Surgical treatment in epilepsy related to vascular diseases and abnormalities

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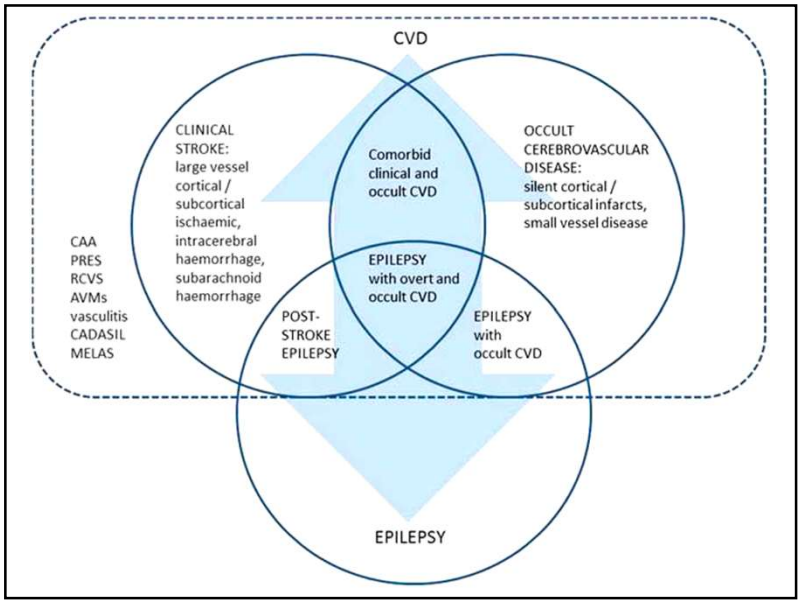
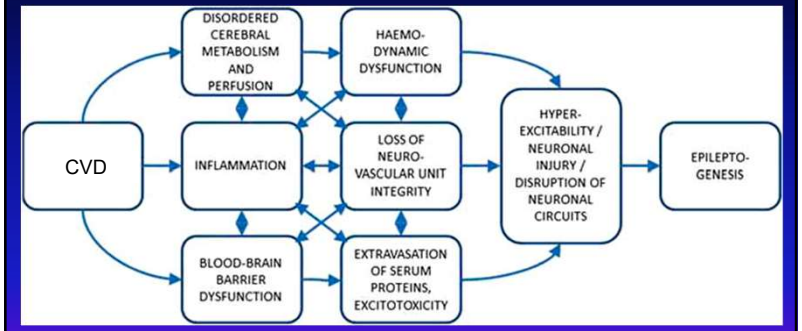


Overview

- Cerebrovascular diseases (CVD) and epilepsy
- Cerebral cavernous malformation (CCM)
- Sturge-Weber syndrome (SWS)
- Arteriovenous malformation (AVM)
- Post-stroke epilepsy

Pathophysiology



Cerebral cavernous malformation (CCM)

AKA "cavernoma" or "cavernous angioma"

Vascular malformation

Multilobulated appearance

Endothelium-lined caverns without mature vessel walls

Dynamic nature: progressive growth, repeated hemorrhages

Most CCMs (48%) are asymptomatic

CCM-related epilepsy: risk factors

Location

- Supratentorial: seizures in 50-63% of patients
- Infratentorial: seizure in 0-18% of patients

Cortical involvement

- Cortical lesion: 57-70% associated with epilepsy
- Deeper lesion: 14-20% associated with epilepsy

Mesial temporal

- Mesial temporal: seizures in 8/9 (88.9%)
- Neocortical temporal: seizures in 41/72 (56.9%)

CCM-related epilepsy: definition

Definite CCM-related epilepsy

- At least 1 CCM + evidence of SOZ in area of in the immediate vicinity of the CCM

Probable CCM-related epilepsy

- At least 1 CCM + evidence that the epilepsy is focal and arises from the same hemisphere as CCM but not necessarily in its immediate vicinity + no evidence of other causes for epilepsy

CCM unrelated to epilepsy

- At least 1 CCM + evidence that CCM and epilepsy are not causally related

CCM-related epilepsy: risk factors

Controversial risk factors

- Lobar localization
- Number of CCM
- Size of CCM and hemosiderin rim

Presurgical evaluation for CCM

5-year risk of seizure is low in CCM patients without seizures

- Incidental CCM 4%
- CCM presenting with ICH 6%
- Prophylactic AED is not recommended for CCM patients without seizures

Presurgical evaluation for CCM

CCM with single seizure

- Most doctors prefer AED > surgery

Early surgery

- High risk of bleeding
- Patients with poor drug compliance
- Patients with strong desire to eventually stop AED

Presurgical evaluation for CCM

CCM presenting with seizures

- 5-year risk of seizure is high 94%
- AED is recommended

Recommendations

- All CCM patients presenting with seizures should be transferred to epileptologists
- Semiology and EEG
- Classification: definite/probable/unrelated

Presurgical evaluation for CCM

CCM patients with seizure free on AED

- 47-60% of patients with newly diagnosed CCM
- Regular follow-up is recommended

Presurgical evaluation for CCM

CCM patients with persistent seizures

- Should NOT wait until fulfill the criteria of drug resistant epilepsy (DRE)
- Presurgical evaluation for epilepsy surgery is considered in cases with failure of single AED trial
- In CCM patients with rare seizures who have semiology, inter-ictal EEG concordant with location of CCM, patients may be referred to surgery (no need of ictal EEG)

Presurgical evaluation for CCM

Dual pathology (CCM + HS or other lesions)

- Most patients will not achieve seizure-free by removal of a single lesion
- Identify that which one is epileptogenic lesion
- In patients with mesial temporal CCM + ipsilateral HS, lesionectomy + amygdalohippocampectomy is recommended

Intraoperative electrocorticography (ECoG)

- Controversial in improving surgical outcome

Presurgical evaluation for CCM

CCM patients with multiple CCMs and persistent seizures

- Video-EEG is necessary
- Identify that which one of CCM corresponding to occurrence of seizure (epileptogenic CCM)
- Seizure outcome in patient with multiple CCMs and single epileptogenic CCM, is favorable

Surgery for CCM

Indications

- Epilepsy
- Bleeding
- Progressive growth with pressure effect

Surgery for CCM

Microsurgical resection

- Lesionectomy with removal of surrounding epileptogenic brain tissue (hemosiderin-stained brain tissue)

Neuronavigation-guided

Intraoperative mapping for CCM located in or near the eloquent cortex

- Awake surgery with electrocortical stimulation

Sturge-Weber syndrome

Pathophysiology of epilepsy in SWS

- Increased number of leptomeningeal vessels
- Atrophy and calcification of cerebral cortex, particularly around blood vessels
- Decreased number of cortical vessels
- Gliosis, neuronal loss and tissue loss
- Calcification, neuronal loss and gliosis are secondary to brain injury from venous stasis and impaired brain perfusion

Sturge-Weber syndrome

Vascular malformation

1. Skin: port-wine stain (capillary malformation)
2. Brain: leptomeningeal angioma
3. Eye: choroidal angioma

Leptomeningeal angioma

- Epilepsy: 75% within 1 year of age, 90% within 2 years of age
- Neurological impairment: intellectual disability, hemiparesis, visual impairment, severe migraine

Sturge-Weber syndrome

Epilepsy in SWS can be difficult to control

- Frequent clusters of seizure
- Episodes of status epilepticus

Seizure control results in the best chance of optimal cognitive and neurologic development

DRE in SWS

- Surgery should be considered especially when hemiparesis and visual deficits already occur

Sturge-Weber syndrome

Hemispherectomy is very successful (90%) in eliminating seizures in most cases and focal resection is less effective

Bilateral extensive SWS with severe brain involvement

- The highest risk of very poor neurologic and cognitive outcome
- Not good surgical candidates
- Hemispherectomy is recommended only in bilaterally SWS with very severe disabling seizures primarily coming from 1 hemisphere
- Surgery is considered palliative > curative

Brain AVM and epilepsy

Nowadays, endovascular treatment > surgery

Surgery for AVM with DRE

- Presurgical evaluation for identifying epileptic focus
- Resection of brain AVM and epileptogenic brain, gliotic brain and hemisiderin deposition
- Electrocortical stimulation and mapping in AVM close to the eloquent cortex

Brain AVM and epilepsy

DRE is rare in brain AVM

Pathogenesis of epilepsy in brain AVM

- Focal cerebral ischemia secondary to AV shunting
- Gliosis of surrounding brain
- Hemosiderin deposit
- Focal hemorrhage
- Secondary epileptogenesis in temporal lobe

Post-stroke epilepsy

Pathogenesis of epilepsy after ischemic stroke

- Increased intracellular Ca2+ and Na+ resulting in lower threshold for depolarization
- Glutamate excitotoxicity
- Hypoxia,
- Metabolic dysfunction
- Global hypoperfusion
- Hyperperfusion injury
- Gliosis

Post-stroke epilepsy

Pathogenesis of epilepsy after hemorrhagic stroke

- Irritation caused by products of blood metabolism
- Hemosiderin deposition
- Gliosis

Post-stroke epilepsy: risk factors

After hemorrhagic stroke

- Subarachnoid hemorrhage
- Middle cerebral artery aneurysm
- Intraparenchymal hemorrhage
- Presence of structural brain lesion
- EEG abnormalities
- Partial type seizure

Post-stroke epilepsy: risk factors

After ischemic stroke

- Severe initial neurological deficit
- Severe persistent disability
- Multiple infarctions
- Large infarction
- Cortical involvement
- Hippocampal involvement
- Embolic stroke

Post-stroke epilepsy

DRE is rare in post-stroke epilepsy

Surgical management

- Presurgical evaluation for epilepsy surgery
- Surgical options: focal resection to hemispherotomy
- Patients' age, underlying diseases and use of anticoagulant/antiplatelet are major limitations for surgery

Conclusions

Cerebral cavernous malformation

- Most common vascular lesion related to epilepsy
- No seizure, no AED
- Surgery in cases with epilepsy and failure of AED, poor drug compliance, desire to stop AED, high risk of bleeding
- Lesionectomy with removal of hemosiderin

Sturge-Weber syndrome

- Surgery is considered in cases with DRE + neurological deficit (motor and visual)
- Hemispherectomy renders the best result

Conclusions

Brain AVM and post-stroke epilepsy

- Rare DRE
- Surgery is considered in cases with DRE
- Surgery for brain AVM: resection of AVM, gliotic brain and hemosiderin
- Surgery for post-stroke epilepsy depends on extension of epileptic focus: focal resection, lobectomy, hemispherotomy