Area Postrema Syndrome: A Case That Mimics Simple Infection

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Case Presentation

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Disclosure: None

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History and Presentation

- A 9-year old girl with no known medical illness presented with **2 weeks** history of **vomiting, hiccups** and intensifying respiratory distress with low grade temperature.
- She has been treated by general practitioner for gastritis, urinary tract infection and pneumonia.
- She eventually required intubation and ventilation due to **bulbar palsy** and **poor respiratory effort** despite with full consciousness and not requiring sedation while on ventilator.

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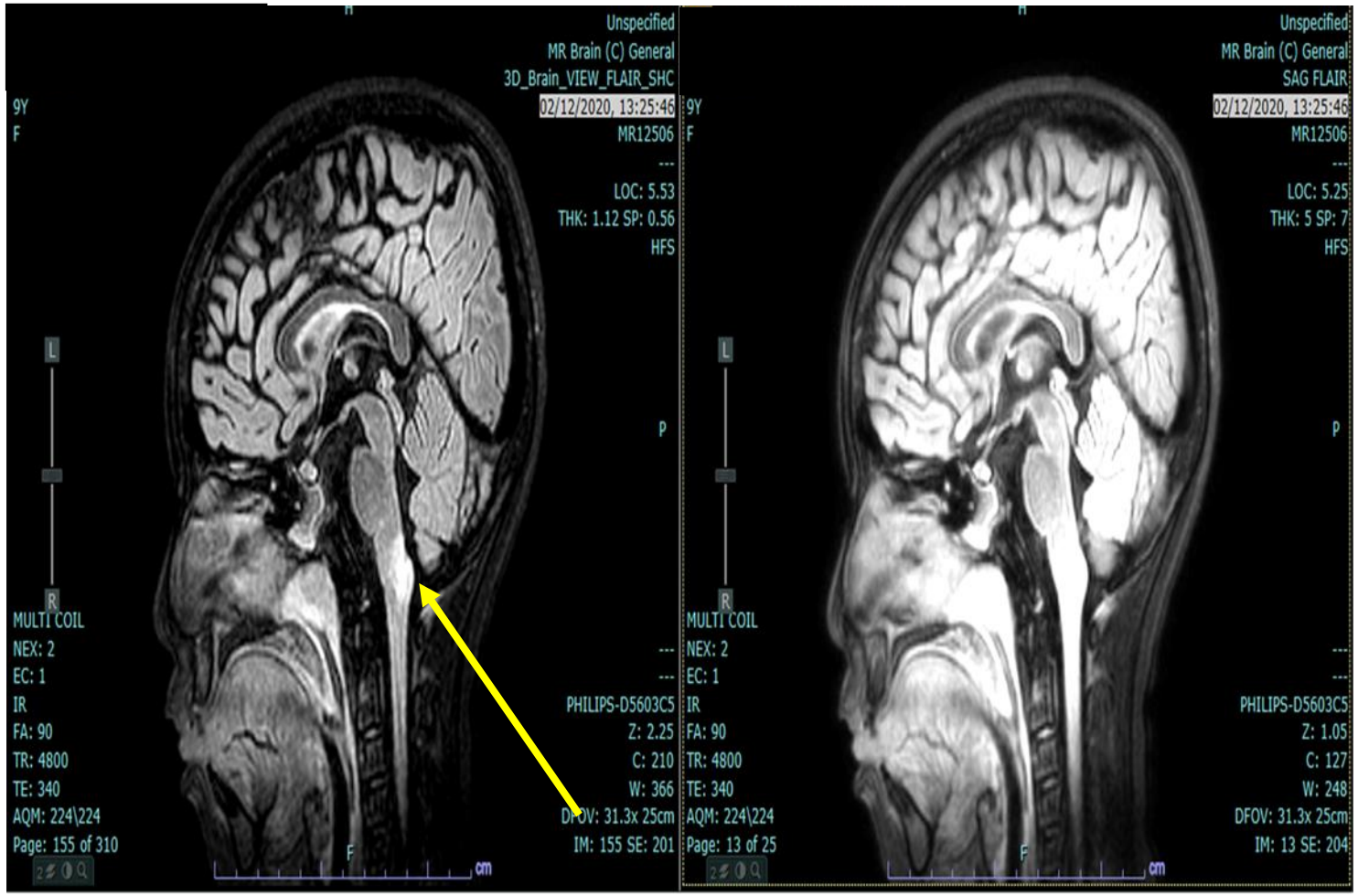
Neuroimaging

Hyperintensity signal over dorsal region of medulla oblongata (area postrema)



Serological Investigations

- FBC: Hb 14, WBC 9.3 (**Neut 81.4%,** Lymp 17%)
- Mycoplasma titre **positive 1:160**
- Serum amylase 1682 , urine diasterase 2248
- Aquaporin-4 receptor antibody **positive**
- Myelin oligodendrocyte glycoprotein (MOG) protein **negative**

Treatment and Management

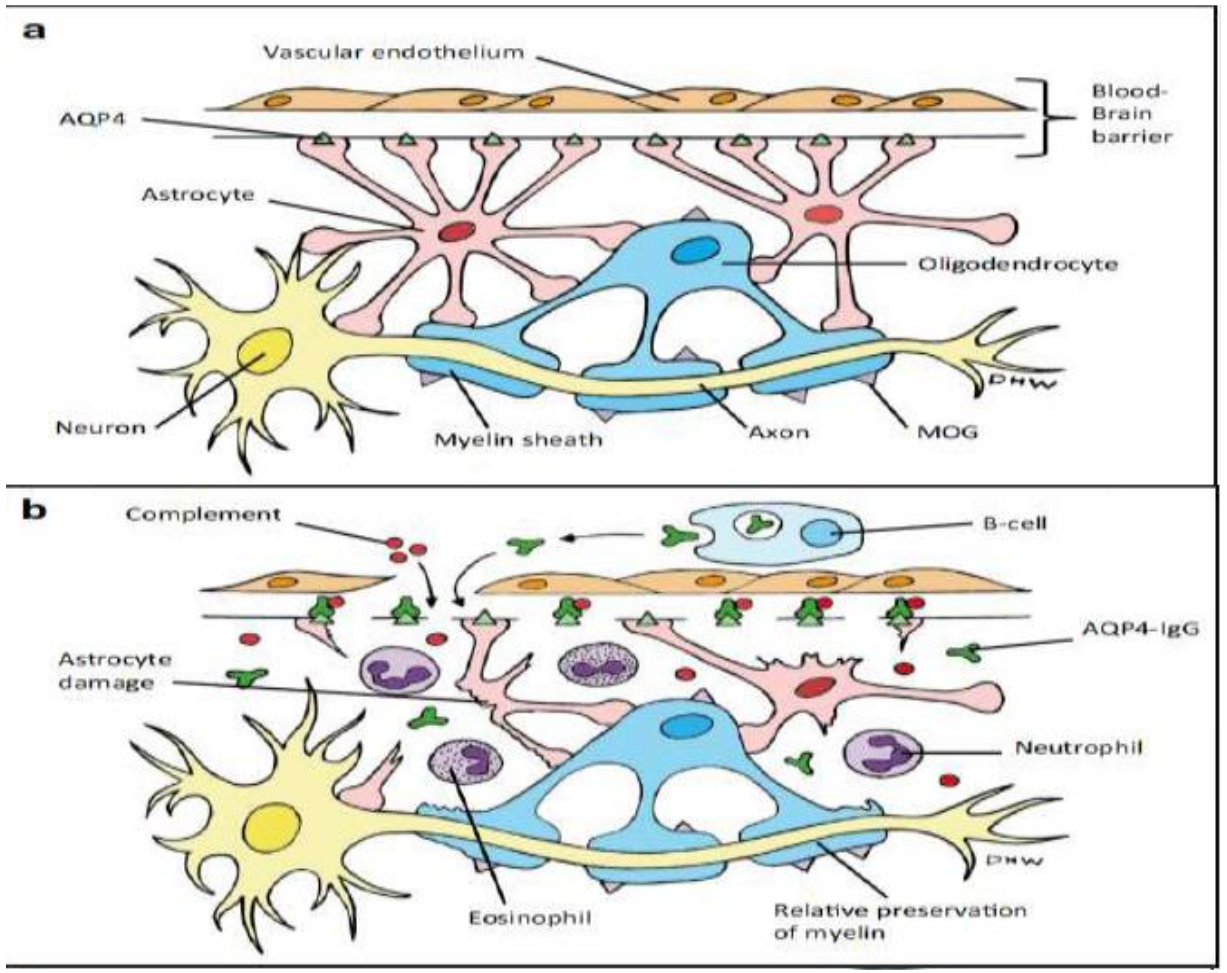
- She was started on IVIg 2g/kg over 2 days
- This was followed by high dose IV methylprednisolone 30mg/kg/day for 5 days (with tapering off dose orally)
- Poor remission / progression hence proceed with plasma exchange procedure
- There were planned for further immunotherapies if plasma exchange not effective

Acute Life Threatening Events / Complications

- She had acute life threatening events within 30 mins of plasma exchange twice that required CPR
- Since then she has poor GCS recovery, developed severe renal injury and liver derangement and diabetes insipidus
- Echocardiography showed significant grade 3 HOCM LVOT obstruction on ventricle contraction
- EEG finding showed generalised attenuated cerebral activity
- CT brain showed generalised oedema

Area Postrema Syndrome (APS)

- APS are associated with loss of AQP4 immunoreactivity and with inflammation
- Misdiagnosis is common and many patients underwent extensive workup.
- Shosha et al presented APS diagnostic criteria and phenotypic assessment (nausea, vomiting and hiccups) as well as severity scale for APS.
- Lack of response to symptomatic therapies may be a clue to APS.



1. The spectrum of neuromyelitis optica, LANCET Neurology 2007;6:805
2. Area postrema syndrome, Neurology 2018;91:1642-1651

Discussion and Learning Points

- **Prolonged** vomiting and hiccups are hints for atypical brainstem/medulla dysfunction
- Early immunotherapy can be effective
- Echo should be considered after ALTE
- The struggle in this case with complication making it difficult to manage the NMOSD/APS
- It is a **rare** case that can **mimic** other typical conditions in paediatrics
- Meticulous history is mandatory and early suspicion should be a sixth sense in recognising the syndrome
- Bradycardia and hypotension are indicators for obstructive element in cardiac disorder hence echo should be mandatory

References

- The spectrum of neuromyelitis optica, Lancet Neurology 2007;6:805
- International consensus diagnostic criteria for neuromyelitis optica spectrum disorders 2015, Neurology;85:177-189
- EFNS Guideline, European Journal of Neurology 2010:17”1019-1032
- Multisystem involvement in neuromyelitis optica. Ann Indian Acad Neurol 2015;18supplS1:56-8
- Updated diagnostic criteria for neuromyelitis optica spectrum disorder: Similar outcomes of previously separate cohorts. M McCreary et al 2018
- Palace et al 2019. Outcome prediction models in AQP4-IgG positive neuromyelitis optica spectrum disorders