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.
- Progressive multifocal leukoencephalopathy.
- 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.\[9\]
- 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**
Success has been achieved with treatment with steroids plus double filtration plasmapheresis.\[11\] It has also been achieved with immunoglobulin therapy.\[12\] However, there are no randomised controlled trials of immunomodulatory therapy.\[13\]

Prognosis

Although the initial presentation is severe, there is usually a good outcome with complete resolution.\[14\] Cases of recurrent BBE have been reported.\[9\]

Further reading & references


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