Clinical History:
We report 4 cases of young children, including two females and two males, with ages ranging from 4 to 9 years, who presented with early onset seizure disorders refractory to medical treatment. They all displayed mild to moderate developmental delay involving mainly language, fine motor skills and below the average intelligence. Imaging of the brains showed different findings, with two cases displaying focal cortical dysplasia with periventricular nodular heterotopias, one case with small bilateral periventricular nodular heterotopias and one with a focal non-specific “thickening” of the cerebral cortex. Non-neurological anomalies were also recorded with one small septal cardiac defect, bowel malrotation at birth and bilateral abnormalities of upper extremities respectively in three of the patients. The neurological familial medical history was variable from case to case, ranging from unknown to a case with a sibling with Prader-Willi syndrome, one history of seizure in a maternal nephew and maternal migraine. All children underwent neurosurgical intervention with hemispherectomies.

Material submitted:  H&E section cerebral cortex from one of the cases

Points for discussion:  1. Main abnormal neuropathological finding
2. Diagnosis