Epilepsia partialis continua of the face in a 4-year-old girl

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Epilepsia partialis continua (EPC) is defined clinically as a syndrome of continuous focal jerking of a body part, usually localized to a distal limb, occurring over hours, days, or even years. It is an extremely rare condition with a prevalence estimated to be less than one per million.[1] In this paper, we report a rare case of EPC confined to the face of a 4-year-old girl.

A 4-year-old girl was recently referred to our clinic due to jerks in the right side of her face especially in the lips and eyes that started 2 months earlier. On neurological examination, she suffered from continuous rhythmic movements in the right side of her face. Muscle tone and force were diminished on the right side of her body. The plantar reflex was upward on the left and neutral on the right. Physical examination was otherwise, normal. She had mild leukocytosis (white blood cell count 14300 [normal: 4000-10000], neutrophils 72%, lymphocytes 17%, monocytes 8%, eosinophils 3%) and anemia (hemoglobin = 10.7 [normal: 12-18 g/dl]). Serum biochemistry, blood culture, urinalysis, urine culture, and liver as well as thyroid function tests were all normal. Electroencephalogram revealed nonspecific findings. An MRI of the brain showed thinning of white matter in the frontal, parietal, and occipital lobes with abnormal signal in T2-weighted images, suggesting periventricular leukomalacia. Single photon emission computed tomography (SPECT) of the brain showed significant hypoperfusion in the left frontal cortex (**Figure 1**). A diagnosis of EPC was made, and she started medical therapy. Various regimens were tried including phenobarbital (60 mg/intravenously [IV]/twice daily [bid]), phenytoin (60mg/IV/bid), lamotrigine (50 mg/orally [PO]/bid), carbamazepine (200 mg/PO/bid), clonazepam (2 mg/PO/bid), gabapentin (100 mg/PO/bid) and general anesthesia. However, no improvement occurred in her condition. Gradually, the seizure became generalized over a few weeks and as the epilepsy was resistant to all available medications at maximal dosage, a decision of corpus callosotomy was finally made. However, her general condition exacerbated rapidly and she died after severe aspiration pneumonia despite aggressive antibiotic therapy.

Epilepsia partialis continua can present in 2 clinical forms: type 1 is usually symptomatic of a focal brain lesion, and the prognosis is the same as that of the associated lesion; type 2 is associated with progressive, idiopathic, neurological deterioration, usually in the form of Rasmussen's encephalitis. Our patient could be best categorized as a case of type 2 EPC. The list of proposed etiologies for EPC is extensive and includes cerebral neoplasms, cortical dysplasias, infections, trauma, drugs, metabolic abnormalities, and idiopathic conditions.[2,3] Idiopathic conditions have sometimes been attributed to genetic and autoimmune causes as well as Rasmussen's chronic encephalitis.[3] Clonic activity in EPC is most common in the upper extremities and typically remains localized to a single muscle group, however, Jacksonian spread of the seizure, sometimes leading to secondary generalized seizure has been reported as well. After a thorough literature review, we could find only approximately 30 reports of EPC cases presenting with jerks confined to the face. Our case therefore introduces a rare presentation of EPC. Electroencephalogram has not been a successful diagnostic modality overall and may be normal or abnormal.[4] In our case, EEG revealed nonspecific findings only. Although the primary imaging modality in the management of epilepsy is MRI, functional neuroimaging often provides complementary information and, in several situations, provides unique information that cannot be obtained with MRI. The functional neuroimaging methods most commonly used in children with drug-resistant epilepsy include positron emission tomography (PET) and SPECT.[4] Both PET and SPECT are emerging as useful research tools in the evaluation of the metabolic effects of EPC, especially when CT and MRI findings are normal. The result of MRI in our case was suggestive of periventricular leukomalacia. However, we proceeded to perfusion studies by SPECT in an attempt to discover the site of abnormal perfusion. The SPECT seems to be helpful for the early diagnosis of EPC, before MRI and EEG reveal any abnormal findings.[4] Regional hyperperfusion accompanied by more diffuse hypoperfusion has been previously described, especially in non-idiopathic

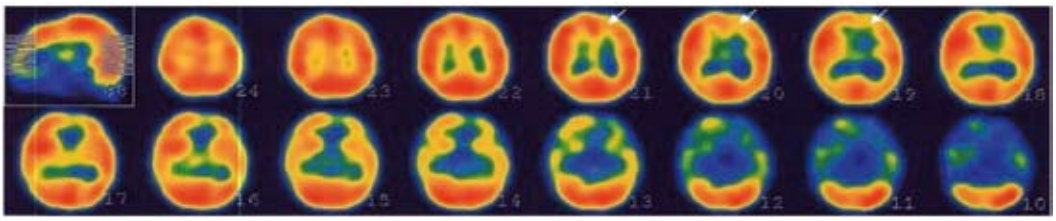

Figure 1 - Single photon emission computed tomography (SPECT) of the brain showing significant hypoperfusion in the left frontal cortex (arrows).

EPC cases.[4] The present report, which is one of very few reports of idiopathic EPC with SPECT showing localized hypoperfusion,[4] emphasizes the potential role of SPECT in localizing the lesion when EEG and MRI fail to reveal specific findings.

The best treatment strategy for idiopathic EPC remains controversial. Antiepileptic drugs must be tried to prevent the spread of EPC, however, they have been generally unsuccessful in altering the course of EPC. New treatments have been developed with variable success. Examples include immunomodulation with corticosteroids and intravenous gamma globulins, transcranial magnetic stimulation and surgery as the final choice. Types and syndromes of epilepsy encountered for palliative surgery are mainly Lennox-Gastaut syndrome, infantile spasms/West syndrome, severe epilepsy multiple independent spike foci, hemi convulsions-hemiplegia-epilepsy syndrome, and other symptomatic partial epilepsies and secondary generalized epilepsy.[5] Corpus callosotomy is among the palliative surgeries for this purpose. Studies in children reveal that callosotomy performed before puberty is not followed by permanent disconnection deficits.[5] For neurosurgeons who are familiar with the procedure of callosotomy, the acute surgical complication will be minimal and acceptable. In fact, patients who have an uncomplicated operative and postoperative course do not experience functionally significant intellectual, emotional, or social impairment.[5] Early identification of medically resistant epilepsy and candidates for epilepsy surgery are important to help in relieving seizures and stabilizing comorbidities in children. Our report demonstrates a case of idiopathic EPC with some rarely reported features, which were refractory to all available antiepileptic drugs, quickly generalized and led to a catastrophic outcome.

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