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A RARE CASE OF PANAYIOTOPOULOS SYNDROME IN 1 YR 10 MONTHS BOY AT DEPARTMENT OF PEDIATRICS AND NEONATOLOGY, ETERNAL HOSPITAL, JAIPUR

Paediatrics

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ABSTRACT

Epilepsy affects 1% of the general population and 4% of children, encompassing heterogeneous seizure syndromes (1). These are defined by distinct aetiology, age at onset, seizure type, and electroencephalographic features, which taken together provide the key to diagnosis, prognosis, and optimal management. Over the past two decades various distinct paediatric epilepsy syndromes, such as Rolandic epilepsy, have been formally recognised (2). Panayiotopoulos syndrome is a new idiopathic childhood epilepsy, recently recognised by the International League Against Epilepsy (2,3). It is common, benign, and may mimic other common illnesses. The EEG does not reflect clinical course and severity. Panayiotopoulos syndrome can be best defined as idiopathic susceptibility to early onset benign childhood seizures with electroencephalographic occipital or extra occipital spikes, and manifests mainly with autonomic seizures also called as Benign occipital epilepsy (4). Large independent studies have accumulated impressively concordant information on the clinical and electroencephalographic features of this syndrome (4-9).

KEYWORDS

CASE REPORT

A, 1 yr10 m old boy, was presented with complaints of breathing difficulty for last 5 days & seizure like activity X 5 days, focal to begin with then generalised, manifested with deviation of eyes with lip smacking movements and rapid blinking of eyes associated with retching, vomiting and excessive salivation, multiple episodes, lasting for 20-30 seconds followed by crying and frothing from mouth, occurs specially when patient goes to sleep. On examination, HR - 100/ min (rhythmic, regular, without radio-radial or radio femoral delay) RR - 20/min (rhythmic, regular, thoracoabdominal), BP – 100/70 mmHg (right arm supine position), Temp - 98 o F (axilla), SPO2 96% on room air. No pallor, icterus, cyanosis, clubbing, edema, lymphadenopathy. In view of breathing difficulty foreign body was suspected for which virtual bronchoscopy was done which was within normal limits. In view of seizures MRI brain and EEG was planned. EEG showed, This EEG is abnormal. The abnormality in the form of diffuse slowing in theta and delta range. Paroxysmal generalized high voltage pseudo periodic slow wave discharges. MRI brain reveals, Patchy focal areas of T2/FLAIR hyperintensity, not showing any restricted diffusion /blooming, seen in bilateral peri-trigonal regions and subcortical regions of bilateral parieto-occipital lobes. MRI findings are suggestive of Patchy focal areas of T2/FLAIR hyperintensity, not showing any restricted diffusion /blooming, in bilateral peri-trigonal regions and subcortical regions of bilateral parieto-occipital lobes – likely representing terminal zones of myelination- Possibility of Panayiotopoulos Syndrome. A diagnosis of Panayiotopoulos Syndrome was made, after taking neurologist opinion. Initially one antiepileptic was started, and seizure episodes continued. So another antiepileptic drugs were added, and dosage were increased to control the seizures. After giving 3 Anti-epileptics drugs his seizures was under control. Lumbar puncture was done to rule out meningitis/ encephalitis, which was normal. He needed Oxcarbazepine, Levetiracetam and Gabapentin for stabilization.

He remained asymptomatic for 14 months on these drugs on follow-up. But again, in the month of April 2021, his seizures recurs and the dose of antiepileptics were increased as per the weight of the child. Seizures were kept on increasing and he was again admitted at Pediatric Department Eternal Hospital, Jaipur, for breakthrough seizures. He was evaluated to rule out infections and other causes of seizures. EEG showed abnormal showing occipital discharges maximum lasting for 17 seconds. After taking Neurology opinion antiepileptics dosage and drugs were revised and Gabapentin was stopped and Lacosamide was added to the treatment along with Levetiracetam and Oxcarbazepine. After 4 days his seizures were controlled, and he was discharged.

Gibbs and Gibbs (1952) (10-11) documented those occipital spikes are common in children with or without seizures. Peak age at first discovery is 4 years, with both EEG abnormalities and seizures usually disappearing in adult life (12,13). They also described, amongst the “atypical seizures of 38 patients with a spike focus in the occipital area” syncope (4%), strabismus and diplopia (1.5%), staring (2.2%), fortification figures (1.5%), formed visual hallucinations (0.7%), pressing hands to eyes (1.5%), vomiting (1.5%), headache (0.7%), and blindness (0.7%).

C.P. Panayiotopoulos studied (14) that 24 out of 900 adult and children patients with epilepsy, were found to have vomiting during an seizure. All the 24 patients were children before puberty with a similar clinical pattern consisting of partial seizures which were mainly nocturnal which is like in our case the child had seizures when he goes to sleep. Ictal vomiting was always concurrent with other epileptic manifestations, more often deviation of the eyes and impairment of consciousness. The initial part of the ictus was short or prolonged for hours with frequent "marching" to hemi-convulsions and generalised seizures. Seventeen of the 24 children suffered from benign childhood epilepsies (BCE) with complete remission in long follow-up; in our case child remained in remission for 14 months then he developed breakthrough seizures. A significantly higher association was found between ictal vomiting and the syndrome of BCE with occipital spikes (p less than 0.001) but not with centro-temporal spikes (p less than 0.2). The recognition of this association may have important theoretical implications. On clinical grounds, it may prevent unnecessary investigations and undue concern.

Concluding that Early-onset benign childhood occipital seizures is probably one of the most precisely described and studied common syndrome of childhood epilepsies and should be made widely known amongst pediatricians, neurologists and electroencephalographers & multiple anti-epileptic drugs may be required to control seizures. The prognosis of this illness is rather good then looks. Panayiotopoulos syndrome is remarkably benign in terms of seizure frequency and evolution. Autonomic status epilepticus imparts no residual neurologic deficit. The risk of epilepsy in adult life seems to be no higher than in the general population.

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