Bickerstaff's Brainstem Encephalitis

This disease is notifiable in the UK, see [1]

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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