7.5.2. Orbital Neoplasms and Malformations (II): Vascular Tumors (II)

Arteriovenous Fistula

introduction

- abnormal direct communication between an artery and a vein
- blood does not pass through an intervening capillary bed
- 2 forms of acquired arterial diversion that affect the cavernous sinus
  - direct carotid-cavernous fistulas
  - indirect (dural) carotid-cavernous fistula
- etiology
  - trauma
  - degeneration

direct carotid-cavernous fistulas

- high rate of blood flow
- connection between the internal carotid artery and the cavernous sinus
- etiology
  - trauma
    - causes a tear or hole in a branch artery of the internal carotid
  - iatrogenic
    - during neurosurgical or neuroradiologic procedures
- tortuous epibulbar vessels

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- clinical presentation
  - bruit
    - ± audible to the examiner and the patient
  - pulsatile proptosis
  - ischemic ocular damage
    - diversion of arterialized blood into the venous system
      - venous outflow obstruction
  - elevated IOP
  - choroidal effusions
  - blood in the Schlemm canal
  - nongranulomatous anterior uveitis
  - ocular motility abnormalities
    - congestion within the orbit
    - increased pressure in the cavernous sinus
      - compression of cranial nerves III, IV, or, most commonly, VI,
- CT scans
  - diffuse enlargement of some or all of extraocular muscles
  - characteristically enlarged superior ophthalmic vein

indirect (dural) carotid-cavernous fistulas

- lower rate of blood flow
- connection between meningeal branches of internal or external carotid artery and cavernous sinus
- degenerative process in older patients with
  - systemic hypertension
  - vascular disease
  - atherosclerosis
- clinical presentation
  - insidious onset
  - orbital congestion
  - proptosis
  - pain
  - arterialization of the conjunctival veins
    - chronic red eye
  - increased episcleral venous pressure
    - IOP elevation on the ipsilateral side
      - glaucomatous optic disc damage
  - mild

diagnostic workup

- magnetic resonance angiography (MRA)
  - not as sensitive as conventional angiography
- conventional angiography
  - gold standard
  - stroke is a rare but possible adverse effect

treatment

- endovascular treatment
  - coils

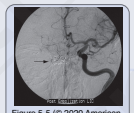

Figure 5-5 (© 2020 American Academy of Ophthalmology)

  - glue
- direct carotid-cavernous fistula
  - usually requires intervention
  - more common
    - transarterial approach
      - may require an orbitotomy to directly access the vein
      - transvenous approach by transcutaneous canalization of the superior ophthalmic vein
- indirect (dural) carotid-cavernous fistula
  - may initially be observed
    - small dural carotid-cavernous fistulas often close spontaneously
  - some investigators have recommended more aggressive management
    - higher risk for intracranial hemorrhage
      - transvenous access

Venous Malformation

- "orbital varices"
- vascular dysgenesis
  - low-flow
- clinical presentation
  - enophthalmos at rest
  - proptosis
    - increases when the patient's head is dependent or after a Valsalva maneuver
- imaging
  - contrast-enhanced rapid spiral CT during a Valsalva maneuver
    - characteristic enlargement of the engorged veins
    - phleboliths
- treatment
  - conservative
    - biopsy should be avoided because of the risk of hemorrhage
  - surgery
    - reserved for
      - significant pain
      - vision-threatening compressive optic neuropathy
    - complete surgical excision is difficult
      - lesions often intertwine with normal orbital structures
    - intra-operative embolization of the lesion may aid surgical removal
  - embolization with coils
    - to diminish symptoms

Arteriovenous Malformation (AVM)

- vascular dysgenesis
  - high-flow
  - abnormally formed anastomosing arteries and veins without an intervening capillary bed
- dilated corkscrew episcleral vessels
- management
  - arteriography
    - selective occlusion of the feeding vessels
      - followed by surgical excision
      - exsanguinating arterial hemorrhage may occur

Orbital Hemorrhage

- etiology
  - trauma
  - surgery
  - vascular malformations
  - sudden increase in venous pressure (eg, Valsalva maneuver)
    - rare
    - spontaneous orbital hemorrhage almost always occurs in the superior subperiosteal space
- treatment
  - observation
  - urgent drainage
    - if there is associated visual compromise

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