Identifying Cerebrovascular Disorders

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Objectives

Review different types of cerebrovascular disorders.

Briefly discuss etiology, clinical presentation, evaluation and management.

Share a few interesting cases
Disclosure

Neurology PI, Wingspan® Stent System Post Market Surveillance Study (WEAVES Study).

Scientific consultant at Stryker Neurovascular.
Stroke

A stroke occurs when a brain vessel occludes or ruptures.

**Ischemic stroke:**
- Cerebral infarction from lack of oxygen due to blockage of blood supply.

**Hemorrhagic stroke:**
- *Intracerebral hemorrhage*: bleeding directly into the brain from the rupture of an abnormal vessel.
- *Subarachnoid hemorrhage*: bleeding beneath the brain membrane (subarachnoid space) from a ruptured aneurysm.
Transient Ischemic Attack (TIA)

**Clinical definition:**
- Focal neurological symptoms or signs that last < 24 hours.
- 30% patients with symptoms lasting < 24 hours have an infarction on MRI.

**Tissue-based definition:**
- A transient episode of neurological dysfunction caused by focal ischemia without acute infarction.

**TIA vs Ischemic Stroke:**
- Distinction not important
- Same pathophysiology
- Same preventative therapy
Common Symptoms of TIA or Stroke

**Anterior circulation**
- Contralateral weakness
- Contralateral sensory deficit
- Contralateral neglect
- Aphasia (left hemisphere)

**Posterior circulation (5 “D”s)**
- Dizziness (vertigo)
- Double vision (diplopia)
- Dysarthria
- Dysphagia
- Disequilibrium (Ataxia)

**Lacunar infarct**
- Pure motor hemiparesis
- Pure sensory hemiparesis
- Ataxic hemiparesis
- Dysarthria-clumsy hand

Blurred vision
Nausea, vomiting, or headache
Major Risk Factors of Stroke

- Hypertension, diabetes, hypercholesterolemia
- Smoking, obesity
- Atrial fibrillation and mechanic valve
- Arterial dissection
- Cocaine or methamphetamine use
- Vasculitis, Moyamoya disease.
- Hypercoagulable state, cerebral venous thrombosis
- Advanced age
Extracranial and Intracranial Atherosclerotic stenosis

• Extracranial stenosis:
  • Carotid or vertebral arteries outside the skull.
  • Proximal internal carotid artery (ICA) stenosis is the most common cause of TIA or Stroke.

• Intracranial stenosis:
  • Account for 10-15% ischemic stroke in the U.S.
  • More common in Asian and Hispanic population
  • A high risk of recurrent or severe stroke.
    - Wang YJ et al; Stroke. 2014;45:663-669
Stroke Prevention for Extracranial and Intracranial Stenosis

1. Life style modification: quit smoking, eat healthy, and lose weight

2. Management of medical illness: HTN, DM and hyperlipidemia

3. Antiplatelet therapy: Aspirin, Plavix, or combination

4. Surgical intervention:
   - Carotid endarterectomy (CEA)
   - Carotid stenting
   - Intracranial stenting (high risk of complication)
Arterial Dissection: Carotid or Vertebral Artery

1. **Etiology**
   
a. Spontaneous
   
   • Develops in patient with Ehlers–Danlos syndrome type 4, Marfan syndrome, fibromuscular dysplasia or atherosclerosis.

   b. Traumatic
   
   • Occurs after car accident, fall, chiropractic manipulation, or hyperextension of the neck.

   C. Drug
   
   • Methamphetamine or cocaine use.

2. **Pathogenesis**
   
   • Begins as a tear in the inner lining of the artery wall.
   
   • Blood enters the dissected wall and forms a clot.
   
   • The clot may block blood flow or break off to cause a stroke.

3. **Treatment**
   
   • Antiplatelet therapy: Aspirin or Plavix.
   
   • Anticoagulation: Coumadin
Vertebral artery (VA) dissection

- Headache or neck pain following a minor head or neck injury.
- Ischemic stroke
  - Focal neurologic signs develop in 85% of patients in a few days,
  - Ipsilateral facial pain and numbness
  - **Dizziness** (vertigo), **Dysarthria**, **Diplopia**, **Dysphagia**, and **Disequilibrium**
  - Nausea, vomiting, hiccups, or unilateral hearing loss
  - Contralateral weakness or numbness
- Subarachnoid hemorrhage (SAH)
  - from intracranial vertebral artery dissection
  - Sudden onset severe headache or decreased level of consciousness
  - Treatment: coil embolization of the dissected artery
Cerebral Amyloid Angiopathy

- Common in elderly and patients with Alzheimer’s disease.
- Amyloid deposits in cortex vessels, causing cracking and rupture of vessel wall.
- Lobar or multi-focal hemorrhage.
- Some patients with diffuse microhemorrhages
Moyamoya Disease

- A rare, progressive cerebrovascular disorder caused by blocked arteries at the base of the brain.
- “Moyamoya” means “puff of smoke” in Japanese and describes the look of the tangle of tiny vessels formed to compensate for the blockage.
- More common in Asian.
- Primarily affects children.
- Initial symptom is often stroke, recurrent TIA, or seizure.
- Adults more often experience hemorrhagic stroke due to bleeding into the brain from the abnormal vessels.
Moyamoya Disease: Treatment

**Medical therapy:**
- Baby Aspirin
- Good hydration
- Headache and seizure management

**Angioplasty and stent:**
- Open narrowed artery, unproven efficacy

**Revascularization surgery:**
- Direct superficial temporal artery to middle cerebral artery (STA-MCA) bypass
- Indirect encephaloduroarteriosynangiosis (EDAS) bypass in children
Cerebral Vasculitis

Inflammation (swelling) of small, medium-size or large vessels in the brain.

**Etiology**
- Primary: no underlying cause
- Secondary:
  - Methamphetamine or cocaine use
  - Infection
  - Connective tissue diseases: systemic lupus erythematosus, rheumatoid arthritis
  - Systemic vasculitis: Wegener’s granulomatosis, polyarteritis nodosa, Behcet’s syndrome.

**Symptoms**
- Severe persistent headaches
- Fever, weight loss, muscles stiffness
- Double vision, partial vision loss or blind spots
- Forgetfulness or confusion
- TIA or strokes

**Treatment**
- High dose steroids
- Immunosuppressant: cyclophosphamide
**Arteriovenous Malformation (AVM)**

- A tangle of abnormal blood vessels connecting arteries and veins in the brain.
- Often located in cerebral hemispheres involving middle cerebral artery (MCA)
- Congenital and typically seen in patients 20-50 years old
- Present with hemorrhage, seizure, or focal deficit
- 1-4 % rate of hemorrhage per year, account for 6-13 % of ICH
Treatment of Arteriovenous Malformation (AVM)

- Medical therapy
  - Treatment of headache, HTN, and seizure
- Surgical resection
  - If the AVM has bled or is in an area that can easily be reached, surgical removal is the treatment of choice
- Endovascular embolization
  - Injects an embolizing agent to the feeding artery to reduce blood flow into the AVM.
  - Less invasive
- Stereotactic radiosurgery (gamma knife)
  - Directs radiation beams to damage AVM vessels and cause scarring.
  - Indicated for small AVMs
Dural Arteriovenous Fistula

An abnormal connection (fistula) between an artery and a vein within the dura (the fibrous cover of the brain).

Etiology
• Venous anomaly or thrombosis,
• Head trauma or infection

Clinical presentation
• Tinnitus (bruits) or headache
• Orbital symptoms (from carotid cavernous fistula)
• Cranial nerve palsies
• Focal neurological deficits from intracranial hemorrhage

Treatment
• Conservative
• Endovascular embolization to occlude the artery
• Surgical resection
• Stereotaxic radiosurgery

Carotid cavernous fistula
Cavernous Angioma  
(Cavernoma, or Cerebral Cavernous malformation (CCM)).

A collection of dilated blood vessels form a benign tumor.  

**Cause**  
- Sporadic, no family history, single lesion.  
- Familial due to CCM gene mutation, typically multiple lesions.

**Clinical presentation**  
- Most asymptomatic  
- Headache, seizure or focal neurological deficits

**Diagnosis**  
- MRI is the most sensitive test: showing small new or old hemorrhages as a rim around the cavernoma.

**Treatment**  
- Conservative: management of headache and seizure  
- Surgical resection: if hemorrhage or refractory seizure.
Hypercoagulable State

Abnormal coagulation that increases the risk of thrombosis.

**Cause:**

1). Inherited (congenital)
   - Homozygous factor V Leiden, prothrombin G20210A mutation,
   - Protein C, protein S, or antithrombin deficiency

2). Acquired
   - Antiphosphlipid syndrome: caused by autoimmune antibodies against lupus anticoagulants, cardiolipin, and β2 glycoprotein.
   - Hematologic conditions such as sick cell disease or thrombocytosis.
   - Estrogen

**Diagnosis:**

- Blood test of coag and thrombophilia panel
- Testing for protein C, protein S, and antithrombin deficiency 2-4 wks after anticoagulation (not useful in acute setting or on warfarin).
Cerebral Venous Thrombosis (CVT)

- Uncommon cause of stroke
- Good outcome from early treatment

**Etiology**

- Infection: sinusitis
- Trauma or surgery
- Hypercoagulable state

**Clinical presentation:**

- Headache
- Seizure
- Cerebral edema or infarction
- Intracerebral or intraventricular hemorrhage
Cerebral Venous Thrombosis (CVT)

**Treatment:**

1. Anticoagulation (iv heparin followed by Coumadin with INR 2-3).
   - For patient with provoked CVT, Coumadin 3-6 month
   - For unprovoked CVT, Coumadin 6-12 months.
   - Recurrent CVT or first CVT with hypercoagulable state, Coumadin indefinitely.
   - For women, full dose LMWH should be used during pregnancy and postpartum.

2. Thrombectomy for patient contraindicated for anticoagulation

3. Other therapy
   - Good hydration
   - Treatment of infection
   - Management of headache and seizure
Case 1

50 y/o gentleman with history of hypertension presented with sudden onset of headache, neck pain, and decreased consciousness.

CT head showed diffuse subarachnoid hemorrhage (A & B).

Fat-saturated T1 MRI (C & D) and MRA (E) showed intracranial vertebral artery dissection with intramural thrombus
Hospital Course

The distal L-VA dissection was treated with coil embolization. He improved significantly in 2 weeks and was discharged to ARU.
25 yo woman with history of migraines and chiropractic neck manipulation presented to outside hospital with neck pain, dizziness and unsteady gait.

Imaging studies showed L cerebellar infarct (A) and bilateral vertebral artery dissection at C₁-C₂ level (B).

Treated with ASA and Lipitor for stroke prevention.

One day later, she developed hearing loss from bilateral cerebellar infarcts (C&D).

Transferred to UCI Stroke Service on iv heparin.
Hospital Course

On arrival, cerebral angiogram confirmed bilateral vertebral artery dissection at $V_3$ segment (A and B). There was no evidence of vasculitis, atherosclerosis or fibromuscular dysplasia (C and D).

She was started on Coumadin for stroke prevention and discharged to ARU and then home in stable condition.
Day 15

Developed L sided weakness, vertigo, and diplopia. MRI showed new infarcts in b/l middle cerebellar peduncles and right pons.

Baby ASA was added to Coumadin therapy (INR 2.6-2.8).
"I think we should call the doctor. This doesn't look good!"
Locked-in from Acute Basilar Artery Occlusion

Aphasic and quadriplegic: NIHSS 23.
Emergently intubated for airway protection.
Stat CTA showed acute basilar artery occlusion (A).
After urgent reversal of coagulopathy (INR 2.6) with PCC, she underwent thrombectomy with excellent BA recanalization (C).
Hospital Course

Awake and moving all extremities post endovascular therapy
Discharge to ARU and then home 4 weeks later with Aspirin and Plavix
Excellent recovery and get married 2 months later.
Cerebrovascular disorders are very complex. Diagnosis is relatively easy due to advanced technologies. Treatment is constantly improving due to multidisciplinary collaboration. Academic Comprehensive Stroke Centers are the best place for patients with complex cerebrovascular disorders.
University of California Irvine Comprehensive Stroke Center

Stroke Neurologists

Neurointerventionalists

Neurointensivists

Cerebrovascular Surgeons

Program manager and assistant

Neurology Residents and Fellows
Neuro ICU/Step-down/Ward Nursing Staff
Emergency Medicine
Neuroradiology
Physical Therapy & Rehabilitation