Bilateral perisylvian polymicrogyria

The advent of MRI technique has enabled the diagnosis of neuronal migration disorders(NMD) and made it possible to make “in vivo” diagnosis. Congenital bilateral perisylvian syndrome(CBPS) is a characterized by pseudobulbar palsy, epilepsy, mental retardation, and migration disorders in the bilateral perisylvian area.

Case series

2013

Schilling et al. report a series of patients with epilepsy and pseudobulbar palsy due to bilateral perisylvian ULG (BP-ULG), show that hippocampal sclerosis (HS) is often associated and highlight the fact that in this entity, unlike in malformative bilateral perisylvian PMG, seizures may be surgically treated.

The motor, cognitive, epileptologic, and imaging features of 12 patients with perisylvian ULG followed at three institutions are described. For patients with refractory seizures, we detail extracranial and intracranial electrographic recordings, surgical strategies, histopathologic analyses of the resected tissue, and outcome of surgical treatment. Descriptive statistics were used for quantitative and categorical variables. Student's t-test was used to compare means, and a $p < 0.05$ was considered significant.

Pseudobulbar palsy and mental retardation were present in all patients with symmetrical BP-ULG. Five had refractory seizures. There was no relationship between the severity of the pseudobulbar palsy or of the mental retardation and the degree of seizure control with medication. The five patients in whom seizures were refractory to medication had significantly earlier age of onset and longer duration of epilepsy ($p < 0.05$). Dual pathology with associated unilateral HS was present in four. One patient with dual pathology had a temporolimbic electroclinical picture and had an anterior temporal lobectomy (ATL) based upon noninvasive evaluation. The other four had ictal semiology suggesting involvement of both temporolimbic and perisylvian cortex. Intracranial electroencephalography (EEG) showed concomitant seizure onset in the anterior temporal region and in the ipsilateral ULG in three of the four with dual pathology and in the ulegyric cortex in the one without HS. Resection guided by a combination of semiology, MRI, and extra and intracranial EEG led to complete seizure control in two and almost complete seizure control (Engel class II) in two other patients. The only surgical failure was an isolated ATL in a patient with dual pathology, and concomitant seizure onset in both lesions according to semiology and intracranial EEG.

The findings suggest that BP-ULG mimics the clinical features of bilateral perisylvian PMG. In patients with refractory seizures, recognition of this entity should lead to consideration of resective surgery despite the bilateral ULG [1)].

2005

In a series of 14 patients, Jansen et al. demonstrated that only a minority had extremely low intelligence, and that some aspects of cognitive function correlated with the extent of the cortical disorganization. Early age at seizure onset correlated positively with Performance IQ scores ($P<0.05$)

and negatively with the extent of the lesion (P<0.01), reflecting that patients with more severe BPP are more likely to have early seizure onset, resulting in greater interference with ongoing cognitive development. Receptive and expressive language skills were found to be equally poor. Frontal lobe function and memory abilities were relatively well preserved, suggesting that the observed cognitive profiles were related, at least in part, to specific areas of cortical dysfunction and not only to global dysfunction [2)]

1998

Miller et al. report three children with pure congenital hemiplegia found to have congenital bilateral perisylvian polymicrogyria (CBPP). None of our patients had the seizures, oromotor dysfunction, or cognitive impairment usually associated with CBPP. CBPP may be more common and heterogeneous than previously thought, is easily recognized by MRI, and should be included in the differential diagnosis of the young child presenting with congenital hemiplegia [3)].

1994

Kim et al. identified four CBPS patients based on neuroimaging and dysarthria patterns among the candidates for epilepsy surgery. All the patients had orofacial diplegia and variable degrees of mental retardation. In the spectrographic analysis of dysarthria, the loss of specific characteristics of formants of vowels and increment of noise in the high frequency formants were observed. Epilepsy was present in all, but only one patient showed intractable seizure requiring surgical intervention. MRI was most helpful in identifying NMD and polymicrogyria in both centroparietal areas in this context. Great alertness is needed to identify this disorder to determine the etiology of epilepsy and dysarthria of uncertain origin [4)].

1)
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