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Case study

Ictal pouting ('Chapeau de gendarme') in three pediatric cases with cortical dysplasia

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ABSTRACT

Ictal pouting (Chapeau de gendarme sign) can be described as an inverted smile. It consists of a turned down mouth with the contraction of the chin, wrinkling of the lips and symmetrical lowering of the labial commissures. This shape resembles the gendarme's hat during Napoléon I's time. Chapeau de gendarme sign is frequently seen in frontal and temporal lobe seizures. Focal cortical dysplasias are intrinsically epileptogenic foci and are frequently seen in patients with ictal pouting in seizure semiology. In this report, we analyzed clinical data, video EEG recordings and brain imagings of three children presenting with ictal pouting semiology in whom patients' magnetic resonans images (MRIs) or positron emission tomographies (PETs) were positive or doubtful for FCD in all. In case 1 and 2 the epileptogenic zones were temporal or temporoinsular. In these patients, with involvement of temporal lobe, dystonia and automatisms were seen in the seizure semiology after chapeau de gendarme sign. In case 3 with frontal lobe origin, hypermotor movements were seen after ictal pouting. In the patients 1 and 2, the cortical dysplasias were in temporal lobe. In patient 3, PET demonstrated hypometabolism on left inferior frontal gyrus but we couldn't verify this finding with MRIs. Ictal pouting (Chapeau de gendarme sign) is a distinct seizure semiology that can often be overlooked and coexist with focal cortical thickening. We suggest that focal cortical dysplasias should be searched in patients with ictal pouting particularly in those with refractory focal seizures.

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1. Introduction

Ictal pouting, 'Chapeau de gendarme' sign defined by turned-down mouth with contraction of the lip and chin symmetrically, resembling the gendarme's hat during Napoléon I's time, is an extremely rare feature in epileptic seizures in the pediatric age group. Only four pediatric cases have been reported in the literature.[1–3] In these reports, frontal, temporal lobe and anterior cingulate cortex associated seizures were defined. Focal cortical dysplasias (FCDs) are intrinsically epileptogenic foci[4] which may be seen in patients with ictal pouting. A study of 11 patients with ictal pouting demonstrated five patients with typical features of FCD located in the mesial part of the frontal lobe and doubtful for FCD in the other five patients.[1]

We report three cases who presented with ictal pouting during prolonged video-EEG monitoring for the purpose of pre-surgical evaluation of medically refractory epilepsy. We analyzed clinical data, video EEG recordings and brain imagings of these three children in whom MRI's or positron emission tomographies (PETs) were positive or doubtful for FCD in all. There have been reports of ictal pouting associated with focal cortical dsyplasia[1,5,6] though documentation of these seizures is seldom available.

2. Case 1

A 15-year-old girl with a mild intellectual disability was admitted to our video-EEG monitoring (VEM) unit with seizures starting at the age of two. She received valproate, levetiracetam,

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carbamazepine and lamotrigine previously without seizure control. In interictal EEG, sharp and wave discharges and double phase reversals in left frontotemporal regions were seen (Fig. 1a). The seizures were characterized by loss of consciousness, ictal pouting ("Chapeau de gendarme" sign) (Fig. 2), nonversive head turning to the left, dystonia of right hand and then bilateral hand and mouth automatisms. During the ictal pouting her eyes were opened and frightened expression was observed. The epileptogenic zone of ictal activity was left frontal lobe. Ictal activitiy started with diffuse desynchronization and teta slowing on left frontal regions and than low-voltage fast activity on left frontotemporal lobe was seen (Fig. 1b). The brain MRI revealed left hippocampal sclerosis and asymmetric hyperintensity of left temporal lobe with reduced delineation of the gray–white matter interfaces representing temporal lobe cortical dysplasia (Fig. 3). PET demonstrated hypometabolism in the mesial and lateral segment of the left temporal lobe (Fig. 4). The patient wasn't found to be appropriate for surgery because of the wide extension of the cortical dysplasia. Consequently antiepileptic drug regimen was arranged.

3. Case 2

A 16-year-old girl presented with seizures for the last 2 years. Seizures started with an aura with a feeling of discomfort with nausea. She had loss of consciousness, eye blinking and grimacing. Despite the use of carbamazepine and topiramate treatments, the

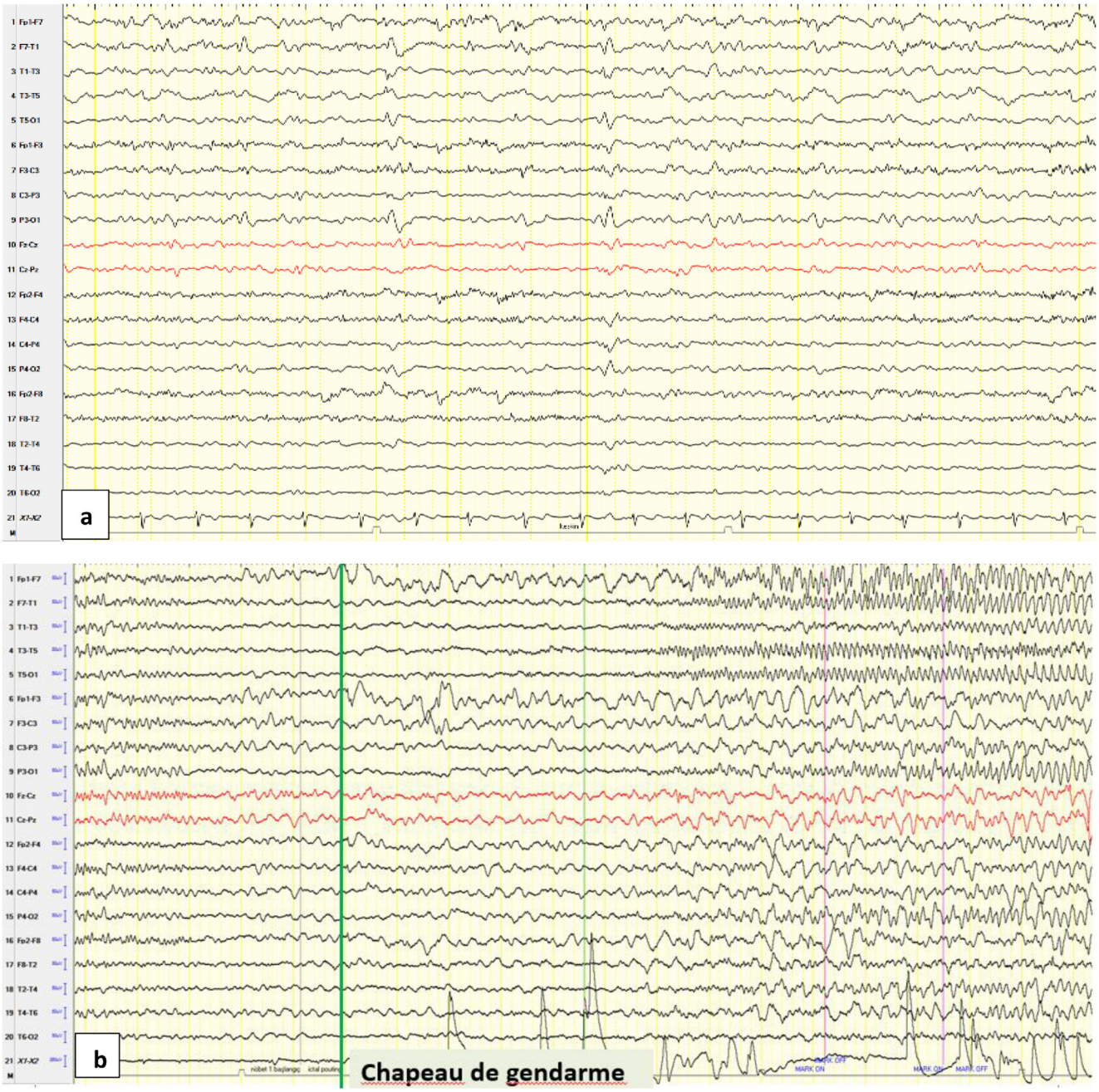


Fig. 1. a: Interictal EEG with sharp and wave discharges, double phase reversals in left frontotemporal region. b: Ictal onset with diffuse desynchronization and teta slowing on left frontal regions and than low-voltage fast activity on left frontotemporal lobe.



Fig. 2. Patients with ictal pouting (IP). In patients 1 and 2, eyes are open with an expression of fear (patient 1) and disgust (patient 2), contrary patient 3's eyes were closed with an expression of discontent.

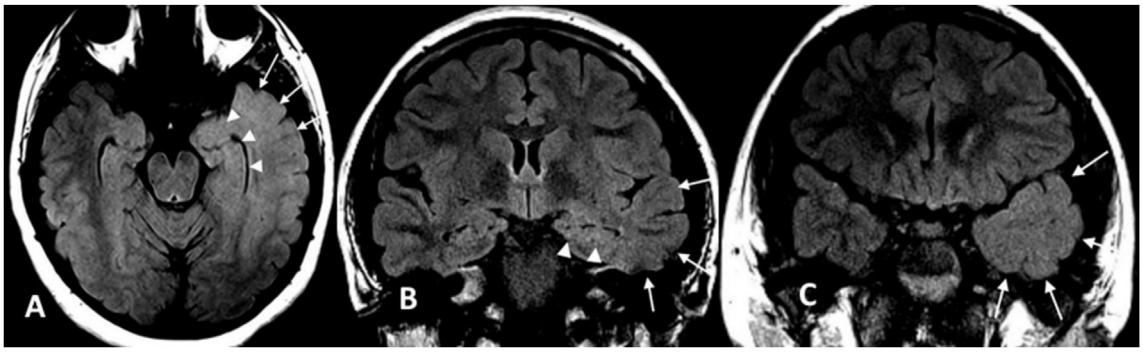


Fig. 3. Axial **(A)** and coronal **(B and C)** plain FLAIR images show left hippocampal sclerosis (arrowheads) and asymmetric hyperintensity of left temporal lobe with reduced delineation of the gray–white matter interfaces representing temporal lobe cortical dysplasia **(arrows.)**.

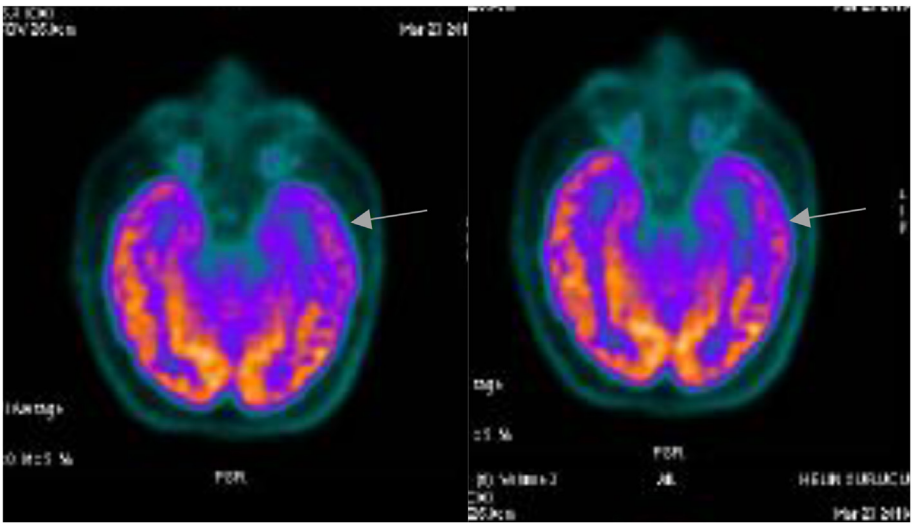


Fig. 4. PET images show hypometabolism in the mesial and lateral segments of the left temporal lobe.

patient had daily seizures, she was hospitalized in the VEM unit. There was no epileptiform discharges in interictal EEG. The seizures were characterized by loss of consciousness, ictal pouting ("Chapeau de gendarme" sign) (Fig. 2), opened eyes with eye blinking and feeling of disgust, dystonia of right hand and bilateral hand and mouth automatisms. The epileptogenic zone was localized to the left frontal lobe (Fig. 5). Brain MRI revealed cortical dysplasia in the left mesial temporal lobe extending to the left insular region (Fig. 6a and b). PET showed hypometabolism in the mesial segments of the left temporal lobe (Fig. 6c). The patient was scheduled for surgery, but the family did not accept. The seizures were partially controlled with lacosamide treatment.

4. Case 3

A 9-year-old male patient was admitted to VEM unit for seizures

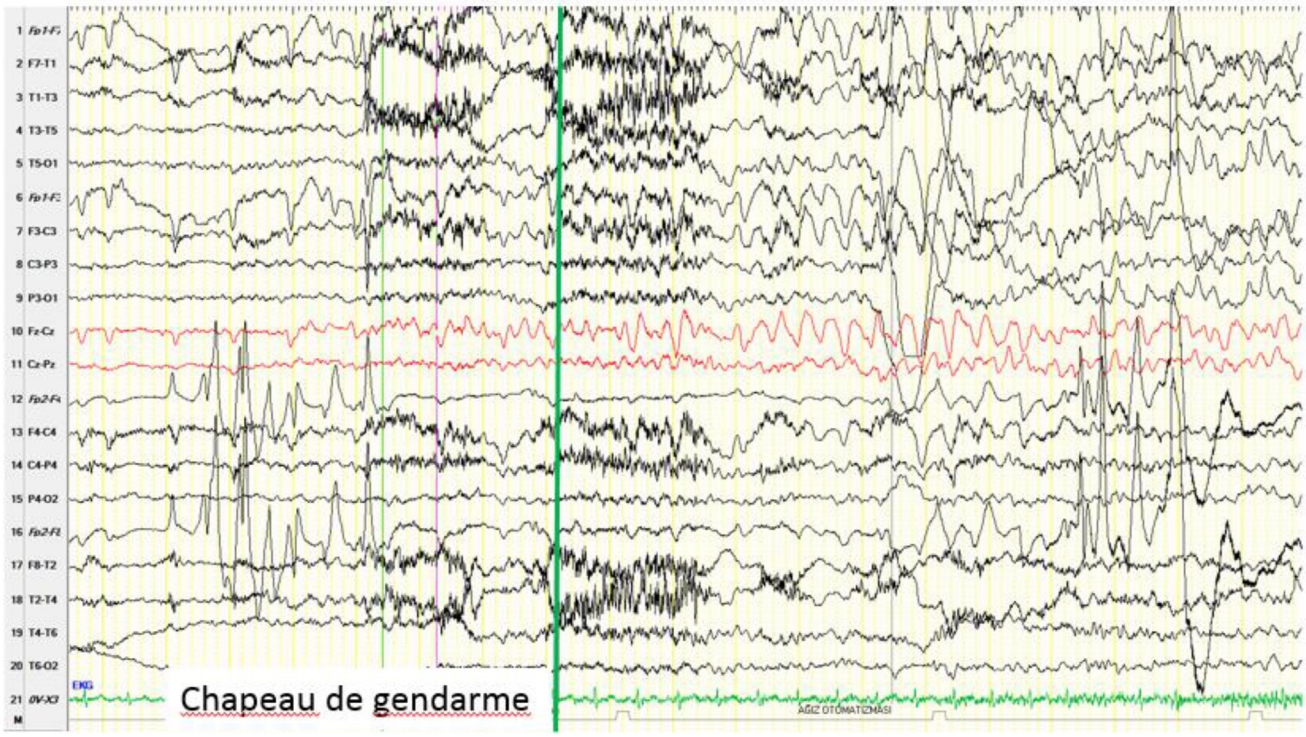


Fig. 5. Ictal onset with teta activity on left frontal and central regions.

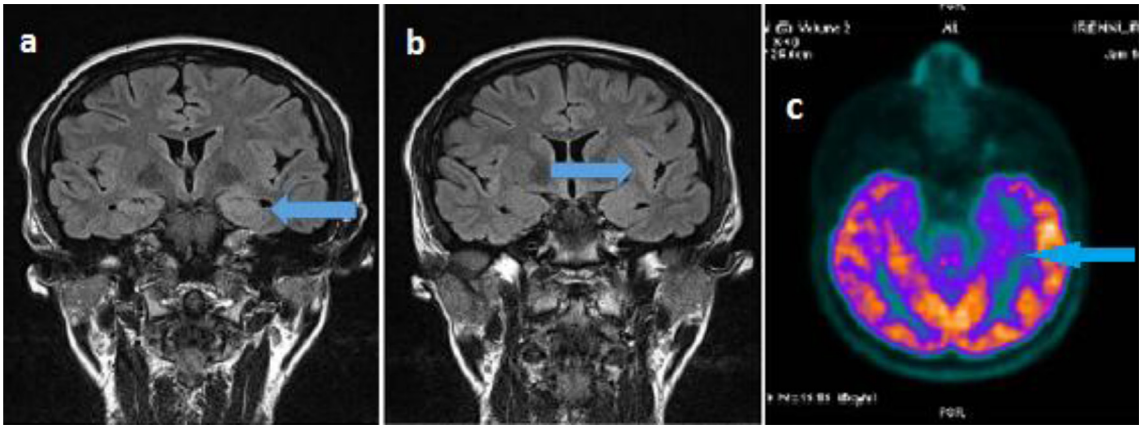


Fig. 6. a,b: Brain MRI revealed cortical dysplasia in the left mesial temporal lobe extending to the left insular region. c: Hypometabolism in the mesial segments of the left temporal lobe in PET images.

beginning after 6 years of age. Valproate, levetiracetam, phenytoin and carbamazepine have been used previously, but seizures were drug resistant. The interictal pattern seen on VEM was sharp and slow waves and generalized periodic discharges with a marked frontal predominance (Fig. 7a). The seizures were characterized by forced expirium, unconsciousness, turned-down mouth (ictal pouting) ("Chapeau de gendarme" sign) (Fig. 2), closed eyes, a discontent expression followed by hypermotor seizure lasting for 15–20 s. The electrographic pattern of the ictal activity was evident in the left hemisphere but were present in both hemispheres at bilateral frontal and parasagittal regions (Fig. 7b). MRI study was negative for focal cortical dysplasia but PET demonstrated hypometabolism on the left inferior frontal gyrus (Fig. 8). The patient was right-handed and functional MRI showed Broca and Wernicke activities on left hemisphere. For these reasons the patient was found not eligible for epilepsy surgery and medical treatment was rearranged. Clobazam treatment resulted in a significant reduction in the seizures.

5. Discussion

Ictal pouting (Chapeau de gendarme sign) can be described as an inverted smile. It consists of a turned down mouth with the contraction of the chin, wrinkling of the lips and symmetrical lowering of the labial commissures.[6]

In previous studiesictal pouting was indicated to be frontal lobe origin especially anterior cingulate cortex (ACC).[7] Souirti et al. analysed 11 patients with chapeau de gendarme sign and the results provide evidence of the involvement of mesial frontal structures and the crucial role of the anterior cingulate cortex in ictal



Fig. 7. a: Sharp and slow waves and generalized periodic discharges with a marked frontal predominance in interictal EEG. b: ictal activity in both hemispheres at bilateral frontal and parasagittal regions, evident in the left hemisphere.

pouting generation.[1] Unlike these, in the case reported by Leitinger et al. intracranial monitoring was performed but no frontomesial focus was detected.[8] In a 16-year-old boy with ictal pouting, MRI showed a brain tumour originating from the right temporal lobe.[2] In the case reported by Tedrus et al., the seizure semiology was ictal pouting in a patient with centrotemporal epileptogenic zone.[3] In our case 1 and 2 the epileptogenic zones were temporal or temporoinsular. In case 3 with frontal lobe origin, hypermotor movements were seen after ictal pouting. In case 1 and 2 with involvement of temporal lobe, dystonia and automatisms were seen in the seizure semiology after chapeau de gendarme sign. Ictal pouting was the early semiology in all our three patients. Although two of our cases have dysplasias in the temporal lobe, ictal pouting was seen in the beginning of seizure with frontal origin in ictal analyzes. This finding supports the frontal origin of ictal pouting in previous studies. Therefore, it may be thought that the patients have frontal lobe involvement, even though their lesions are temporal. A case similar to our patients was reported by Grunwald et al..[9] During the presurgical evaluation of their patient, the results from non-invasive electrophysiological and functional imaging studies all pointed to the seizures originating within the right temporal lobe. The use of invasive electrodes evaluted the localization of the primary epileptogenic area of anterior cingulate cortex. On the other hand, the anterior cingulate gyrus maintains massive interconnections, both with the limbic system and with the frontal, temporal, and parietal neocortex.[9] Thus, the ictal

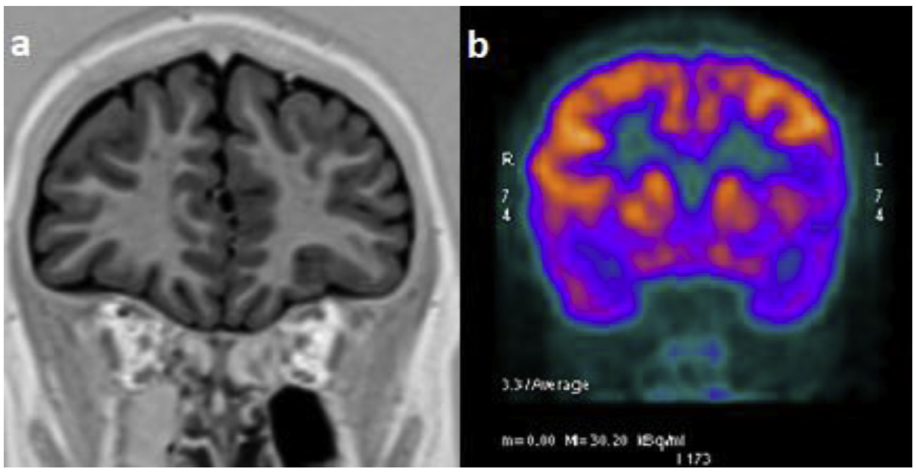


Fig. 8. a: Normal brain MRI b: Hypometabolism at left inferior frontal gyrus in PET images.

activity may start from temporal lobe and rapidly spread to ACC, so that the ictal zone looks like the frontal lobe. However we could not demonstrate whether the chapeau de gendarme sign's origin is ACC or any part of frontal or temporal lobe, as we do not have invasive EEG recordings.

FCDs are intrinsically epileptogenic areas, and stereo-EEG studies have demonstrated both ictal and interictal discharges originating from dysplastic cortex.[4] Previous studies demonstrated an association between ictal pouting and FCDs in mesial part of frontal lobe, frontal operculum extending into the insula, anterior cingulate cortex and temporal lob.[2,5,6] Souirti et al. reported 11 patients with ictal pouting. In five of them, MRI showed typical features of FCD and in other five the images were doubtful for cortical dysplasia.[1] In our patients 1 and 2, the cortical dysplasias were in temporal lobe. In patient 3, PET demonstrated hypometabolism on left inferior frontal gyrus but we couldn't verify this finding with MRIs. This may be due to the low tesla values (1.5 T) of MRI.

Previous studies reported the patients with FCDs and ictal pouting who became seizure-free after surgery.[1,5,6] On the other hand, none of our patients were operated. So we can not mention if patients would be seizure free when surgical resection was performed. Actually Sourti et al. reported globally 11 patients out of 36 seizure free frontal lobe epilepsy patients with ictal pouting and all had FCD.[1] We do not know if the 25 seizure free patients with frontal lobe epilepsy without ictal pouting had an FCD or not. This may be due to intense interconnections between anterior cingulate gyrus and limbic system in the regions of cortical dysplasia.

In the study with the largest patient group with ictal pouting, they suggested that the chapeau de gendarme sign could be associated with the involvement of mesial frontal areas, especially the anterior cingulate cortex (ACC).[1] Four of their 11 patients had mainly closed eyes during the initial part of the seizure, whereas the other patients had their eyes opened. The epileptogenic zone assessed by SEEG involved the rostroventral part of the anterior cingulate cortex (affective division) in the patients who had their eyes wide open, whereas when the patients had their eyes closed, the dorsal part (cognitive division) was involved. The clinical expression is characterized by an intense emotional pattern like fear or aggressive behavior in the patients with the ictal onset of affective divison. In the patients with the ictal onset of cognitive divison, the affective component may be less intense and more 'cognitive' with the facial expression of disagreement, discontent, disappointment or doubt and often closed eyes. In our cases, third patient's eyes, with epileptogenic zone of frontal lobe, were closed during ictal pouting. In the patients with temporal lobe involvement, both in case 1 and 2, the eyes were open. In case 1, during the ictal pouting her eyes were opened and a frightening expression was observed. Previous researches reported that, fear with or without a matching facial expression may be an early manifestation of mesial frontal lobe seizures.[10,11] Our patient has a widespread cortical dysplasia in temporal lobe, however there may be frontal lobe extension that we cannot demonstrate with MRI. In case 2, she had a feeling of disgust with wide opened eyes. Previous studies demonstrated the fact that the anterior insula is necessary for the capability of feeling disgust and recognize the same emotion in others[12–15] So in case 2, the cortical dysplasia extending to the insula can be the cause of the expression of disgust with ictal pouting. In case 3, he had a discontent expression with the chapeau de gendarme sign. This is similar with the patients whose eyes are closed and have a neutral facial expression in the study of Sourti et al..[1]

6. Conclusion

Ictal pouting (Chapeau de gendarme sign) is a very uncommon type of seizure semiology originating predominantly from frontal and temporal lobe. As we have described here, focal cortical dysplasias in the frontal and temporal region is probably a causal association in these patients. We suggest that focal cortical dysplasias should be searched in patients with ictal pouting particularly in those with refractory focal seizures.

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