CASE OF HEMICONVULSION HEMIPLEGIA–EPILEPSY SYNDROME ASSOCIATED WITH INFLAMMATORY–DEGENERATIVE IMAGING FINDINGS IN A TODDLER WITH ADRENAL INSUFFICIENCY.

ABSTRACT

Hemiconvulsion-Hemiplegia-Epilepsy (HHE) syndrome is an uncommon outcome of prolonged focal status epilepticus in childhood usually occurring during febrile illness followed by hemiplegia ipsilateral to the side of convulsions which is associated with seizures, cerebral hemisphere atrophy, global atrophy of one hemisphere and transient or permanent epilepsy. This is a report of 4 year male child with right sided hemiparesis, vacant stare and adrenal insufficiency with past history of altered sensorium and focal dyscognitive seizures. This is accompanied by radiologic evidence of acute cytotoxic edema in the affected hemisphere followed by chronic atrophy. MRI brain shows gliosis, restricted diffusion in the left cerebral hemisphere with reduced caliber of the left ICA, anterior, middle and posterior cerebral arteries.

KEYWORDS

Hemiconvulsion–Hemiplegia–Epilepsy; Adrenal insufficiency; status epilepticus.

CASE HISTORY:

We describe the case of a 4 year-old male child with adrenal insufficiency presented with vacant stares (~2min/episode) and right hemiparesis. The child was admitted in a tertiary care hospital at 3 years of age for altered sensorium, focal dyscognitive seizures and right hemiparesis.

IMAGING FINDINGS:

Initial MR showed cytotoxic edema involving the complete left cerebellar hemisphere with apparently normal MRA. MRI after 1 year showed gliosis with secondary volume loss; ex-vacuo dilatation of left lateral ventricle with deviated falx. There was reduced calibre of left ICA, anterior, middle and posterior cerebral arteries.

REFERENCES:


