CAROLINAS REHABILITATION



Introduction

Status dystonicus is a rare medical emergency which manifests as a life-threatening hyperkinetic movement disorder. While this complication typically occurs in patients with a known history of dystonia for a mean duration of 6 years, this case demonstrates the development of status dystonicus in a patient with a history of dystonia for only weeks.

Case Report

A 15-year-old male s/p 4 days intubation and sedation following cardiac arrest and CPR due to status asthmaticus presented to inpatient rehabilitation with apraxic speech, poor visual acuity, and bilateral upper greater than lower extremity weakness. His outside hospital discharge noted significant agitation which was difficult to control despite liberal use of benzodiazepines and antipsychotic medications. Shortly after rehab admission, he developed increasing bilateral lower extremity tone ultimately manifesting as dystonia. Follow up MRI (Figure 1) demonstrated stable findings of diffuse cortical infarction in the bilateral frontal, parietal, and occipital lobes as well as the bilateral basal ganglia. MRI of the total spine, CSF studies, and infectious workup were unremarkable. The patient was started on baclofen, gabapentin, and clonidine, but developed rhabdomyolysis, acute kidney injury, and fever, prompting transfer to the ICU for aggressive medication titration and monitoring. The patient required aggressive IV hydration, sedation with IV lorazepam, and PEG tube was placed for nutrition. The patient's dystonia slowly improved with further medication titration and supportive care. He was able to return to the rehab floor where he made significant progress with therapy but remained mod-max assist for ADLs and mod assist for ambulation with a rolling walker at the time of inpatient rehabilitation discharge.

Early Presentation of Status Dystonicus in a Child with Hypoxic Brain Injury: A Case Report.

Hardgrove, W., Ference E., Westerkam, D., Tsai T. Carolinas Rehabilitation, Charlotte, NC

Discussion

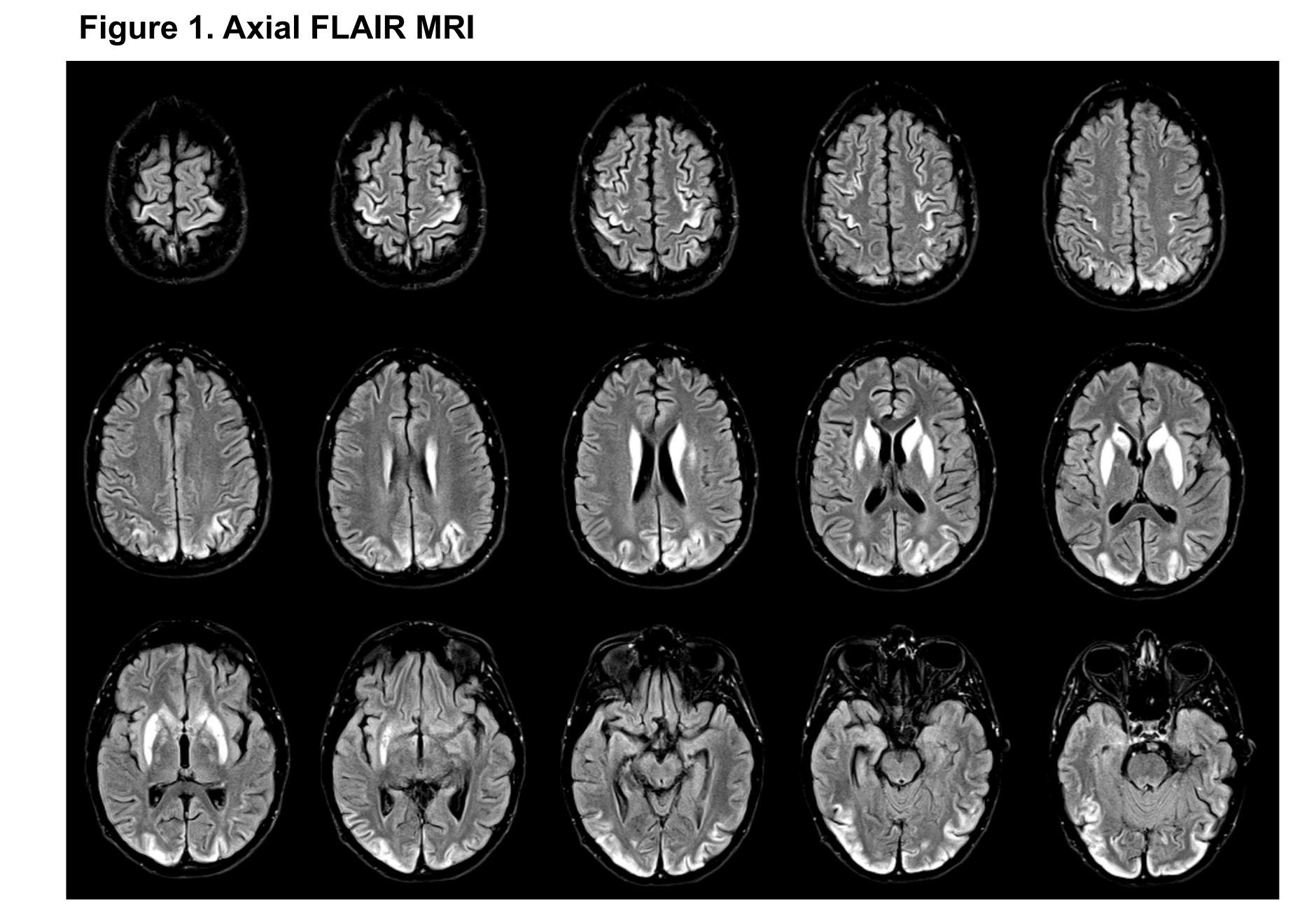


Figure 1. Axial FLAIR demonstrating widespread hyperintensities in the bilateral frontal, parietal, and occipital lobes and intense bright signal in the bilateral basal ganglia consistent with severe hypoxic injury.

Figure 2. Presentation Timeline

- Day 0
 - Status asthmaticus, intubated on scene and taken to OSH ICU
 - - Admitted to inpatient rehabilitation
 - Baclofen, gabapentin, and Clonidine initiated and titrated for worsening tone
- Day
- Transferred to ICU for monitoring in status dystonicus.
- Ativan titrated, baclofen and clonidine weaned. No new findings on work up.
- 25
 - Returned to inpatient rehabilitation
 - Clonidine was eventually discontinued 2/2 bradycardia.
- Day
- Discharged home on Ativan, baclofen, and gabapentin.
- Mod-Max assist for ADLs and ambulation with a rolling walker

Conclusions

- Dystonic storm should be considered as a possible diagnosis in any pediatric patient with worsening dystonia.
- With a mortality rate of approximately 10%, a high clinical index of suspension is required for an early diagnosis of dystonic storm.
- In severe refractory cases, supportive care should be conducted in the ICU for proper monitoring.
- Treatment focuses on temporizing measures with sedative hypnotics, GABA agonists, anticholinergics, antipsychotics, and catecholamine depleters (tetrabenazine).

References

- Allen NM, Lin JP, Lynch T, King MD. Status dystonicus: a practice guide. Dev Med Child Neurol. 2014 Feb;56(2):105-12. doi: 10.1111/dmcn.12339. Epub 2013 Dec 4. PMID: 24304390.
- 2. Fasano A, Ricciardi L, Bentivoglio AR, Canavese C, Zorzi G, Petrovic I, Kresojevic N, Kostic VS, Svetel M, Kovacs N, Balas I, Roubertie A, Mishra D, Mariotti P, Temudo T, Nardocci N. Status dystonicus: predictors of outcome and progression patterns of underlying disease. Mov Disord. 2012;27:783–8
- 3. Termsarasab, P., Frucht, S.J. Dystonic storm: a practical clinical and video review. J Clin Mov Disord 4, 10 (2017).

Contact Info

Wesley.Hardgrove@atriumhealth.org
Tobias.Tsai@atriumhealth.org

This case was reviewed by the Atrium Health Institutional Review Board and was found to be exempt. We wish to thank the Department of Physical Medicine and Rehabilitation at Carolinas Medical Center for supporting this project and funding our conference attendance.