

Leucoencephalopathy with Bitemporal Lobe Cysts in a Child with Developmental Delay

Almundher Al-Maawali,¹ *Amna Al-Futasi,² Raghad Abdwani,² Dilip Sankhla³

أعتلال المادة الدماغية البيضاء مع تجوفات ثنائية الجانب في الفص الصدغي لدى طفل يعاني من تأخر التطور الطبيعي

المنذر المعولي، آمنة الفطيسي، رغد عبدواني، ديليب سنكهلا

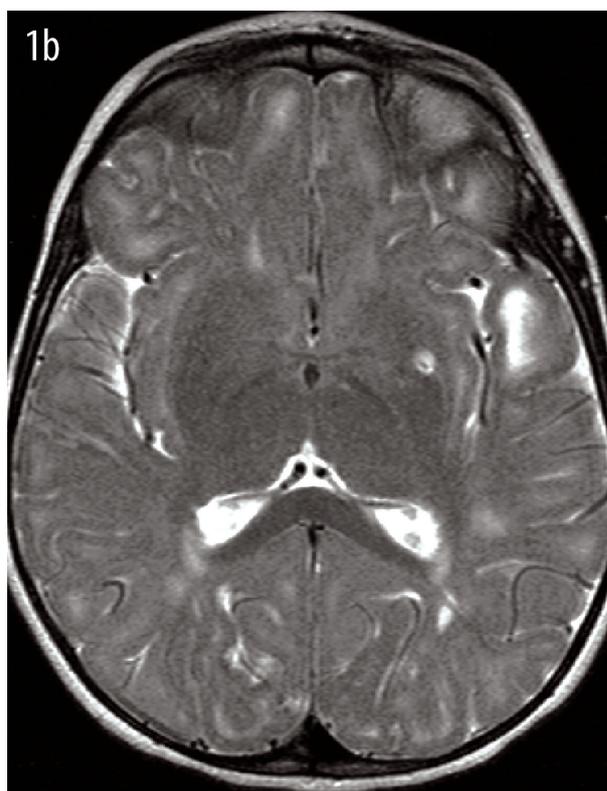


Figure 1a and 1b : T2W axial images showing bilateral diffuse periventricular white matter, hyper intense lesions in both cerebral hemispheres with hyper intense bilateral cystic lesions in anterior temporal lobe and in parietal lobe. Corpus callosum, basal ganglia, thalami and internal capsule are spared

¹Department of Genetics, College of Medicine and Health Sciences, Sultan Qaboos University Hospital, P. O. Box 35, Al-Khod 123, Muscat, Sultanate of Oman; ²Department of Child Health, College of Medicine and Health Sciences Sultan Qaboos University, P. O. Box 35, Al-Khod 35, Muscat, Sultanate of Oman; ³Department of Radiology and molecular imaging, Sultan Qaboos University Hospital, P. O. Box 35, Al-Khod 35, Muscat, Sultanate of Oman

*To whom correspondence should be addressed. Email: amnaf@squ.edu.om

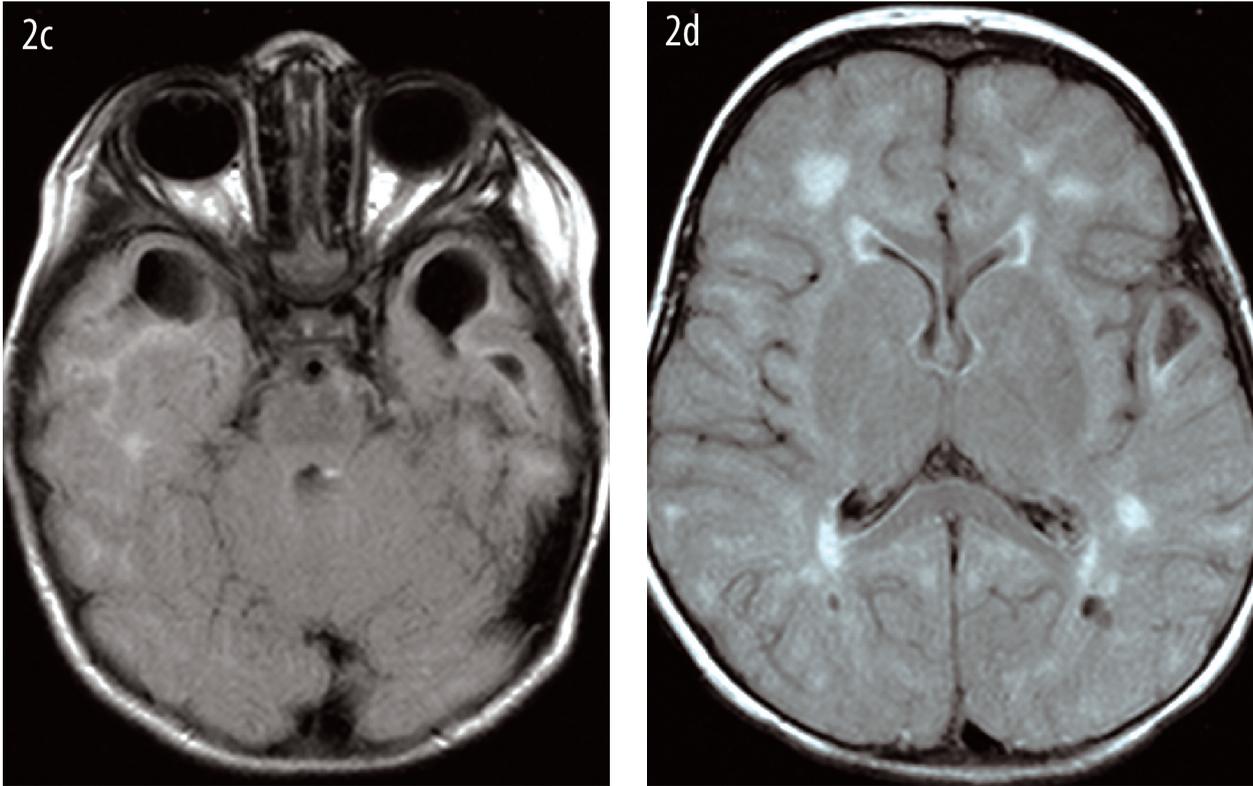


Figure 2c and 2d: FLAIR sequence axial images show cystic lesions in anterior temporal which are isointense to cerebrospinal fluid (CSF)

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 magnetic resonance imaging (MRI) suggestive of white matter changes.¹ This disease, which is called leukoencephalopathy 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,² and the ‘vanishing white matter’ disease.³ 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.³ To our knowledge, this is the first case to be described in the Arabian Peninsula. This condition differs from those previously known cystic leukoencephalopathies and appears to be new, distinct entity.⁴ 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

1. Bodensteiner JB, Kerrigan JF, Johnsen SD. Leukoencephalopathy with bilateral anterior temporal lobe cysts. *J Child Neurol* 2006; 21:419-422.
2. Singhal BS, Gorospe JR, Naidu S. Megalencephalic leukoencephalopathy with subcortical cysts. *J Child Neurol* 2003; 18:646-652.

3. Grosso S, Cerase A, De Stefano N, Marco LD, Galluzzi P, Galimberti D, et al. Non-progressive leukoencephalopathy with bilateral anterior temporal cysts: a case report and review of the literature. *Brain Dev* 2005; 27:73-77.
4. Sener RN. Leukoencephalopathy with patchy lesions, temporal cystic degeneration, and normal head size, *Comput Med Imaging Graph.* 2003; 27:87-91.