Leucoencephalopathy with Bitemporal Lobe Cysts in a Child with Developmental Delay

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Figure 1a and 1b: T2W axial images showing bilateral diffuse periventricular white matter, hyper intense lesions in both cerebral hemispheres with hyper intense bilateral cystic lesions in anterior temporal lobe and in parietal lobe. Corpus callosum, basal ganglia, thalami and internal capsule are spared

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AN Omani girl, born to consanguineous parents at full term with an insignificant peri-natal history, presented at 2 years and 8 months old at Sultan Qaboos University Hospital, Oman. She had had delayed psychomotor development during her first year of life. She had no history of trauma, seizures, or infection during infancy. She had walked at the age of 2 ½ years and was still not speaking although she seemed to understand simple commands. Physical examination showed only brisk reflexes at the lower extremities, and normal head circumference. Her routine and metabolic investigations were normal. Her MRI findings are shown.

**COMMENT**

Many reports have been published recently, describing patients with a new disease entity. These patients characteristically present with a non-progressive clinical course, associated with cystic changes in the anterior temporal lobes and increased signals on magnetric resonance imaging (MRI) suggestive of white matter changes. This disease, which is called leukencephalopathy with bilateral anterior temporal lobe cysts, can be differentiated clinically and radiologically from those of the better known progressive disorders presenting with white matter cysts: megalencephalic leukoencephalopathy with subcortical cysts, and the ‘vanishing white matter’ disease. The neuroradiological findings are distinct and vital in establishing the diagnosis. In clinical practice, variability of the phenotype has been noted. Patients may present with symptoms ranging from only mild spasticity in the lower limbs associated with normal cognitive function to severe disabling motor handicap with mental retardation and microcephaly.

To our knowledge, this is the first case to be described in the Arabian Peninsula. This condition differs from those previously known cystic leukencephalopathies and appears to be new, distinct entity. The pathological cause has not yet been defined and it is thought to be genetic in origin with autosomal recessive inheritance, though no gene has yet been identified.

**REFERENCES**
