A Case of a Splitting Headache: Paraneoplastic Rhombencephalitis

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Introduction
- Paraneoplastic neurologic syndromes (PNS)
  - Overall: no true incidence
  - Classic Syndrome: incidence of 0.86 per 100,000 cases
- Paraneoplastic Rhombencephalitis
  - Subset of Paraneoplastic Encephalitis (30%)
  - Inflammation of hindbrain (brainstem and cerebellum)
- We present a case of a female who acutely developed paraneoplastic rhombencephalitis

Case Presentation
- 67 year old female with metastatic small cell lung cancer to the bone, liver, and brain
- Due to poor prognosis of patient, family decided to place patient on comfort cares
- Developed status epilepticus with minimal response to IVIG and solumedrol treatment started
- Paraneoplastic panel of the CSF fluid
- Lumbar puncture
- Initial supportive treatment with antiemetics and fluids
- CBC, CMP, and UA unremarkable
- Unremarkable neuro exam. Gait not formerly assessed
- Physical exam: acutely ill female. Slurred speech noted.
- Hemodynamically stable
- MRI two weeks prior showed no metastatic disease
- Overall: no true incidence
- Inflammation of hindbrain (brainstem and cerebellum)
- • Cancer Dx within past 5 years of Neuro sx

Discussion
- Pathophysiology
  - Well Characterized Onconeuro antibodies:
    - anti-Hu, Yo, CV2, Ri, Ma2, or amphyphian
      - This case = Anticholinergic receptor ganglionic neuronal Ab
  - Overall poor prognosis and acutely aggressive
    - No clear guidelines on treatment
    - Tumor directed treatment
    - Immunomodulation
      - Antibody removal therapy – questionable results in literature
    - 1 cell mediated therapy – better success

2004 Definitions of PNS
- Classic Syndromes
  - Subacute cerebellar Degeneration
  - Limic encephalitis
  - Encephalomyelitis
  - Opsoclonus,
  - Myoclonus
  - Cancer Dx within past 5 years of Neuro sx
- Non-Classic Syndrome
  - Resolves/Improves after cancer Tx, w/o immunotherapy
  - Cancer Dx AND paraneoplastic Abs within past 5 years of Neuro sx

References
- Graus et al. (2004). Recommended diagnostic criteria for paraneoplastic neurological syndromes. doi.org/10.1136/jnnp.2003.034447