CASE CONFFRENCE

Presenter : R2周光緯
Supervisor : VS 吳柏衡

2011.10.19

Discussion

Neuromuscular weakness & Guillain–Barré syndrome

Table 166-1 Differentiating Central from Peripheral Nervous System Disorders

	Central	Peripheral
History	Cognitive changes	Weakness confined to one limb
	Sudden weakness	Weakness with pain associated
	Nausea, vomiting	Posture- or movement-dependent pain
	Headache	Weakness after prolonged period in one position
Physical examination		
Reflexes	Brisk reflexes (hyperreflexia)	Hypoactive reflexes
	Babinski sign	Areflexia
	Hoffman sign	
Motor	Asymmetric weakness of ipsilateral upper and lower extremity	Symmetric proximal weakness
	Facial droop	
	Slurred speech	
Sensory	Asymmetric sensory loss in ipsilateral upper and lower extremity	Reproduction of symptoms with movement (compressive neuropathy)
		All sensory modalities involved
Coordination	Discoordination without weakness	Loss of proprioception

GBS

- Acute immune-mediated polyneuropathy
- AIDP is the most common form
- Usually progress over a period about 2 weeks
- Worst at 4th week since initial symptoms – 90 %
- If S/S persist more than 8 weeks → CIDP

Pathogenesis

- An immune response to a preceding infection that cross-reacts with peripheral nerve components
- Target : myelin or the axon of peripheral nerve, resulting in demyelinating and axonal forms of GBS

Cause

- Campylobacter jejuni infection
- Cytomegalovirus
- Epstein-Barr virus
- Human immunodeficiency virus (HIV)
- Another triggering event
  - Immunization
  - Surgery
  - Trauma
  - Bone-marrow transplantation

Clinical Feature

- Progressive, fairly symmetric muscle weakness
- Depressed or absent DTR
- Onset : days to a week
- Severity : mild difficulty with walking to respiratory paralysis

Con't

- Weakness start from arm & face – 10 %
- Respiratory failure – 10~30 %
- Facial weakness & oropharyngeal weakness – 50 %
- Oculomotor weakness – 15 %
- Paraesthesia – 80 %

CSF study

- Albuminocytologic dissociation
  - Elevated CSF protein
  - Normal CSF WBC
- 80 ~ 90 % 1 week after the onset of symptoms
- If WBC increases (> 10/mm^3), check for concurrent HIV infection

Neurophysiologic Study

- Nerve conduction studies (NCS)
  - Absent responses or slowed conduction velocities
- Needle electromyography (EMG)
  - Reduced recruitment
- Demyelination at the level of the nerve roots

Miller Fisher syndrome (MFS)

- GBS variant
- Ophthalmoplegia with ataxia
- Areflexia
- Diagnosis : serum IgG Ab to GQ1b

Diagnostic Criteria

- National Institute of Neurological Disorders and Stroke (NINDS)
- Required features :
  1. Progressive weakness of more than one limb
  2. Areflexia

- Supportive features :
  1. Progression of symptoms over days to four weeks
  2. Relative symmetry
  3. Mild sensory symptoms or signs
  4. Cranial nerve involvement, especially bilateral facial nerve weakness
  5. Recovery starting two to four weeks after progression halts
  6. Autonomic dysfunction
  7. No fever at the onset
  8. Elevated protein in CSF with a cell count <10 mm3
  9. Electrodiagnostic abnormalities consistent with GBS

Treatment

- Supportive care
- Evaluation of respiratory function
- Intubation and ICU admission as indicated
- IVIG 2 g/kg over 2 days
- Plasmaphoresis
- Steroid → no benefit and may be harmful
- Consult neurologist for admission

Table 166-3 Managing Respiratory Failure in Guillain-Barré Syndrome
Indications for intubation
Vital capacity <15 mL/kg
PaO_2 <70 mm Hg on room air
Bulbar dysfunction (difficulty with breathing, swallowing, or speech)
Aspiration
Indications for admission to intensive care unit
Autonomic dysfunction (**Table 166-2**)
Bulbar dysfunction
Initial vital capacity <20 mL/kg
Initial negative inspiratory force <–30 cm of water
Decrease of >30% of vital capacity or negative inspiratory force
Inability to ambulate
Treatment with plasmapheresis

DDx

- Chronic inflammatory demyelinating polyneuropathy
  - > 8 weeks
- Acute polyneuropathies
- Spinal cord disorders
- Neuromuscular junction disorders
- Muscle disorders