The rare association of leukoencephalopathy, cerebral calcifications, and cysts (LCC) is a very rare entity described in 1996. Since then, approximately a dozen cases have been reported in the current medical literature. A 9-year-old male was admitted to our hospital for the evaluation of generalized tonic-clonic seizures. There were no complications during pregnancy. His parents were healthy and not consanguineous. No abnormality was found on general physical and ophthalmologic examinations. Neurologic evaluation showed dysbasia. Complete blood exam, liver, renal and thyroid function tests, phosphate, alkaline phosphatase, and lactate levels were within normal limits. Serological tests for cytomegalovirus, Toxoplasma gondii, and HIV1/2 were all negative. Cervical, thoracic, abdominal CT and EEG examinations showed no abnormality. CT showed numerous foci of calcifications scattered through the basal ganglia, MRI Axial T2-weighted MRI image through the brainstem shows the pontine cyst. Coronal T1-weighted image shows the pontine cyst and its ring enhancement. The same findings shown in Axial T2, Axial T1 IR and sagittal T2. A telovelar approach to the fourth ventricle was performed and a supracollicular puncture was done and reoperated with improved symptoms. The clinical manifestations of LCC are according to lesion topography. Progressive calcifications in the basal nuclei, cerebellum and subcortical white matter are very accurately outlined by computed tomography. Diffuse white matter abnormalities and cysts are best appreciated on T2-weighted sequences; there is relative sparing of U-shaped fibers. Contrast-enhanced MRI presents with high signal intensities adjacent to cysts and calcifications, sometimes with mass effect, suggesting blood-brain barrier disruption.