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Mega Corpus Callosum, Polymicrogyria, and Psychomotor Retardation Syndrome

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Letters to the Editor

History

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ABSTRACT

The mega corpus callosum (MCC), polymicrogyria (PMG), and psychomotor retardation (PMR) is an extremely rare cerebral malformation syndrome. It may also be associated with megalocephaly and other dysmorphic features. There are only a few case reports of MCC-PMG-PMR syndrome in the literature.

Keywords: Corpus callosum, Polymicrogyria, Motor retardation, Magnetic resonance imaging

Mega Korpus Kallozum, Polimikrogri ve Psikomotor Retardasyon Sendromu

ÖZ

Mega korpus kallozum (MCC), polimikrogiri (PMG) ve psikomotor retardasyon (PMR) son derece nadir görülen bir serebral malformasyon sendromudur. Ayrıca megalosefali ve diğer dismorfik özelliklerle de ilişkili olabilir. Literatürde MCC-PMG-PMR sendromu ile ilgili sadece birkaç olgu bildirimi bulunmaktadır.

Süreç

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Anahtar sözcükler: Korpuz Kallozum, Polimikrogiri, Motor Retardasyon, Manyetik Rezonans Görüntüleme

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To the Editor

The mega corpus callosum (MCC), polymicrogyria (PMG), and psychomotor retardation (PMR) is an extremely rare cerebral malformation syndrome. It may also be associated with megalocephaly and other dysmorphic features. There are only a few case reports of MCC-PMG-PMR syndrome in the literature (1, 2, 3, 4).

An eight-year-old male patient presented with recurrent seizure episodes and psychomotor retardation. He was the third child of non-consanguineous parents. His gestational and perinatal histories were non-remarkable for any abnormality. His age-based psychomotor development was delayed, and he was mentally retarded. He had secondary generalized seizures of left side origin. He was taking antiepileptic treatment. Physical examination showed no marked dysmorphic features. There was, however, no cutaneous signs. He had a normal visual and auditory function. Serum and urinary laboratory analyses were within normal limits, as was his electroencephalogram. His genetic analysis revealed 46 XY. A brain magnetic resonance imaging (MRI) with 1.5 Tesla magnetic field was performed under mild sedation. MRI showed that the patient's corpus callosum was thicker than normal. Morphometric analysis was performed to calculate the length, thickness, and area of CC as well as the supratentorial-supracallosal compartment area. CC's morphometric analysis was performed on a midsagittal T1-weighted image. A comparison of his morphometric analysis with that of a normal subject of the same age revealed that our patient had a marked increase in CC thickness, and CC area, with the changes being more prominent in the genu of CC (Figures 1A, B).

Our patient's CC genu thickness was 15 mm. The normal thickness of the genu of CC has been reported to be 8.35±0.80 mm for children aged 4 to 18 years in the literature (1). Furthermore, our patient had bilateral diffuse fronto-temporo-parietal polymicrogyria and pontine hypoplasia (Figures 1C, D). There was also incomplete opercularization at the level of the bilateral Sylvian fissures. Patchy signal changes being prominent at periventricular and subcortical white matter areas were also detected. Bilateral caudate nuclei were enlarged and had a globular appearance. The ventricular system had a normal size. There was an appearance compatible with megalencephaly of the cerebral hemispheres and cerebellum. Patient's consent was obtained for this study.

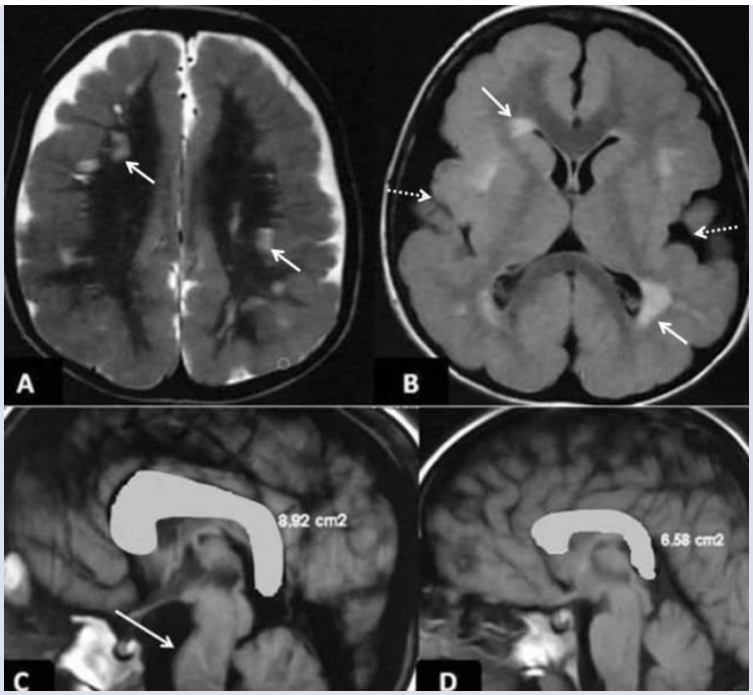

Figure 1. Axial (A) T2-weighted, and (B) FLAIR MR images show bilateral polymicrogyria; FLAIR image shows incomplete opercularization of Sylvian fissures (dashed arrows), and subcortical, and periventricular signal alterations (white arrows). It is also observed in megalencephalic appearance. (C) Sagittal T1-weighted MR image shows mega corpus callosum and pontine hypoplasia (white arrow). Corpus callosum area is 8.92 cm^2. (D) Sagittal T1-weighted MR image of a healthy subject of the same age shows that the corpus callosum area is 6.58 cm^2, and the pons appears normal.

Corpus callosum develops between the 8th and 20th weeks of gestation. Although the splenium of the corpus callosum is the thickest part in the normal morphology of corpus callosum, the genu of the corpus callosum was thicker than the splenium in our patient. Neuronal migration begins simultaneously with the CC

development. Thus, CC anomalies typically coexist with other brain anomalies, mainly neuronal migration anomalies. In contrast to corpus callosum anomalies like partial or complete agenesis, MCC is quite rare. Although MCC may occur incidentally, it may also be associated with certain other conditions (childhood schizophrenia, musicians). It may also develop as a result of trauma, inflammation, or non-inflammatory primary conditions such as megalencephaly, microcephaly, and polymicrogyria. CC thickening is quite rare in the absence of inflammation or neoplastic infiltration. Only two syndromes display this feature: Cohen syndrome and neurofibromatosis 1 (NF-1). Whereas CC thickening is along with a normal head size in NF-1, it is associated with microcephaly in Cohen syndrome. Among conditions causing CC thickening, inflammatory conditions (demyelinating pathologies, acute disseminated encephalomyelitis, diffuse axonal injury, etc.) are more common than non-inflammatory ones. CC thickening may also be associated with global developmental delay, facial dysmorphism, mental retardation, microcephaly, and visual symptoms (2, 3, 4). MCC may rarely coexist with PMG and PMR, as in our case.

MCC-PMG-PMR syndrome should be considered in the differential diagnosis of cases with mega corpus callosum, and polymicrogyria on MRI examination. An increase in the rate of detection of CC and associated anomalies have been noted with the widespread use of MRI. A careful evaluation of midsagittal MR images is of particular importance for the detection of CC anomalies among children with mental-motor retardation, facial dysmorphism, and a history of seizures.

Conflict of interest

There is no a conflict of interest

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