**Abstract**

A 3 month old male infant presented with intractable seizures following administration of neem oil. Seizures were refractory in nature with features of raised intracranial tension and were managed with thiopentone, mechanical ventilation, midazolam infusion and mannitol.

Investigations revealed REYE’s syndrome like picture and diffuse cerebral oedema with frontal cortical atrophy in CT brain. Subsequently the child had presented with loss of gained milestone (social smile) and nonattainment of further developmental milestones, persistent hypotonia and myoclonic seizures on follow up at 5th month of life. Inview of myoclonic seizures, hypotonia and developmental regression with parental consanguinity, inborn error of metabolism was suspected.On evaluation, child was found to have significantly elevated levels of 2-hydroxyglutarate, succinate & adipate on Gas chromatography mass spectrometry (GCMS) which was suggestive of gutaric aciduria type II.

**Key words:** neem oil poisoning, glutaric aciduria.