Research Paper

Volume : 3 | Issue : 11 | November 2014 • ISSN No 2277 - 8179

REFLEX SEIZURES

Pediatric Neurology

KEYWORDS : STARLE REFLEX, REFLEX SEIZURE,STARTLE EPILEPSY,STARLE DISEASE

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ABSTRACT

Reflex seizures are epileptic seizures which are consistently elicited by a specific stimulus(1). These seizures can arise in a 'spontaneous' unpredictable fashion without detectable precipitating factors, or they can be provoked by certain recognisable stimuli. These seizures are the result of sensory stimulation caused by the environment and hence it is also termed as environment epilepsy(2).Startle reflex, a protective brain stem reflectory reaction, is one among the many precipitating events resulting in reflex seizures. Exaggerated startle reflex is seen in a few disorders in infants and children.Careful history, thorough physical examination and relevant investigations to confirm the specific diagnosis is important as,both the treatment and prognosis varies between different diseases.Here we report a case of reflex seizures precipitated by startle reflex in a child.

CASE REPORT

A one year six months old female child, first born to non-consanguinous parents, was brought by the mother with complaints of abnormal movements following any sudden stimuli for the past 3 months like jerky movements of all 4 limbs with uprolling of eyes which recovers immediately within 10 seconds for about 10-15 episodes per day. The stimuli being any sudden noise like even the sound produced while eating crispy snacks or sudden touch like even spilling of water while drinking. There is no associated urinary or stool incontinence and no involuntary movements noticed. Child is developmentally normal for age with uneventful birth history.

On examination, child was comfortable with mother with vitals and anthropometry being normal for age. Systemic examination including fundus was normal. When a sudden stimuli was produced,child had exaggerated startle reflex. A working diagnosis of startle disease,reflex seizure,and tay-sachs disease was considered and evaluated. MRI BRAIN was normal and EEG showed epileptiform activity. Hence a diagnosis of reflex seizure was confirmed and child was started on sodium valproate. On follow up,during second week. Child showed marked improvement to treatment.

DISCUSSION

Reflex seizure is a condition in which seizures can be provoked by an external stimulus or, less commonly, internal mental processes. Individuals with pure reflex seizure do not suffer spontaneous seizures, but they may co-exist with spontaneously occurring seizures(3). Reflex seizure of different types have been known for centuries. The first reference to reflex epilepsy is attributed to Apuleius lucius, a roman philosopher, in his APOLOGIA AND FLORIDA, long back in 150 A.D. The oldest clear reference to photosensitive epilepsy is by a Greek pediatrician, Soranus of Ephesus in 2nd century A.D. Later it was documented in animals by Italian school of neurophysiologists in 1920s and 1930s.But it was only in 1929 with the advent of EEG by Berger , a new era started for the study of reflex and photosensitive epilepsies(4). Reflex seizures have a prevalence of 4 – 7% among patients with epilepsies(1,5).Etiologically reflex seizures are idiopathic, symptomatic, or probably symptomatic and are determined by the specific precipitating stimulus and the clinical/EEG response.

'Epilepsies characterised by seizures with specific modes of precipitation' is the term used in the 1989 ILAE classification.*'Reflex'* is the preferred name in the new ILAE diagnostic scheme. According to new INTERNATIONAL LEAGUE AGAINST EPILEPSY(ILAE), reflex seizure is defined as "Objectively and consistently demonstrated to be evoked by a specific afferent stimulus or by activity of the patient. Afferent stimuli can be: elementary, i.e. unstructured (light flashes, startle, a monotone) or elaborate i.e. structured. Activity may be elementary, e.g. motor (a movement); or elaborate, e.g. cognitive function (reading, chess playing), or both (reading aloud)." The stimulus evoking an epileptic seizure is specific for a given patient and may be extrinsic, intrinsic or both.Extrinsic stimuli are simple, such as flashes of light, elimination of visual fixation and tactile stimuli &complex, such as reading or eating.The latency from the stimulus onset to the clinical or EEG response is typically short (1–3 s) with simple stimuli and long (usually many minutes) with complex stimuli.Intrinsic stimuli are elementary, such as movements & elaborate, such as those involving higher brain function, emotions and cognition (thinking, calculating, music, or decision-making). The response to the stimulus consists of clinical and EEG manifestations, alone or in combination. EEG activation may be subclinical only, that is without overt clinical manifestations. Conversely, ictal clinical manifestations may be triggered without conspicuous surface EEG changes. Startle is one among the various precipitating stimulus for reflex seizures(6).*Reflex seizures* may be generalised, such as absences, myoclonic jerks or generalised tonic clonic seizures &focal, such as visual, motor or sensory. Myoclonic jerks are by far the most common form of reflex seizures. The role of the EEG is fundamental in establishing the precipitating stimulus in reflex epilepsies, because it allows subclinical EEG, or minor clinical ictal events to be reproduced on demand by application of the appropriate stimulus. Some types of reflex seizures can occur in the context of symptomatic, localization-related epilepsy, a brain magnetic resonance imaging (MRI) scan should be obtained to identify the etiology and potential structural abnormalities underlying the epilepsy. The treament of reflex seizures is dependant mainly on avoiding the stimulus or either by preventing or modifying the stimulus. A standard antiepileptic drug has to be started preferably sodium valproate . sodium valproate monotheraphy has a success rate of 73-86%. Levitracetam, ethosuximide, lamotrigine are second choice drugs. The prognosis is excellent in reflex seizures. The following table helps to differentiate reflex seizure by startle from startle disease and startle epilepsy as the etiology, pathology and treatment varies for each disease.(table 1)

Conclusion:

Startle reflex, though is a normal reflex in individuals, exaggerat-

ed startle has to be evaluated. If untreated it will lead to neurological and psychological problems, both to the parents and the kids. Avoidance, prevention, or modification of the provocative stimulus is the key point of management. Careful evaluation is essential as prognosis differs among different etiologies

TABLE 1 : Difference between REFLES SEIZURE BY STARTLE with STARTLE EPILEPSY & STARTLE DISEASE.

Name	Reflex seizure	Startle epilepsy (6)	Startle disease (7,8)
Age	Infants &childhood	Variable	At birth (stiff baby syndrome)
Pathology	Decreased epileptogenic threshold	Cortical dysplastic lesions, anoxic encephalopathy	Abnormal gene in glycine receptor
Response	Mainly myoclonic jerks	Tonic phase after startle	Increased startle
EEG	Generalised epileptic discharges	Discharges reflecting the brain lesions	Always normal
Treatment	Avoid stimuli, sodium valproate	Carbamazepine, surgery	Clonazepam
Prognosis	Excellent	Guarded	Variable

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