Introduction

- Bickerstaff's brainstem encephalitis (BBE) is a rare post-infectious neurological and immunological syndrome characterized by:
  - ophthalmoplegia;
  - ataxia;
  - impaired consciousness;
  - hyperreflexia; and/or
  - presence of pathological reflexes.
- Annual incidence of BBE: 0.078 per 100,000 habitants.
- Male-to-female ratio of 1.3:1 and a mean age of 39 years.
- This paper presents the first documented case of BBE associated with *Burkholderia cepacia* septicemia as to its clinical manifestations, laboratory features, diagnostic criteria, and management.

Case Presentation


- 3 weeks prior to admission
  - Productive cough, fever, body malaise
  - No headache, vomiting, or neck stiffness.
  - Admitted for community-acquired pneumonia
  - Treated with piperacillin-tazobactam IV
  - Condition improved thus opted to go home against medical advice
- 1 day prior to admission
  - Slurred speech
  - Facial asymmetry
  - Diplopia
  - Gait disturbance
  - Nausea and vomiting
- Upon admission: neurologic findings included
  - o altered mental status
  - o bilateral convergent strabismus
  - o facial asymmetry
  - o dysarthria
  - o ataxia
  - o hyperreflexia.
- On the 3rd hospital day:
  Ventilatory failure ensued which prompted respiratory support.

Diagnostics:

- Sputum & blood cultures: (+) for *B. cepacia* as isolate.
- Crania CT scan: no evidence of infarct, hemorrhage or mass noted.
- Cranial magnetic resonance imaging (MRI): Specifically high T2W/FLAIR signal intensities involving the right side of midbrain, pons, and medulla (see Figure 1).
- Cerebrospinal fluid (CSF) analysis: Elevated protein level of 75 mg/dL; white blood count of 0-2 cells/hpf, indicating albuminocytological dissociation.
- Work up for other causes of CNS infections revealed negative.

Treatment:

- Meropenem + methylprednisolone IV 1 gm/day for 3 days
  - Improvement of symptoms was noted

Discussion

- The basis for diagnosing BBE includes:
  - Main Features
    - Progressive symmetric ophthalmoplegia and ataxia
    - Either consciousness disturbance or pyramidal signs
    - Motor strength with 5 or 4
  - Supporting Features
    - History of infectious symptoms within 4 weeks of onset
    - CSF albuminocytological dissociation
    - Presence of anti-GQ1b IgG antibody
    - Not done
  - The above clinical features together with the abnormal cranial MRI findings are consistent with BBE. However, testing for anti-GQ1b antibody was not performed since it was not available in the local setting. Yet, it should be noted that that negative antibody testing does not preclude the diagnosis. Odaka *et al* in 2003 reported that the positive rate of the said antibody is only 66%.
  - In our patient who is diabetic and likely to be immunocompromised, this must have favored the growth of *B. cepacia* as documented by positive sputum and blood cultures.

Conclusion

- Bickerstaff brainstem encephalitis should be considered in patients presenting with impaired consciousness, ophthalmoplegia, ataxia, and respiratory difficulty.
- This will ensure prompt diagnosis and appropriate therapeutic management since this is a relatively fatal illness if left undetected.

References