Dystonic Storm: An Atypical Presentation of Subacute Sclerosing Panencephalitis

Subacute Sclerosing Panencephalitis (SSPE) typically presents with progressive myoclonic epilepsy and cognitive regression. We report the case of a child with SSPE who presented with dystonic crisis.

An 8-year-old unimmunized, previously asymptomatic girl, presented with dystonic posturing involving right sided limbs and cognitive regression. Dystonia had started acutely – two weeks prior to hospital admission. At onset, dystonia occurred intermittently at 15-30 minute intervals, each episode lasting about 2-3 minutes. Severity and frequency of dystonia progressed rapidly. At presentation, the child had near-continuous hemidystonia involving right-sided limbs in awake state, which abated during sleep. There was no history of fever, rash, myoclonic jerks, animal bite or exposure to toxins. On examination, the child was unresponsive, apathetic and did not recognize her parents. Her vital parameters and anthropometry were unremarkable, and meningeal signs were absent. She had brisk deep tendon reflexes and extensor plantar reflex. Kayser Fleischer ring (KF ring) was not detected in ocular slit-lamp examination .

Her hemogram, liver and kidney function tests, serum ceruloplasmin, thyroid profile and anti-nuclear antibodies (ANA) were normal. Magnetic resonance imaging (MRI) of brain revealed discrete, scattered hyperintensities involving bilateral occipital lobes and left globus pallidus. Electroencephalography (EEG) showed generalized slowing, absent sleep-markers and quasi-periodic high-amplitude generalized slow waves (left more than right). The child was diagnosed to have SSPE in view of her highly elevated CSF and serum anti-measles IgG titres (250 and 125 times, respectively; normal being <2.0).

Her dystonia responded partially to sequential treatment with increasing doses of oral trihexyphenidyl hydrochloride, lorazepam injection and midazolam infusion. Initiation of oral carbamazepine (10 mg/kg/day) led to marked symptomatic control. Child did not have any features of rhabdomyolysis. She was initiated on isoprinosine besides other supportive therapy. Parents were counseled regarding child's prognosis and she is being followed-up on out-patient basis.

Prominent dystonia and dystonic storms are atypical presentations of SSPE [1,2]. Hemidystonia, as seen in index child, has also been reported in SSPE [3]. Carbamazepine is known to be an appropriate drug for effective control of myoclonus in SSPE [4]. However, dramatic response to carbamazepine as seen in the index child, is rare.

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