NEUROMUSCULAR DISEASE AND ANESTHESIA

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Charcot-Marie-Tooth Disease (CMT)

- Most common inherited cause of chronic motor and sensory PERIPHERAL neuropathy, leading to distal muscle atrophy

- Slow and PROGRESSIVE degeneration/demyelination of peripheral nerves and roots
  - Motor and sensory loss of distal portion of lower then upper extremities

- Progressive distal skeletal muscle weakness and wasting
  - loss of tendon reflexes

- Generally manifests in teenage years
CMT

• Affects 1 in 2,500 people

• Seven types with multiple subtypes

• CMT 1 and 2 are most common

• Classically, neuropathy restricted to lower legs:
  • Foot deformities (foot drop)
  • Reduced deep tendon reflexes
  • Peroneal muscle atrophy (peroneal muscle affected early)
  • Frequent trips and falls
CMT

• Rarely affects muscles of respiration, however it’s difficult to assess

• May have no symptoms despite abnormal PFT’s (restrictive lung pattern)

• Proximal muscle weakness in arms may be predictive of respiratory muscle weakness
Anesthesia Implications: CMT

• Chart, Chart, Chart!
  • Pre-op AND post-op neuromuscular assessment

• TIVA and volatiles have been used safely

• Epidural, spinal and combined spinal/epidural techniques have been used successfully

• Response to non-depolarizing muscle relaxants can be unpredictable: exacerbated or attenuated. Varies from patient to patient
Anesthesia Implications: CMT

• Evidence base is limited and controversy exists regarding muscle relaxants use

• Decreased number of acetylcholine receptors
  • Decrease dose of non-depolarizing muscle relaxants
  • Completely antagonize non-depolarizing relaxants
  • Some case studies have shown no prolongation of relaxant with atracurium and mivacurum
  • Prudent to stay away from succinylcholine
Anesthesia Implications: CMT

- Risk of Malignant Hyperthermia has been speculated but NEVER ESTABLISHED
  - Assess personal and familial hx of MH
Anesthesia Implications: CMT

- Restrictive Lung Impairment is associated with:
  - Phrenic nerve dysfunction
  - Diaphragm dysfunction
  - Thoracic cage abnormalities

- Central Sleep Apnea may be associated with diaphragm dysfunction and hypercapnia

- OSA has been reported as possibly due to pharyngeal neuropathy

- Consider post-op BiPAP or CPAP if pt does have associated pulmonary disease
Anesthesia Implications: CMT

• Autonomic denervation is common and should be assessed:
  • Orthostatic hypotension
  • Heart rate variability
  • Resting tachycardia
  • Nocturnal HTN

• Assessment of comorbidities such as diabetes since this can lead to further deterioration in neuropathy
Anesthesia Implications: CMT

• Case Studies
  • LMA
  • Thoracotomy and use of vecuronium and sugammadex

• OB Study
  • Higher occurrence of presentation anomalies
  • 2x the rate of PPH and operative delivery
  • Forcep use 3x as often
  • Majority of CMT cesareans were emergent
OB Anesthesia Implications: CMT

- Limited research regarding use of regional anesthesia in CMT pt’s
- Opinions on regional anesthesia are often contradictory and based on theory rather than documented practice
- Developing consensus that regional anesthesia is safe alternative to general anesthesia
OB Anesthesia Implications: CMT

• Literature review:
  • 8 reports of spinal/eidural
  • Half were parturients
  • 6 out of 8 had NO untoward affects
  • Zero reported affects beyond discharge
OB Anesthesia Implications: CMT

• DELAYED RESOLUTION OF SENSORY BLOCK following epidural anesthesia is a consistent sequela in these patients

• Neurologic complications following labor occur in ABSENCE of anesthesia and are 5 TIMES MORE COMMON after childbirth itself independent of regional blockade!
OB Anesthesia Implications: CMT

- Document, document, document!

- Peroneal muscle atrophy may worsen with pregnancy
  - ? Fluid retention/edema around nerve ?

- Assess respiratory function
  - Decreased respiratory reserve, residual volume, FRC, total lung capacity
Transverse Myelitis

- Acute inflammation of the spinal cord, usually involving a limited segment
- Results in sensory, motor, and autonomic dysfunction
- Clinical signs are due to the involvement of the ascending and descending tracts in the transverse plane of the spinal cord
- Often damages the myelin
- Incidence of 1-5 million/year
Transverse Myelitis

- Pain, muscle weakness, paralysis, sensory problems, or bladder and bowel dysfunction

- Most people with transverse myelitis recover at least partially. Those with severe attacks sometimes are left with major disabilities.

- S/S are acute (hours to days) or subacute (1-4 weeks)
Transverse Myelitis

• Initial Symptoms:
  • Localized lower back pain
  • Sudden Paresthesias in legs
  • Sensory loss
  • Paraparesis (partial paralysis of legs)
  • Paraparesis may progress to paralegia
  • Bladder and Bowel dysfunction is commons
  • Muscle spasms and general discomfort
  • Headache, fever, loss of appetite
  • Respiratory problems, depending on segment of spinal cord involved
Transverse Myelitis

• Four Classic Features
  1. Weakness of legs/arms
  2. Pain
  3. Sensory Alteration
  4. Bowel and Bladder dysfunction

• Depending on complete or partial transverse involvement of the cord, the distribution of signs and symptoms may be symmetric or asymmetric below the affected level
Transverse Myelitis

• Most common cause is autoimmune

• Other causes:
  • Viral and other infections
  • Multiple Sclerosis
  • Neuromyelitis Optica
  • Autoimmune d/o such as Lupus, Sjrogren’s, Sarcoidosis

• 15-36% are idiopathic
Transverse Myelitis

• Diagnosis by Exclusion
  • History and neuro exam
  • MRI to rule out structural lesion (tumor, stenosis, herniated disc, abscess etc.)
  • CSF examination to exclude infection and check for MS
  • Blood and serologic tests to rule out autoimmune or infectious etiology) (B12 deficiency, Lupus, HIV, NMO etc.)
Transverse Myelitis

• Treatment
  • Reduce spinal cord inflammation

  • Manage and alleviate symptoms (narcotics, NSAIDS, corticosteroids)

  • Plasma exchange and immunosuppressive medications in severe cases
Transverse Myelitis

• Prognosis
  • Recovery usually begins 2-12 weeks after onset of symptoms and may continue for 2 years, sometimes longer
  • If no improvement within 3-6 months, complete recovery unlikely
  • 1/3 fully recover, 1/3 fair recovery, 1/3 no recovery
  • Rapid onset of symptoms generally results in poorer recovery outcomes
Anesthesia Implications: TM

• Preop documentation:
  • Motor function
  • Sensory level
  • Autonomic dysfunction

• Question about drug therapy
  • Preoperative tests should include a complete blood count, liver and renal function tests, as the drug therapy can alter these tests
Anesthesia Implications: TM

• Choice of anesthesia technique remains debatable.....

• In the literature, transverse myelitis has been attributed to the use of spinal and epidural anaesthesia... BUT, so has general anesthesia!!

• NO CAUSAL RELATIONSHIP HAS BEEN ESTABLISHED
Anesthesia Implications: TM

• Performing regional anesthesia in pt’s with pre-existing CNS disorders has been debatable and should be based on the risks and potential benefits of each individual case

• Avoid succinylcholine

• Pt’s are extremely sensitive to non-depolarizing relaxants
Anesthesia Implications: TM

• Postoperative documentation of motor and sensory function is essential for these patients and they should be monitored for any haemodynamic alterations.

• In cases of progressive symptoms, a neurology consult prior to anesthesia is prudent. Can serve as a baseline should symptoms worsen after anesthesia.
OB Anesthesia Implications: TM

- Spinal and epidural anesthesia have been used without incidence

- Same preop assessment/workup as non-OB pt

- Risk of general anesthesia and autonomic dysreflexia well outweigh the minimal potential risk that epidural or spinal anesthesia may exacerbate TM or trigger inflammation
Transverse Myelitis

• Autonomic Dysreflexia
  • Paroxysmal HTN, bradycardia
  • Lesion above T6, although can occur with a lesion as low as T10
  • Acute reaction of the autonomic nervous system to overstimulation