Congenital Bilateral Perisylvian Syndrome: Differential Diagnosis

Konjenital Bilateral Perisilviyan Sendrom

Hanefi YILDIRIM, MD,[a]
Mustafa KOÇ, MD,[a]
Kürşad POYRAZ, MD,[a]
Erkin OĞUR, MD,[a]
Nimet KABAKUŞ, MD[b]

[a]Department of Radiology,
[b]Department of Pediatrics Neurology,
Medical Faculty, Fırat University,
Elazığ

Geliş Tarihi/*Received:* 09.03.2007
Kabul Tarihi/*Accepted:* 22.04.2007

Yazışma Adresi/*Correspondence:*
Mustafa KOÇ, MD
Medical Faculty, Fırat University
Department of Radiology, Elazığ
TÜRKİYE/TURKEY
mkoc@firat.edu.tr

ABSTRACT Congenital bilateral perisylvian syndrome is a structural malformation of the brain which normal pattern of folds on the surface of the brain is replaced with many small abnormal folds. The underlying anomaly is polymicrogyria, a malformation of the cerebral cortex. Polymicrogyria refers to excessive number of prominent and small convolutions composed from shallow and enlarged sulci. This anomaly constitutes lobular appearance of cerebral cortex. We present cranial magnetic resonance imaging (MRI) findings of a 3-year old male patient who was admitted with asymmetric facial paralysis, chewing and swallowing difficulties and intractable seizures. Brain MRI revealed bilateral perisylvian cortical dysplasia including polymicrogyria and local cortical thickening that extends to the level of vertex.

Key Words: Congenital; brain; epilepsy, rolandic; magnetic resonance imaging

ÖZET Konjenital bilateral perisilviyan sendrom beyinin yapısal bir malformasyonudur ve normal girus yapısının yerini çok sayıda küçük anormal giral yapı almıştır. Esas anomali polimikrogri olup sığ ve genişlemiş sulkuslardan, multipl küçük giruslardan oluşan serebral korteksin malformasyonudur ve serebral kortekse lobüler bir görünüm verir. Biz bu yazıda, 3 yaşında, asimetrik fasiyal paralizi, çiğneme ve yutma güçlüğü ve tedaviye cevap vermeyen epilepsi bulguları ile başvuran erkek olgunun, kranial manyetik rezonans görüntüleme (MRG) bulgularını sunduk. MRG görüntüleme, bilateral perisilviyan kortikal displazi, polimikrogri ve verteks düzeyine uzanan lokal kortikal kalınlaşmayı gösterdi.

Anahtar Kelimeler: Konjenital; beyin; rolandik epilepsi; manyetik rezonans görüntüleme

Turkiye Klinikleri J Med Sci 2008, 28:419-421

The congenital bilateral perisylvian syndrome (CBPS) is a congenital neurological syndrome characterized by pseudobulbar palsy, cognitive deficits and bilateral perisylvian abnormalities observed on imaging.[1] Patients have neurologic dysfunction, primarily pseudobulbar paresis. Dysarthria and severe restriction of tongue movements are present. Motor milestones are delayed in most patients and language milestones in all. Mild to moderate intellectual deficits were documented and also pyramidal signs were observed. Advances in neuroimaging techniques, especially in MRI studies, have enabled recognition of CBPS as a clinicoradiologically specific syndrome.[2]

Epilepsy is seen in more than 85% of cases and is frequently intractable.[1]

A 3-year-old boy was admitted due to intractable epilepsy and partial paralysis of muscles on both sides (diplegia) of the face. The patient has had difficulties in speaking (dysarthria), chewing (mastication), and swallowing (dysphagia) since birth. Her motor development was delayed. There was no family history of epilepsy or other neurological disorders. The medical history was unremarkable and laboratory data on admission were within normal limits.

Imaging was obtained at 1.5T MRI. Sedation of the patient was required. On cranial MRI, axial 3D SPGR (spoiled pradient recolled) and coronal morphology images showed infolding of the outer layer of the brain. The perisylvian cortex was bilaterally thicker than normal, suggesting polymicrogyria (Figure 1, 2). Sagittal T1W MR images showed that both sylvian fissures were continuous with the Rolandic sulci (Figure 3). There was no another pathological finding.

Medical treatment was used for his epilepsy and other complaints and the patient was followed-up for 6 months.

DISCUSSION

Polymicrogyria is a malformation of cortical development that is characterised by abnormal arrangement and excessive folding of cerebral surfaces. Acquired lesions such as cerebrovascular diseases or virus encephalitis and congenital lesions such as polymicrogyria may be implied as etiological factors. Most reported families provide evidence suggestive of X-linked transmission and autosomal dominant.

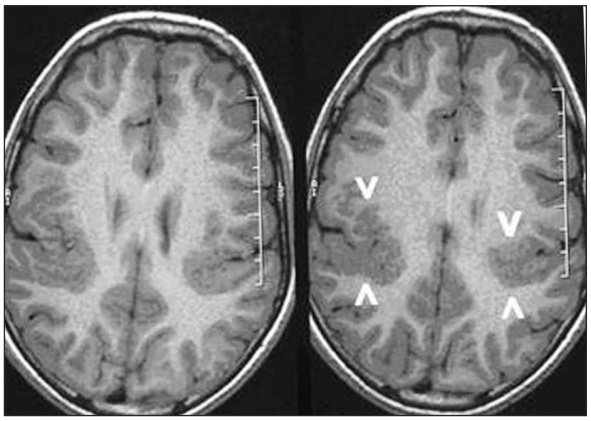

FIGURE 1: Axial 3D SPGR MR image shows bilateral cerebral cortical in folding and thickening extending to vertex (arrow heads).

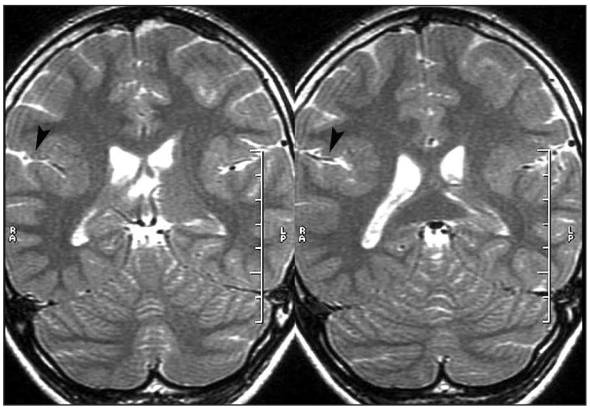

FIGURE 2: Coronal T2W MRI shows sylvian fissures extending posteriorly (arrowheads).

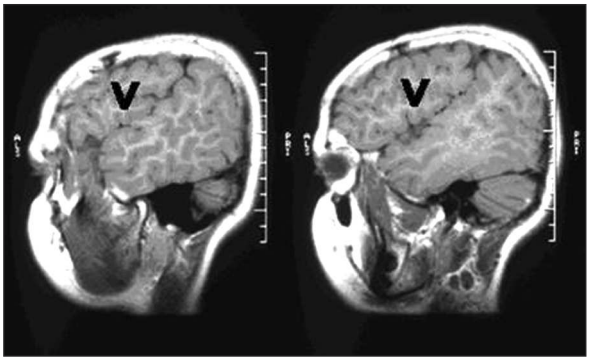

FIGURE 3: Sagittal T1W MR image revealed perisylvian fissures continuous with Rolandic sulci (arrowheads).

Polymicrogyria reveals a combination of three characteristic findings on imaging: abnormal gyral pattern, increased cortical thickness, and irregularity of the cortical white-matter junction. The most common location of polymicrogyria is around the sylvian fissure, particularly the posterior aspect. Polymicrogyria involving the frontal, parietal or temporal opercula is classified as perisylvian polymicrogyria.[3] When bilateral and associated with pseudobulbar palsy (oropharyngeal dysfunction and dysarthria) and epilepsy, it is called CBPS.[1,2]

Patients with polymicrogyria restricted to the posterior portion of the sylvian fissure at the parietooccipital regions are difficult to diagnose due to the lack of neurological signs, relatively late seizure onset, difficulty in localizing seizure onset, and inability to recognize the cortical abnormality on computed tomography scans.[4] MRI reveals bilateral perisylvian and perirolandic malformations with exposure of the insula. The malformations are symmetrical in 80% of cases. Pathologic correlation reveals four-layered polymicrogyria in the affected areas.

Early onset of seizures was associated with the fact that patients with larger lesions were more likely to have earlier seizure onset, resulting in greater interference with ongoing cognitive development. To our knowledge, in contrast to previous case reports, our case was diagnosed at an early age.

Clinical findings of CBPS emphasized the importance of long-term follow-up, suggesting that the prognosis for epilepsy may not be predicted based on the early response to treatment or on the presence of structural encephalic abnormalities.

In conclusion, CBPS is a rare clinical-radiological entity characterized by pseudobulbar palsy, cognitive deficits and epilepsy associated with bilateral perisylvian cortical dysplasia on neuroimaging studies. The underlying abnormality is probably polymicrogyria. MRI has an important role in the diagnosis of this syndrome and associated other lesions.

REFERENCES

1. Kuzniecky R, Andermann F, Guerrini R. Congenital bilateral perisylvian syndrome: study of 31 patients. The CBPS Multicenter Collaborative Study. Lancet 1993;341:608-12.
2. Sejima H, Takusa Y, Kimura M, Tamaoki Y, Kishi K, Yamaguchi S. A variant case of congenital bilateral perisylvian syndrome with asymmetric findings on neuroimaging and septum pellucidum defect. Brain Dev 2001;23:131-4.
3. Barkovich AJ, Hevner R, Guerrini R. Syndromes of bilateral symmetrical polymicrogyria AJNR Am J Neuroradiol 1999;20: 1814-21.
4. Guerrini R, Dubeau F, Dulac O, Barkovich AJ, Kuzniecky R, Fett C, et al. Bilateral parasagittal parietooccipital polymicrogyria and epilepsy. Ann Neurol 1997;41:65-73.