Hemifacial Seizures and Cerebellar Tumor: A Rare Co-existence

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We report a 4-year-old boy with multiple daily episodes of abnormal eye movements, hemifacial spasms. Neuro-imaging revealed a cerebellar tumor. We believe that this association constitutes a rare but important syndrome of epilepsy characterized by seizures of cerebellar origin.

Key words: *cerebellum, epilepsy, Seizure, Tumor.*

Hemifacial spasms occur most commonly in adults above the age of 40 years and are secondary to compression of facial nerve by vascular loop. In children this entity is very rare and is usually secondary to tumors in cerebello-pontine angle(1). Cerebellar ganglioglioma is also rare in pediatric populations. Coexistence of these two rare entities constitutes a unique clinicopathological syndrome of sub-cortical epilepsy.

We report a case with hemifacial spasms and cerebellar ganglioglioma, which is a co-existence of two rare entities and may constitute unique syndrome of sub-cortical epilepsy.

Case Report

A is a 4-year-old boy, product of a normal delivery, presented with nystagmoid jerks of eyes multiple times a day noticed by the parents since the age of two years. His development was normal for the age.

There were paroxysmal nystagmoid jerks noted in his right eye multiple times in a day. The eye deviation was in semicircular manner. These jerks had no directional preponderance. It was associated with hemifacial spasms of the right side. There were no limb movements, precipitating factors or aura. The episodes used to last briefly for few seconds. The child was conscious and responsive during the seizures. There were no post ictal phenomena. These occurred in awake as well as during sleep state. The seizures were uncontrolled with medications.

The neurological and ophthalmological examinations were essentially normal.

CT brain revealed calcified lesion without perifocal edema in right cerebellum, causing mass effect over 4th ventricle. EEG showed frequent spike and wave activity from ipsilateral temporal cortex, without focal features and without sub-cortical spread or generalization.

Magnetic resonance imaging revealed well defined non-enhancing mass lesion in right cerebellar hemisphere reported as cerebellar ganglioglioma or gangliocytoma.

Positron emission tomography scan revealed increased metabolism in the right paramedian area corresponding to the calcified area on CT scan.

Seizures responded to a combination of valproic acid 40 mg/kg/d and Clobazam 0.5 mg/kg/d. The neurosurgeon was consulted. The decision of surgery was postponed in view of good seizure control and no mass effect of tumor on vital structures along with its surgical inaccessibility. The patient was monitored with serial scans and repeat neuroimaging did not reveal any increase in size of the tumor.

Discussion

Hemifacial spasms and cerebellar ganglioglioma are rare entities in children (1). Till date this coexistence is reported in ten patients with striking similarity in the presentation(2).

John Hughlings Jackson was singularly responsible for developing the modern concept of epilepsy in early years of 19th century(3). Classic doctrine dictates that epileptic seizures arise only in the cerebral cortex and subcortical structures such as cerebellum, brainstems have only neuromodulatory

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influence on cerebral epileptic activity(2,3). The issue whether seizures arise from cerebellum is controversial. Sub-cortical structures are thought to have inhibitory effects on cortical epileptic activity through neurochemical modulation(4).

Epilepsy of sub-cortical origin has been demonstrated in animal models(3). More recently seizures of subcortical origin have been documented in humans with hypothalamic hamartomas and cerebellar dysplastic lesions(3). The epilepto-genicity of these hamartomas comes from aberrant connectivity of neuronal structures or from intrinsic epileptogenicity of dysplastic neurons(4-6).

Introduction of depth electrodes suggested the cerebellar origin of seizures. Typical motor manifestation in response to cerebellar stimulation includes ipsilateral facial grimacing, ipsilateral and contra lateral head and eye deviation, nystagmus and alteration of tone and posture and autonomic changes(1). The ictal semiology of hemi facial seizures *i.e.*, spasm of hemifacial muscles, head and eye deviation, nystagmoid jerks and cardio respiratory dysfunctions are consistent with dysfunction of the cerebello pontine region. And it can be corroborated by imaging, depth recordings & PET scans. These movements were also shown to resolve after surgical excision of these lesions.

Review of literature has identified 10 children with clinical and radiological finding similar to our patient. All had hemifacial spasms; few had additional features of tonic head and/or eye deviation and arm movements. Surgical exploration was obtained in 4 out of which 3 had ganglioglioma and one had low-grade astrocytoma(1).

Both hemifacial seizures and cerebellar ganglioglioma are rare entities in pediatrics. This co-existence is unique clinicopathological syndrome and has an important implication in current nosologic and pathogenetic concept of epilepsy and should be included as a new epileptic syndrome.

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