Movement Disorder Emergencies: When "Too Much" or "Too Little" Movement Needs Urgent Attention!!

Although most patients with movement disorders (MD) present in outpatient services and are managed in ambulatory settings, there are instances when they land up in neurological emergencies, either due to a rapidly appearing new MD or acute exacerbation of a chronic MD.[1] MD emergencies (MDEs) include acute or subacute onset progressive neurological disorders having MD as their predominant manifestation, with a likelihood of significant morbidity or mortality if not managed urgently.[2] Based on the predominant MD phenomenology, they can be classified into either hyperkinetic or hypokinetic MDEs.[3,4] While chorea, ballism, dystonia, tremor, myoclonus, and tics comprise the hyperkinetic group, acute parkinsonism, neurolept malignant syndrome (NMS), serotonin syndrome (SS), and malignant catatonia are the major hypokinetic MDEs.[3,4] Larger studies evaluating MDEs are lacking, and most of the literature consists of case reports or case series.

In this issue of the journal, Bhoyar *et al*.[5] have performed a single-center, prospective, observational study to evaluate the clinical profile (disease onset, duration, MD phenomenology, MD severity, and precipitating factors), investigative features (biochemical, neurophysiological, and neuroradiological findings), and outcome of patients presenting with MDEs to the neurology service at a South Indian tertiary care center, over a period of 26 months. The disorders were classified as hyperacute (onset in min to h), acute (onset within a day), subacute (onset within days to weeks), and chronic (onset duration >4 weeks).

Of the 45,000 patients attending the neurology emergency, 90 (0.2%) patients had MDEs. Functional MDEs and those failing to provide complete clinical information were excluded, with 71 patients with MDEs entering the final analysis. The mean (SD) of patients with MDEs was 45.4 (24.4), with a male: female ratio being 1.29. Although symptoms started acutely in two-thirds of patients, the mean duration from onset to presentation was 4.8 days. MDEs in 65 patients (91.5%) were hyperkinetic in nature, with chorea (59.2%) and dystonia including dystonic tremor (16.9%) being the most common. Myoclonus (9.8%), dyskinesia (2.8%), tics (1.4%), and opsoclonus myoclonus syndrome (1.4%) were the other hyperkinetic MDEs. Of the six patients (8.5%) with hypokinetic MDEs, acute parkinsonism was reported in four (5.6%) patients with NMS and SS seen in one (1.4%) patient each. Metabolic factors including hyperglycemia (29.6%), autoimmune including infection-induced autoimmunity (17%), and stroke (12.7%) were the most common etiological factors observed. The clinical status of nearly all patients (97.1%) either improved or remained the same, that is, did not worsen.

One of the recently published Italian studies reported MDEs in 1.4% of all emergency patients requiring neurological attention, with nearly three-fourths of them being hyperkinetic MDEs.[6] However, the phenomenology and etiology spectrum of MDEs in the Italian cohort differed from those of the present study. They reported tremor (19.8%) as the most common MDE, followed by myoclonus (17.7%), dystonia (15.6%), and chorea (11.4%). Acute parkinsonism (15.6%) was the most common hypokinetic MD. Although drug-induced (29.2%), neurodegenerative disorders (15.6%), and brain lesions (11.5%) dominated the responsible aetiological factors, a substantial proportion of cases were functional (19.8%).[6] A higher prevalence of diabetes mellitus (nearly 8% in patients aged above 20 years) and hypertension (one-fourth of the population aged above 18 years) in the India population may account for hyperglycemia and stroke being the common causes of MDEs in the present study.[7,8] Differences in genetic and epigenetic factors of the study populations might have contributed to these dissimilarities. Moreover, the present study did not include functional MDEs.

The present study included 15 patients (21.1%) aged below 18 years, with a mean age of 10.6 years, and all presenting with hyperkinetic MDEs. Dystonia (33.3%), chorea (26.7%), and myoclonus (20%) were common phenomenologies. Hyperkinetic MDEs predominated in the previously reported pediatric studies.[9-11] Although an Indian study reported myoclonus (27%), dystonia (23%), and choreoathetosis (21%) as the most common MDEs,[9] chorea (38.5%) and tics (44.5%) were most frequently reported by Dale *et al*. and Raucci *et al*., respectively.[10,11] Autoimmune and infectious diseases were the most common etiological factors in 60% of the present cohort,[5] but Goraya *et al*.[9] reported the same in only 35% of their cases. More than one-fourth of their cases had nutritional or metabolic aetiological factors, which might have resulted due to the low mean age of their study population (4.9 years).[9] Although neuropsychiatric disorders (51.2%) including tics, functional MDEs, and idiopathic stereotypies predominated in the study by Raucci *et al*.,[11] Dale *et al*.[10] reported inflammatory causes including autoimmune and infections in 42.3% and functional MDEs in 23.1% of their cases.

In the most common MDEs, i.e., choreiform MDEs, the authors reported hyperglycemia (43.9%), stroke (19.5%), and autoimmune (14.5%) as the common causes. Unilateral presentation including hemichorea (50%) and unilateral face (11.9%) or upper limb (7.1%) involvement was common in choreiform MDEs.[5] In patients with diabetic striatopathy, the authors observed hemichorea and generalized chorea in 84.2% and 10.5% of patients, respectively,[5] similar to that reported in a recent descriptive analysis involving

176 patients.[12] In the dystonic MDEs group, drug-induced dystonia was the most common cause, with risperidone, ondansetron, prochlorperazine, and levosulpiride being the offending agents.[5] A recent Indian study reported seven cases of levosulpiride-induced oromandibular dystonia.[13] In the group of myoclonic MDEs, SSPE (42.9%) was the most common cause. A child with tic disorder developed severe motor tics (tic status) and showed an excellent response to tetrabenazine. Tic status had been previously reported by Sachdev *et al*.[14] Amongst the four patients with acute parkinsonism, two patients on deep brain stimulation developed implantable pulse generator depletion, similar to a couple of previous reports.[15,16] Amongst the other two patients manifesting acute parkinsonism, one had multiple neurocysticercoses and the other had osmotic demyelination syndrome.

In addition to the referral bias and reduced neurological emergency visits due to the COVID-19 pandemic overlap with the duration of the study, the lack of inclusion of functional MDEs and anti-psychotic induced MDEs were the major limitations of this study. However, the authors provided an interesting account of MDEs in the Indian context. Although uncommon, MDEs must be identified and managed promptly to reduce morbidity and mortality in the affected patients.

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