Bickerstaff's Brainstem Encephalitis

This disease is notifiable in the UK, see NOIDs article for more details.

Bickerstaff's brainstem encephalitis (BBE) is an immune disorder of unknown aetiology. It is characterised by:

- Acute, progressive cranial nerve dysfunction.
- Associated cerebellar ataxia.
- Coma.

There is a clinical continuum between BBE and Fisher's syndrome.[1]

Bickerstaff reported eight patients who, in addition to acute ophthalmoplegia (diplopia) and ataxia, showed drowsiness, extensor plantar responses or hemisensory loss.[2]

**Epidemiology**

- It is very rare and mostly reported in adults; however, cases affecting children have also been reported.[3]
- Very often it follows an illness, and an association with certain infections, including cytomegalovirus, *Campylobacter jejuni*, typhoid fever and *Mycoplasma pneumoniae*, has been documented.[4, 5, 6]

**Presentation**

- Acute diplopia.
- Ataxia.
- Pyramidal tract paralysis.
- Disturbance of consciousness.
- Headache is common.
- Progressive, symmetrical ophthalmoplegia, ataxia and either disturbance of consciousness or hyperreflexia.
- Facial palsy, extensor plantar reflex, pupillary abnormality, nystagmus and bulbar palsy.
- It may result in apnoea and a reversible brain death picture.

**Differential diagnosis**[7]

- Multiple sclerosis.
- Behçet’s disease.
- Lyme disease.
- Sarcoidosis.
- Whipple’s disease.
- Listeria rhombencephalitis.
- Vasculitis due to systemic lupus erythematosus (SLE).
- Acute disseminated encephalomyelitis.

**Investigations**

- One review of 62 patients found positive serum anti-GQ1b immunoglobulin G (IgG) antibody in 66%, and brain abnormality on MRI scan in 30% of patients.[4, 8]
- The presence of anti-GQ1b antibodies and an abnormal brain MRI scan can help to support its diagnosis but absence of anti-GQ1b antibodies and a normal MRI scan result do not exclude the diagnosis, which remains based on clinical criteria and exclusion of other aetiologies.[8]
- Electrodiagnostic study results suggested peripheral motor axonal degeneration.

**Associated diseases**

- A large number of patients have associated Guillain-Barré syndrome, suggesting that the two disorders are closely related.[8]
- Miller Fisher’s syndrome (ophthalmoplegia, ataxia and absent reflexes).[10]

**Management**

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Success has been achieved with treatment with steroids plus double filtration plasmapheresis. It has also been achieved with immunoglobulin therapy. However, there are no randomised controlled trials of immunomodulatory therapy.

Prognosis

Although the initial presentation is severe, there is usually a good outcome with complete resolution. Cases of recurrent BBE have been reported.

Further reading & references


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