Bahan Ajar 9

SIRINGOMIELIA

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Pendahuluan

- Syringomyelia → gangguan degeneratif, kronik progresif → gejala awal timbul pada usia dewasa awal (25 - 40 tahun).
- Kasus ini sangat jarang ditemukan
- Insiden laki-laki = perempuan
- Gambaran klinis → bervariasi, tergantung arah pelebaran *syrinx* ke arah transversal atau longitudinal. Pelebaran → biasanya ke anterior dari kanalis spinalis

Definisi

- Syringomyelia (Yunani → syrinx artinya pipa atau tube) → kelainan degeneratif progresif yang kronik atau kelainan perkembangan medulla spinalis dengan karakteristik klinis :
  - Painless wealness
  - Brachial amyotrophy
  - Segmental sensory loss of dissociated type (loss of thermal and painful sensation dgn sebagian taktil, posisi sendi, dan getaran).

Etiologi

Tipe mayor syringomyelia berdasarkan etiologi & ada tidaknya hubungan dgn kanalis sentralis.

- *Communicating syringomyelia* :
  - dilatasi kanalis spinalis → bersifat primer → dihubungkan dgn abnormalitas dari foramen magnum seperti *Chiari malformation* tipe 1 atau *basilar arachnoiditis*
  - Ada hubungan langsung antara *syrinx* dgn sistem ventrikular
- *Non-communicating syringomyeli* :
  - kista terbentuk pada substansi dari medula spinalis
  - tidak berhubungan langsung dgn kanalis sentralis atau spatium subarachnoid.

Patogenesis

- **Teori hidrodinamik Gardner** → *"water hammer"-like* transmisi pulsatile tekanan CSF mll sambungan antar ventrikel 4 dgn canalis centralis tulang belakang → Blockade ini berawal dari foramen Magendie.

- **Teori William** → perkembangan siring, khususnya dgn Chiari malformasi → mengikuti perbedaan tekanan antara intracranial dgn spinal yg disebabkan o/ aksi seperti katup di foramen magnum.

Kriteria Bernett

Type	Classification
Type I	Syringomyelia with obstruction of the foramen magnum and dilatation of the central canal (developmental type) A. With type I Chiari malformation B. With other obstructive lesions of the foramen magnum
Type II	Syringomyelia without obstruction of the foramen magnum (idiopathic developmental type)
Type III	Syringomyelia with other disease of the spinal cord (acquired types) A. Spinal cord tumors (usually intramedullary, especially hemangioblastoma) B. Traumatic myeolopathy C. Spinal arachnoiditis and pachymeningitis D. Secondary myelomalacia from cord compression (tumor, spondylosis), infarction, hematomyelia
Type IV	Pure hydromyelia (developmental dilatation of the central canal), with or without hydrocephalus.

Gejala Klinis

Symptoms due to loss funtion	Symptoms due to irritation
Motor symptoms	
Paresis Paralysis	Myoclonia Fasciculations Spasticity
Sensory symptoms	
Hypoaesthesia Hypoalgesia Anosmia Amaurosis Deafness	Paraesthesia Dysaesthesia Allodynia Hyperalgesia Pain Photopsia Tinnitus
Vasodilations Hypo/anhydrosis Loss of pilorection	Vasoconstriction Hyperhyrosis Pilorection
Autonomic symptoms	

Penatalaksanaan

Type	Mode of action	Drug	Side effect
Anticonvulsants	Inhibits opening neuronal voltage –dependent channels (calcium channels, sodium channel) andGABA receptor.	Carbamazepine Gabapentin Pregabalin Topiramate	Hepatotoxicity, drowsiness, fatigue, ataxia, vertigo, gastrointestinal discomfort headache, blurred vision.
Antidepressants	Inhibits the re-uptake of the neurotransmitters norepinephrine and serotonin by neurons.	Amitriptyline Duloxetine Venlafaxine	Mouth dryness, intense sedation, fatigue, diminished libido, weight loss, nausea, insomnia, headache.
Local anaesthetics	Act mainly by inhibiting sodium influx through sodium-specific ion channels in the neuronal cell membrane.	Lidocaine Mexiletine	Dizzine, arrhythmia.
Analgesics	Act through specific receptors, particularly μ receptors distributed throughout the central and peripheral nervous system blocking them.	Tramadol Dextropropoxyphene Buprenorphine Morphine Oxycodone Fentanyl Methadone	Nausea, vomiting, sweating, dizzines, mouth dryness, sedation, vertigo.

Penatalaksanaan

Terapi Pembedahan

- Dekompresi occipital dan cervical
- Laminektomi dan syringotomi
- Shunting ventrikuloperitoneal

Prognosis

- bergantung penyakit dasarnya, besarnya disfungsi neurologis, & perluasan syrinx.
- Beberapa studi menunjukkan pasien meninggal rata-rata diusia 47 tahun → Kemajuan teknologi + teknik pembedahan→ RESIKO rendah

Edukasi

- Hindari → latihan berisiko tinggi, seperti : berlari, melompat → ketidakstabilan servikal.
- Hindari aktivitas yg disertai maneuver Valsava.

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