

ACUTE DEMYELINATION

Introduction

These disorders consist of monophasic and polyphasic (recurrent) diseases with acquired immune injury to the white matter in the central nervous system.

Optic neuritis

- acute loss of vision (decreased visual acuity) of one or both eyes
- often associated with pain on eye movements and colour desaturation
- A relative afferent pupillary defect is present
- CT/MRI may show swelling and abnormal signal of optic nerves.

Acute transverse myelitis

- spinal cord dysfunction, with motor weakness, numbness of both legs and/or arms, often associated with urinary retention
- maximal deficits occurring between 4 hours to 21 days after symptom onset
- MRI may demonstrate swelling +/- abnormal signal in the spinal cord

Acute Disseminated Encephalomyelitis (ADEM)

- acute neurological deficits, often multifocal (weakness, numbness, ataxia) with at least 2 of the following:
 - (1) prodromal illness in the last 28 days
 - (2) fever
 - (3) neck stiffness
 - (4) headache
 - (5) seizures
 - (6) altered level of consciousness or behaviour
- MRI shows multiple areas of abnormal signal in the white matter

Table 1. ADEM: Common differential diagnoses

<i>CNS infection</i>
bacterial, tuberculous meningitis
Herpes simplex encephalitis
<i>Demyelination of the peripheral nervous system</i>
Guillain Barré syndrome
Chronic inflammatory demyelinating polyneuropathy (CIDP)
<i>Acute stroke</i>
<i>Mitochondrial disorders</i>

Other Investigations (as needed)

- cerebrospinal fluid - FEME, cultures, oligoclonal banding, Herpes virus PCR (optional: lactate, viral studies)
- infection screen - virology, mycoplasma, etc.
- vasculitis screen (ESR, C3, C4, antinuclear factor)
- evoked potentials - visual, auditory and somatosensory

Treatment

Supportive measures

- vital sign monitoring, maintain blood pressure
- assisted ventilation for "cerebral / airway protection"
- anticonvulsants for seizures
- antibiotics, Acyclovir for CNS infections if febrile, awaiting cultures, PCR result

Definitive immunotherapy

- IV Methylprednisolone 20 - 30 mg/kg/day (max 1 gm) daily for 3 to 5 days then oral Prednisolone 1 mg/kg/day (max 60 mg) daily to complete 2 weeks.
- Severe episodes of demyelination that respond to initial therapy may benefit from a longer course, tapering over 4 to 6 weeks
- If no response, consider: IV Immunoglobulins 2 gm/kg over 2 - 5 days (or referral to a paediatric neurologist)

If recurring demyelinating episodes, consider referral to a paediatric neurologist