CASE REPORT

Status Dystonicus: A Rare Complication of Dystonia

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A severe episode of dystonia refractory to standard drug therapy has been labeled as status dystonicus or dystonic storm. We report the development of this complication in a 10-year old boy with idiopathic torsion dystonia, the probable precipitating factor being either an infection or introduction of clonazepam.

Key words: *Child, Dystonia, India, Management, Status dystonicus.*

Patients with primary and secondary dystonic conditions occasionally develop severe episodes of generalized dystonia and rigidity, which may be refractory to standard drug therapy(1). This condition has variously been labeled as 'status dystonicus (SD)' or 'dystonic storm'(2), or the patients labeled as 'desperate dystonics'(3). The condition is quite rare with less than 40 episodes reported in the literature at the last count, and none from India(4). We herein describe the clinical presentation and management of an episode of SD in a pediatric patient with idiopathic torsion dystonia.

CASE REPORT

A 10-year-old boy, a known patient of idiopathic torsion dystonia, presented with a one-month history of progressive worsening of dystonia. The child was a product of a non-consanguineous marriage, born of a normal pregnancy and delivery, and had been developing normally till 5 years of age. There was no family history of dystonia, and hepatic or neurological disease. At the age of five years, the parents noted the child to have an abnormal gait. Subsequently, there was gradual onset of dystonia of his legs (left followed by right) apparent over a 6-month period. The dystonia gradually spread to involve all four limbs. There was no diurnal variation. He received treatment at multiple centers with various diagnoses including, seizure disorder, cerebral palsy, conversion reaction, Wilson disease, etc. The various medi-cations received at different times, till one month prior to presentation (mid-march 2008), included multivitamin combinations, Vitamin E, calcium, Vitamin D, zinc, alprazolam, trihexyphenidyl, tizanidine, tetrabenazine, penicillamine and sodium valproate, in addition to many medications from the various traditional Indian systems of medicine (*Unani*, *Ayurveda*, Homeopathy, etc.). He had been investigated for Wilson disease with negative results.

Prior to this episode of dystonia (early march), the child could not walk, could sit without support, could feed himself by hand (with spillage), but had no other abnormal movements and had normal speech. In mid-march 2008 (one month prior to presentation), the child changed physicians and two new drugs were added to the previous treatment *viz*., clonazepam and syndopa (levodpoa with carbidopa). Over the next 2 weeks, the child had increased dystonia, including the axial musculature. In addition, he had choreiform movements, slurring of speech, drooling of saliva and difficulty in swallowing. This was associated with excessive

exhaustion and pain, and the child could not sleep. He had no metabolic derangements, no impairment of respiratory function and no hypoxemia. Ten days prior to admission, the child had developed a gluteal abscess, which was managed with incision and drainage, and oral co-amoxyclav.

At our center, initial medical treatment consisted of increasing the dose of trihexyphenidyl (4.5mg/d) and diazepam (30mg/d), and continuing with clonazepam (1.5 mg/d) and L-dopa (100mg/d). Transfer to ICU could not be arranged. Over the next few days, there was no improvement in the dystonia, and sleep disorder. After a detailed review of the drug history, the possibility of choreiform movements being drug-induced was also entertained, possibly due to trihexyphenidyl or L-dopa. Further, the status dystonicus could also have been precipitated by the drugs. On this premise, trihexyphenidyl and L-dopa were gradually tapered (to avoid the possibility of malignant hyperthermia) and, diazepam and clonazepam increased. Over the next three weeks, the dystonia gradually decreased, and, pain and exhaustion subsided.

DISCUSSION

Status dystonicus (SD), first recognized by Jankovic and Penn in 1982, has been defined as "increasingly frequent and severe episodes of generalized dystonia, which necessitate urgent hospital admission. Patients have one or more of the following life threatening complications: bulbar weakness compro-mising upper airway patency with the risk of pulmonary aspiration; progressive impairment of respiratory function leading to the development of respiratory failure, exhaustion and pain; and metabolic derangements"(1). Although a life-threatening disorder, the condition is not common, with two large series of total 17 patients; 12 over a 10-year period from various centers in UK(1), and 5 patients from four neurology centers from Brazil(5). Rest of the literature consists of multiple case-reports in both adults and children(2,3,6).

Several drugs and surgical procedures in different combinations have been tried in SD, but no definite data seems to be available on the best therapeutic stategy(4). Although orally active drugs may occasionally arrest SD(3,5), the current literature favors using intravenous agents for deep sedation(2,4). As the patients have intense muscle activity, they are liable to develop metabolic complications such as rhabdomyolysis, that may lead to acute renal failure(1). In addition, bulbar and respiratory complications may ensue, thereby requiring tracheal intubation; in addition to hyper-pyrexia, muscle exhaustion, pain and dehydration. In view of these, SD patients should be routinely managed in intensive care settings(4). This also facilitates the currently accepted management strategy for severe SD, which consists of deep sedation under muscle paralysis and assisted ventilation(4). The sedation may be done with increasing doses of intravenous infusion of midazolam (30-100 µg/kg/hour), to which propofol (0.5-2.0 mg/kg/hr) may be added(4). The duration of sedation is determined empirically by intermittently reducing the sedation and evaluating the child. Intrathecal baclofen may also be used, although the literature does not provide clear cut evidence for efficacy(6,7). Second-line strategies, especially in those with progressive disorders, involve electrical deep brain stimulation of globus pallidus internus(8) or bilateral pallidotomy(5,9).

The most important differential diagnoses are neuroleptic malignant syndrome and malignant hyperthermia. This is more important because drugs used in the treatment of dystonia, such as tetrabenazine (being used in this child also) and lithium, as well as levodopa withdrawal, have all been implicated as causing such a malignant syndrome(1,5,10). Of the 30 patients (37 episodes) reported in literature, 25 (86.7%) were males and 17 (56.7%) younger than 14 years, and 3 each worsened and died, respectively(4). Majority of them required treatment in ICUs with intravenous sedation or muscle paralysis with ventilation, but a few have responded to oral therapy also(3,5). Addition of clonazepam and infection/stress (both present in this child) have been implicated as precipitating factors in quite a few patients(4). Although clonazepam addition seems to be incriminated in this child also, but the child improved with continuation of clonazepam; which disfavors this possibility. Coincidental onset of SD with clonazepam has

previously also been reported, with no improvement on stopping the drug(1). Clonazepam has also been used for SD management(1), as in our patient. Trauma, surgery and infection are frequently reported trigger factors(5), and two of these were also present in this child.

We have reported a pediatric patient with status dystonicus, a rare, life-threatening complication of dystonia. Both pediatricians and pediatric neurologists should be aware of the condition, so that it is recognized early and management can be initiated. The management and outcome are highly variable but a general consensus on the use of IV sedation and ICU management is currently evident from literature.

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