# Paroxysmal Dystonia in inflammatory disorders of the central nervous system. A southeastern Michigan cohort study.

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## Background

Paroxysmal Dystonia (PD) is a recurrent, neurological symptom characterized by sustained muscle contraction, frequently causing twisting and repetitive movements, or abnormal postures that persists seconds to minutes. These infrequent symptoms are less recognized in general practice although they have been described in inflammatory disorders of the central nervous system (CNS) specifically, in Multiple Sclerosis (MS) and Neuromyelitis Optica (NMO)

## Objective

To evaluate the presentation and treatment of paroxysmal dystonia (PD) in patients with inflammatory disorders of CNS.

## Design/Methods

Study setting was in a large integrated health care system serving residents of southeastern Michigan. Over the course of 5 years, 17 patients with inflammatory disorders of CNS presented with paroxysmal symptoms. Electronic medical records were reviewed retrospectively and data was collected on diagnosis, socio-demographic factors, neurological symptoms, MRI imaging, treatment regimen and treatment response. Descriptive statistics were used to characterize the PD cases. The Institutional Review Board approved this study.

## Results

### Cohort Characteristics and Socio-demographic Profile

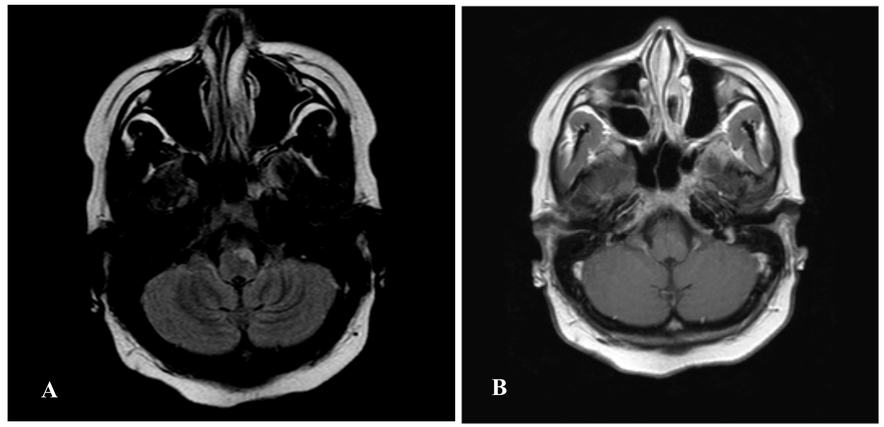
| Characteristic | | MS (N=8) | NMO (N=3) |
|---|---|---|---|
| Age at symptom onset | Mean (S.D.) | 46.1 (10.7) | 47.6 |
| | Median (Range) | (35-55) | (36-60) |
| Gender, N (%) | Male | 4 (50%) | |
| | Female | 4 (50%) | 3 (100%) |
| Race, N (%) | African American | 5 (62.5%) | 2 (67%) |
| | Caucasian | 2 (25%) | 1 (33%) |
| | Other | 1 (12.5%) | |

### Presentation of Paroxysmal Dystonia in Correlation to Stage of Disease

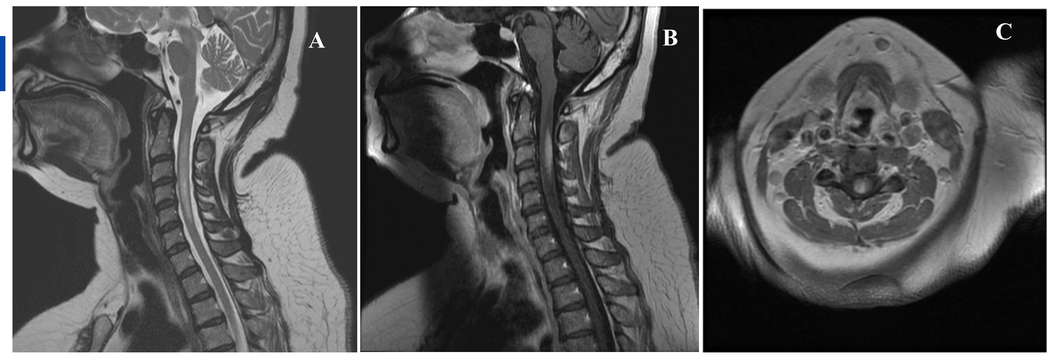
| | PD as initial attack of MS | PD after established diagnosis of MS |
|---|---|---|
| MS (n=8) | 4/8 (50%) | 4/8 (50%) |
| | **PD after initial TM attack** | **PD in patient with established diagnosis of NMO who had suffered a relapse** |
| NMO (n=3) | 2/3 (67%) | 1/3 (33%) |

PD=paroxysmal dystonia, MS= multiple sclerosis, NMO=neuromyelitis optica

## Results




**Case 1:** 35-year-old female who presented with recurrent episodes of intermittent right sided dystonic movements. Her right hand would draw up and her hand would tighten: her toes would also curl up. Episodes lasted about 10 seconds and occurred several times per day. MRI brain shows, new extensive areas of T2 hyperintensity within the brainstem. There is a plaque-like area of T2 hyperintensity along the left ventral medulla (A), which is new with some patchy enhancement as shown in post-contrast T1 image (B).




**Case 2:** 60 year old female has presented initially with shock like sensation over her right shoulder which over the course of five days had then developed right arm and leg weakness. Initial MRI of cervical spine was consistent with extensive transverse myelitis, CSF revealed presence of oligolonal bands and her NMO antibodies were positive concluding a diagnosis of NMOSD. A month later she reported spasms on the right side of the body involving the right hand and she had extension of her right leg with right foot turning in. Spasms occurred at a rate of one every 5 minutes. Sagital T2 MRI of C-spine (A), showes increased T2 signal spanning from the level of C2 to C4 predominantly within the central cord which tapers at its superior and inferior margins. There is also mild cord expansion. T1 images with contrast (B), reveal enhancement of previously noted area of cord expansion spanning from C2-C4. Axial T1 post contrast (C), reveal a right hemicord involvement.

## Results

**MRI Findings:**

T2 FLAIR MR images in MS patients with PD showed new lesions in areas of:

- Midbrain (1/8)
- Pons (1/8)
- Medulla (1/8)
- Posterior limb of internal capsule (1/8)
- Thalamus (1/8)
- Cerebral peduncle (1/8)
- Cerebellar peduncle (1/8)

All NMO patients had longitudinal extensive cervical cord lesion.

### Therapeutic agents and treatment response

| | Monotherapy | N (%) (distribution of patients on specific agent) | Combination therapy | N (%) (distribution of patients on specific agent) | Dose range for specific agent used | Time to symptom resolution |
|---|---|---|---|---|---|---|
| MS (n=7)* | Carbamazepine<br>Oxcarbazepine<br>Gabapentin | 5 (71%)<br>1 (14%)<br>1 (14%) | None | None | Carbamazepine 200-600 mg/day<br>Oxcarbazepine 300 mg/day<br>Gabapentin 1200 mg/day | ≤ 8 weeks |
| NMO (N=3) | None | | Carbamazepine<br>Oxcarbazepine<br>Gabapentin<br>Acetazolamide<br>Baclofen<br>Valium | 2 (67%)<br>1 (33%)<br>2 (67%)<br>2 (67%)<br>2 (33%)<br>2 (33%) | Carbamazepine 400-2400 mg/day<br>Oxcarbazepine 600 mg/day<br>Gabapentin 1500-3600 mg/day<br>Acetazolamide 1000 mg/day<br>Baclofen 30 mg/day<br>Valium 15 mg/day | ≈ 24 months |

* 1/8 patients in MS group, required no treatment given short symptom duration.

## Conclusion

Recognition of paroxysmal dystonia as a neurological symptom in MS is important, especially since this can be a presenting feature of the disease. In NMO it is important to be aware that these symptoms can occur after the active stage of transverse myelitis. Unlike many other manifestations of MS, paroxysmal dystonia is frequently abolished with monotherapy, whereas this manifestation in the NMO group requires a multi-therapeutic approach and a longer duration before symptom resolution.

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