference in the Comprehensive Epilepsy Center of Beijing Children's Hospital. Through a comprehensive analysis of various data, including semiology, scalp EEG, and neuroimaging diagnosis (MRI and positron emission tomography), the patient was undergoing right operculum lesion resection. Intraoperative EEG monitoring showed focal sharp waves. Histopathology demonstrated focal cortical dysplasia type IIa. The patient had been seizure free for 18 months at the last follow-up.

DISCUSSION

We present a patient with pure salivatory seizures and repetitive motor automatisms due to non-dominant right operculum-insular lobe epilepsy. Intraoperative cortical EEG and the favorable post-surgical outcome are further proof of seizure onset zone localization. Isolated salivatory epileptic phenomena are rare. Penfield and Jasper were the first to recognize pure salivatory seizures in 1954. They described a patient who had frequent drooling paroxysms that were accompanied by continuous focal seizures. After the removal of a traumatic cyst in the postcentral gyrus, the patient was seizure-free.[3] It was not until 1963 that two other cases of salivatory seizures were documented.[4]

Saliva is produced by six major salivary glands and several hundred minor salivary glands. The major salivary glands secrete 1.5 L of saliva per day. The neurological stimulus for salivary secretion is parasympathetic, and the contraction of the smooth muscle of the salivary ducts is stimulated by the sympathetic nervous system. Stimulation of beta-adrenergic receptors is responsible for the production of mucous secretions.[6,7]

Hypersalivation is a problem due to increased oral secretion. Increased sialorrhea may be ascribed to the following reasons: excessive production of saliva, decreased clearance of saliva, an inability to keep the mouth closed, or difficulty in swallowing the excess saliva. In neurological conditions, this excessive saliva results from a weakness or poor coordination of the bulbar or facial musculature. Sialorrhea has been found with various neurological conditions, including stroke, neuromuscular diseases such as amyotrophic lateral sclerosis/motor neuron disease, and neurodegenerative diseases such as Parkinson's disease, multiple system atrophy, progressive supranuclear palsy, and dementia with Lewy bodies.[7]

Ictal hypersalivation is a dramatic increase in the production of saliva leading to uncontrollable drooling during seizures. It is semiology that has been reported to be associated with benign Rolandic epilepsy and temporal lobe epilepsy.[8] Reports of ictal hypersalivation are sporadic, and the lateralization value of this complex oral automatism remains uncertain. Shah et al.[9] reported ten patients with hypersalivation; seven patients had nondominant temporal lobe epilepsy, and three patients had dominant temporal lobe epilepsy. Morita et al.[1] described one case with abnormal massive salivation whose left hippocampal atrophy, after left temporal lobectomy was performed, the patient was completely seizure-free with no episode of massive salivation. Satow et al. [5] reported a patient with ictal hypersalivation whose brain MRI showed an atrophic lesion around the left peri-Sylvian area. Nascimento et al.[2] documented a patient with pure salivatory seizures secondary to a subtle malformation limited to a single gyrus at the lower part of the right parietal cortex. By performing stereo-EEG (SEEG), although patients were not having pure salivatory seizures, Isnard et al.[10] reported insulo-opercular symptoms including hypersalivation, clonic facial contractions, and strong laryngeal constriction, which also helped identify the insular and operculum as the origin of seizures. Wang et al.[11] also found ictal discharges in the insula and operculum during ictal hypersalivation. Therefore, based on the literature, the operculum may be the region most implicated in ictal hypersalivation. Our case showed that

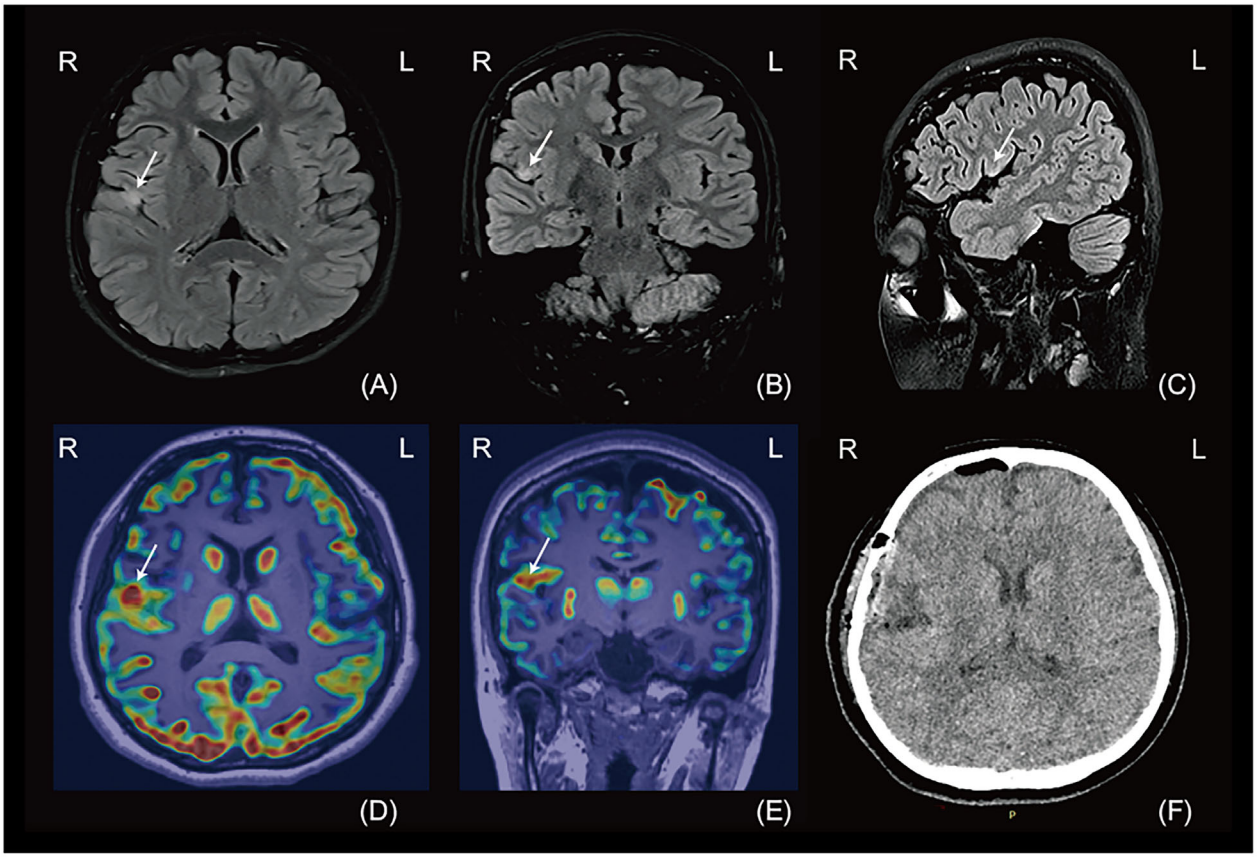

FIGURE 1 The pre-surgical evaluation images and the post-operative image. (A) Axial T2 FLAIR showing a high-intensity signal in the posterior of the right central operculum part (white arrow). (B) Coronary T2 FLAIR showing a high-intensity signal in the posterior of the right central operculum part (white arrow). (C) Sagittal T2 FLAIR showing a high-intensity signal in the posterior of the right central operculum part (white arrow). (D) PET imaging showing focal hypermetabolism in the same area on an axial view (white arrow). (E) PET imaging showing focal hypermetabolism in the same area on a coronary view (white arrow). (F) Post-surgical computed tomography indicating resection of the right centro-operculum region. FLAIR, fluid-attenuated inversion recovery; PET, positron emission tomography.

the nondominant operculum can also lead to pure salivatory seizures, consistent with the prior results. This usually reflects the spread of the ictal discharge to the suprasylvian opercular region and is sometimes accompanied by dysarthria in a cluster analysis of insular epilepsy, the posterior insular can induce the symptom.[12]

Since the widespread use of SEEG, the functions of the operculum have been gradually explored. Various and complicated functions have been reported in the insula and opercula, such as somatosensory, visceral sensory, speech, motor, cardiovascular, auditory, vestibular, and visual, depending on the site of stimulation.[13] Mazzola et al.[14] discovered that electrical stimulations of the area around the central sulcus of the insula could evoke hypersalivation. In addition, a symptom analogous to ictal hypersalivation, ictal spitting, was described from right temporal seizures, but the symptomatogenic area is probably outside the temporal lobe.[15] Therefore, we believe that epileptic hypersalivation is probably a function of a network or networks, perhaps including the central autonomic network, rather than the function of a single anatomic structure or region. The central autonomic network consists of a group of interconnected areas of the telencephalon, diencephalon, and brain stem that control preganglionic sympathetic and parasympathetic visceromotor outputs.[16] Although it can be difficult to determine whether hypersalivation is caused by temporal lobe seizures or insular lobe seizures, some clinical signs can help us differentiate the origin of episodic salivation. In temporal lobe seizures, hypersalivation is caused by oropharyngeal automatisms that manifest as excessive chewing, leading to foaming at the mouth, whereas in seizures involving the insular lobe, hypersalivation is often a result of perioral and pharyngeal dystonia, leading to a more dilute and watery production of saliva.

Based on previous reports in the literature and our patient's clear lesion on MRI and favorable surgical outcome, we highlight the importance of the operculum in patients with

paroxysmal hypersalivation, especially in patients with pure salivatory seizures.

CONSENT FOR PUBLICATION

Consent was obtained from the patient's guardian.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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How to cite this article: Sun K, Yang D, Liang S, Yuan L, Zhang G. Ictal hypersalivation as a prominent symptom in a girl with insulo-opercular epilepsy. *Pediatr Investig*. 2023;7:297–300. https://doi.org/10.1002/ped4.12390