

Hemangiomas and Vascular Malformations

Pathogenesis · morphology · clinicopathologic interpretation

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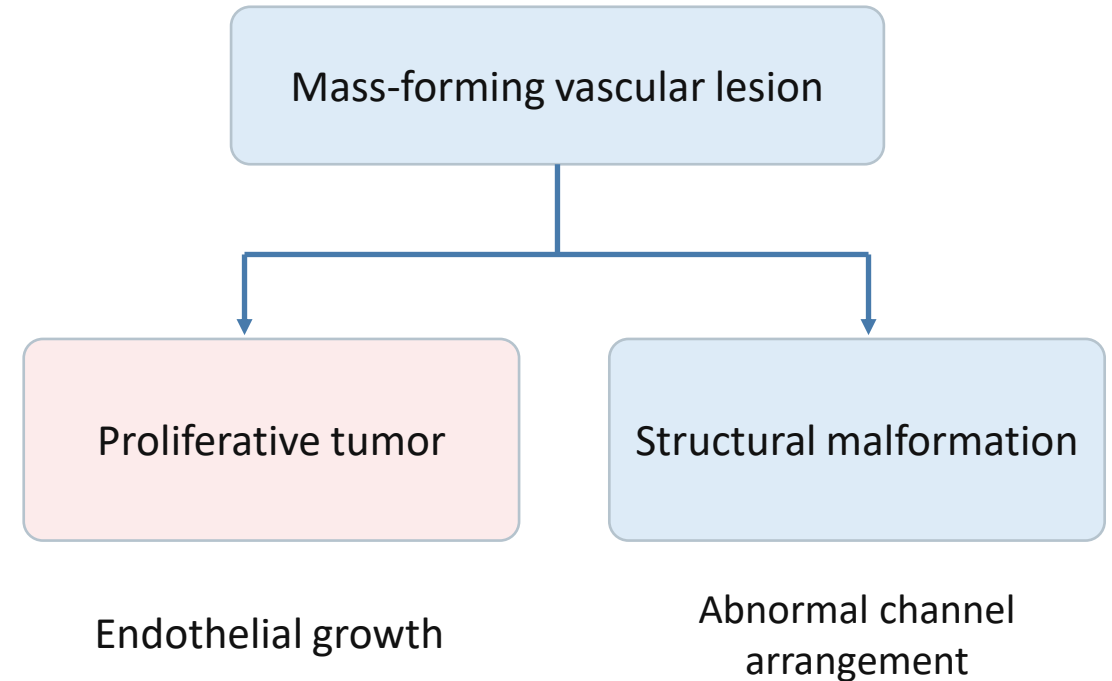
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Learning objectives

- Separate proliferative vascular tumors from structural malformations.
- Recognize capillary, juvenile, cavernous, and lobular capillary patterns.
- Explain clinically important ectasias and lymphatic lesions.
- Distinguish cerebral arteriovenous from cavernous malformations.

Two fundamentally different vascular lesions

- Vascular tumors form masses through endothelial proliferation.
- Vascular malformations comprise structurally abnormal vessels.
- Both may appear red-blue and mass-forming at the surface.
- Biologic category—not color alone—determines interpretation.

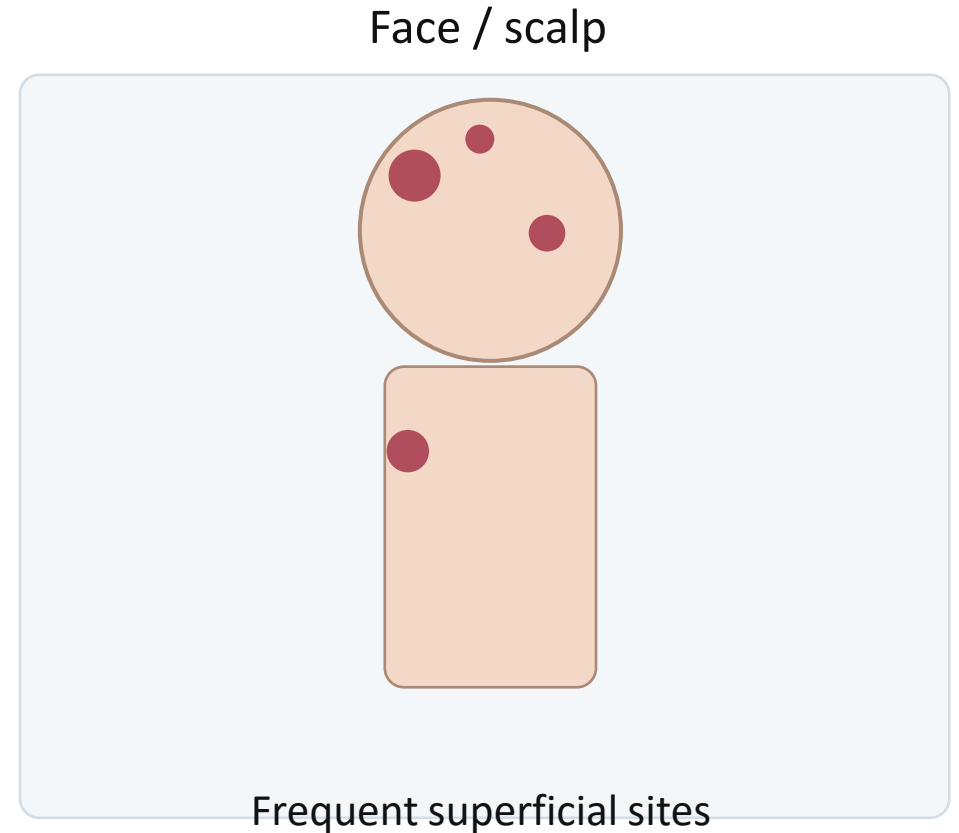


Why the word “hemangioma” is ambiguous

- In strict usage, hemangioma designates a benign vascular tumor.
- Historical usage also labels some malformations “hemangiomas.”
- Similar appearance and architecture encourage diagnostic overlap.
- Describe the lesion and its biologic behavior before naming it.

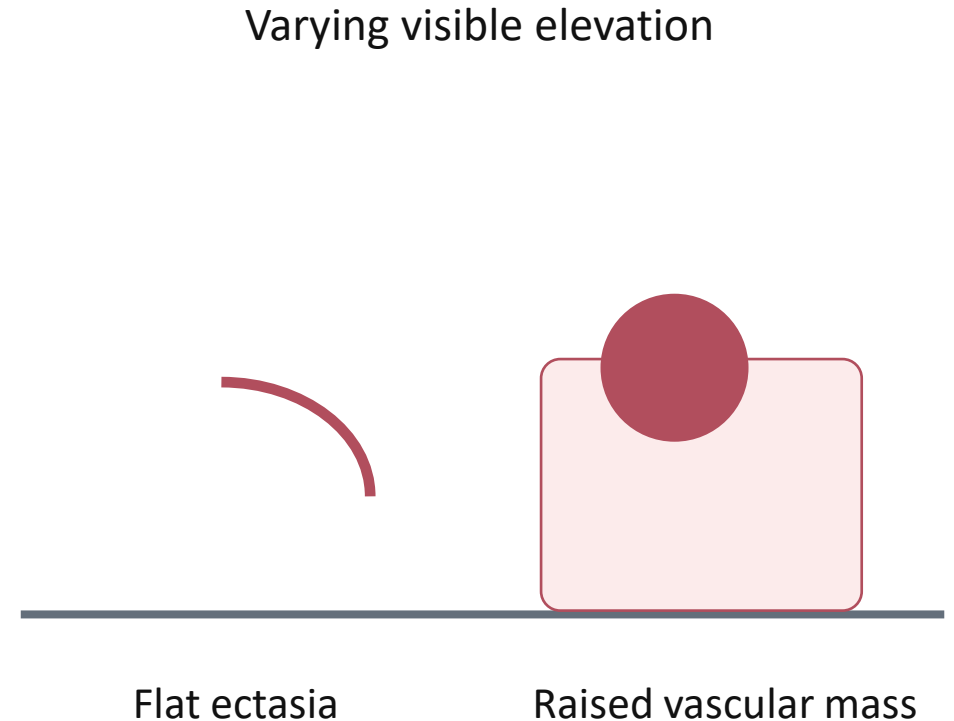
Pediatric vascular lesions: Typical topography

- These are the commonest mass-forming lesions of infancy.
- Skin is a frequent site, particularly the face and scalp.
- Visible lesions range from flat patches to elevated masses.
- Color is commonly red to blue, with irregular contours.



Surface appearance is a continuum

- A flat colored patch may represent ectatic vessels.
- An elevated mass may contain proliferating vascular channels.
- Superficial color does not reliably reveal deep extent.
- Depth and tissue relationships require separate consideration.



Natural history can distinguish entities

- Some childhood hemangiomas initially enlarge and later involute.
- Persistent port-wine stains behave differently from regressing lesions.
- Cavernous lesions generally do not regress.
- A time course adds information beyond a single examination.



Different lesion types follow different courses

Vascular tumor versus malformation

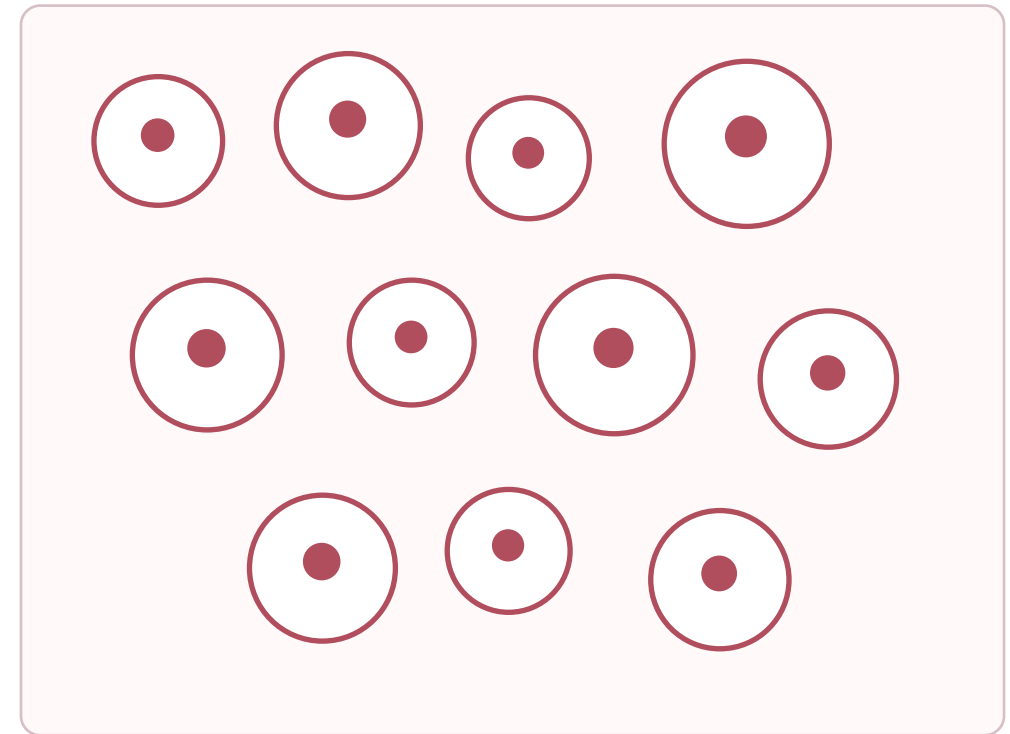
Feature	Proliferative tumor	Vascular malformation
Primary process	Vascular endothelial growth	Abnormal vessel architecture
Term “hemangioma”	Strictly appropriate for benign tumor	Sometimes applied historically
Presentation	May form raised mass	May be flat or mass-forming
Interpretation	Morphology + growth pattern	Morphology + vascular configuration

Why benign childhood masses still matter

- Many benign pediatric tumors are clinically inconsequential.
- Size and anatomic position can nonetheless cause major problems.
- A cosmetically conspicuous lesion may have functional consequences.
- Benign histology does not guarantee a harmless clinical course.

Histologic hallmarks of benign vascular proliferation

- Recognizable lumina contain erythrocytes in blood-vessel lesions.
- A single layer of bland endothelial cells lines the channels.
- Organization as vessels contrasts with solid atypical growth.
- The relevant diagnosis still depends on the specific architecture.



Small endothelial-lined blood spaces

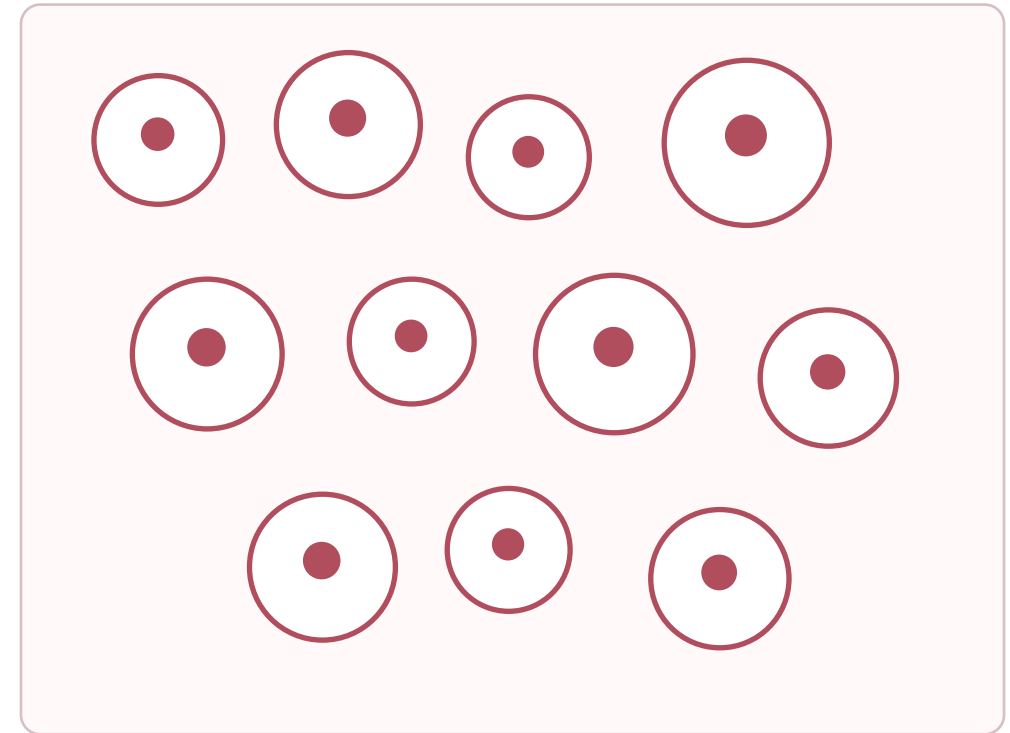
Schematic — not a histologic specimen

What raises concern for malignancy?

- Malignant vascular tumors are more solidly cellular.
- Cytologic atypia can accompany loss of well-formed lumina.
- Normal-looking blood-filled channels favor benign vascular growth.
- Ambiguous cases may require demonstration of endothelial lineage.

Capillary hemangioma: Defining architecture

- This is the most common hemangioma type.
- The lesion contains numerous small, thin-walled capillaries.
- Supporting stroma is relatively scant.
- The compact capillary network explains its often red appearance.



Small endothelial-lined blood spaces

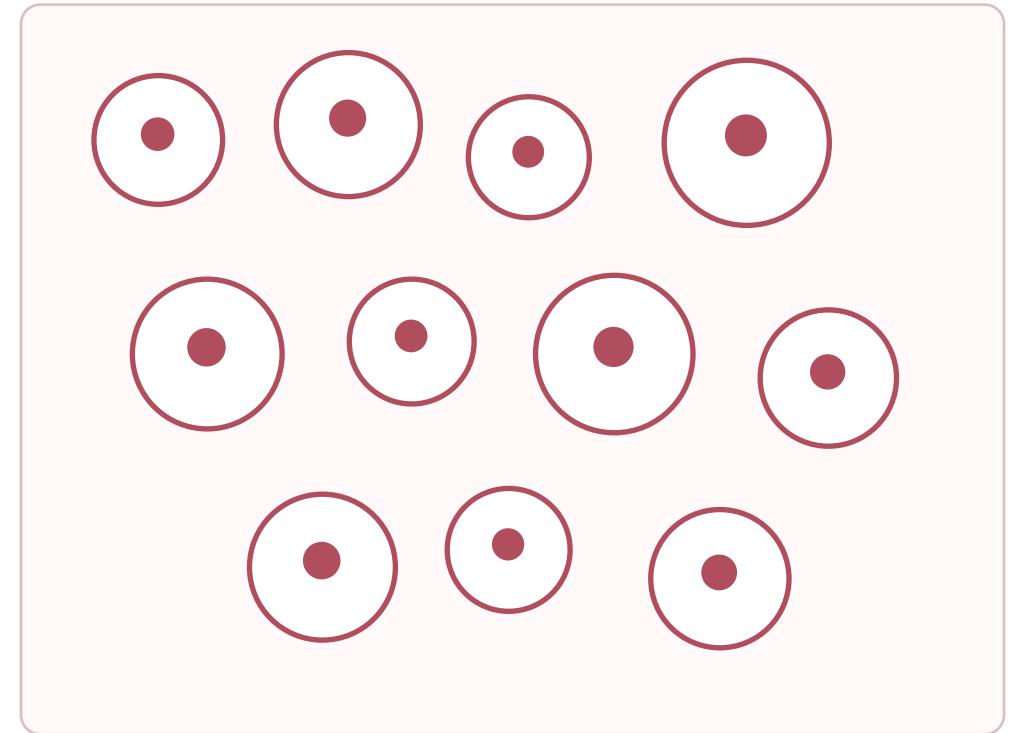
Schematic — not a histologic specimen

Capillary hemangioma: Site distribution

- Commonly affects the skin and subcutaneous tissues.
- Oral mucosa and lips are frequent superficial locations.
- It may also develop in liver, spleen, and kidney.
- Site alone does not establish a specific vascular subtype.

Microscopic reading: Identify the capillary units

- Look for multiple small lumina rather than cavernous spaces.
- Each lumen is bounded by a thin endothelial lining.
- Erythrocytes identify the channels as blood vessels.
- This is a native schematic; No patient histology is shown.



Small endothelial-lined blood spaces

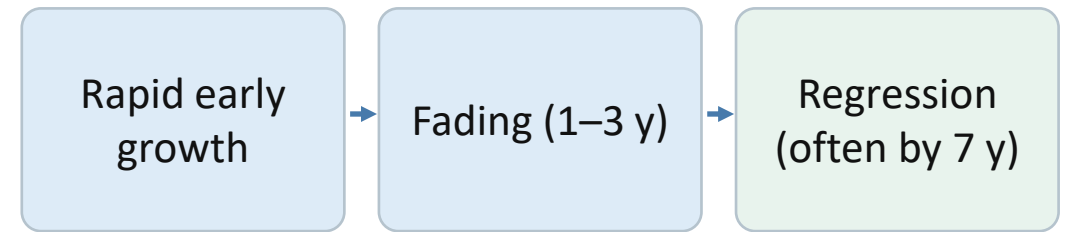
Schematic — not a histologic specimen

Juvenile hemangioma: Dynamic childhood lesion

- The “strawberry” variant commonly involves newborn skin.
- Lesions may be multiple.
- A rapid proliferative phase occurs over the first months.
- Regression follows in many cases rather than continuous expansion.

Juvenile hemangioma: Proliferative-to-involutional course

- Early months: Conspicuous enlargement can be rapid.
- Fading beginning around ages 1–3 years.
- Complete regression is reported by age 7 in most cases.
- These age landmarks describe a pattern, not an individual guarantee.



Illustrative course of juvenile hemangioma

Why a regressing lesion is not a port-wine stain

	Juvenile hemangioma	Port-wine stain
Early course	Proliferative growth	Flat vascular birthmark
Later course	Common spontaneous regression	Usually persists; may thicken
Biologic class	Benign vascular tumor	Vascular ectasia / malformation
Interpretive clue	Changing mass	Persistent patch

Multiplicity does not imply malignancy

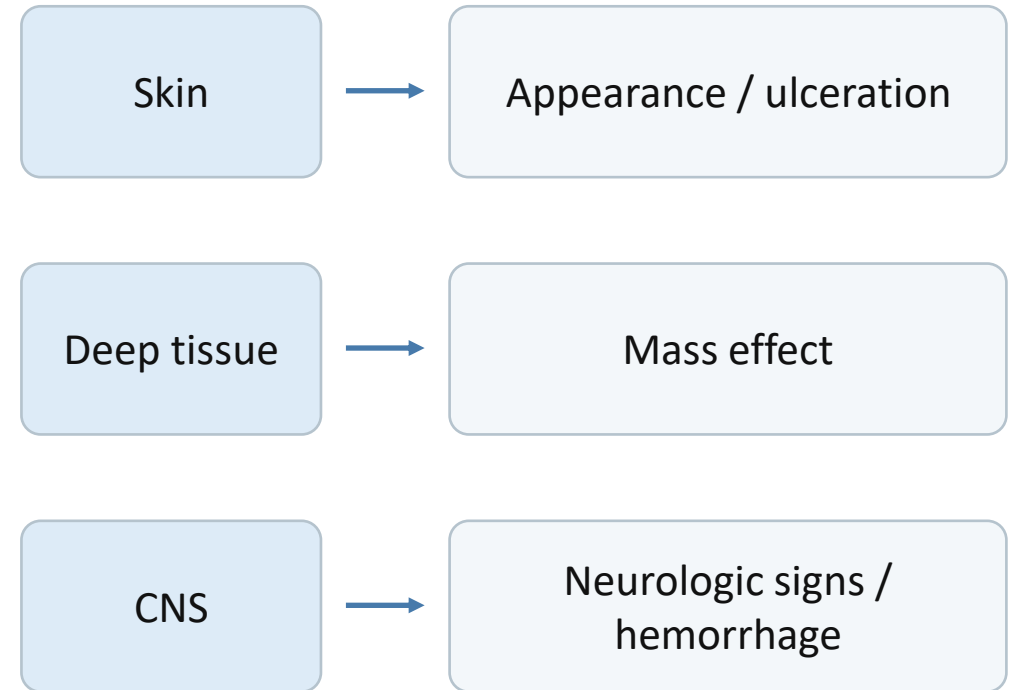
- Juvenile cutaneous hemangiomas may be multiple.
- Multifocality requires interpretation in anatomic context.
- The depth and extent of lesions determine clinical importance.
- Do not equate several benign vascular lesions with aggressive histology.

Hemangiomas may extend beyond the skin

- Internal hemangiomas are recognized in the same vascular spectrum.
- The liver accounts for a substantial share of internal lesions.
- Extensive involvement is sometimes termed angiomatosis.
- Specific complications depend on the involved organ.

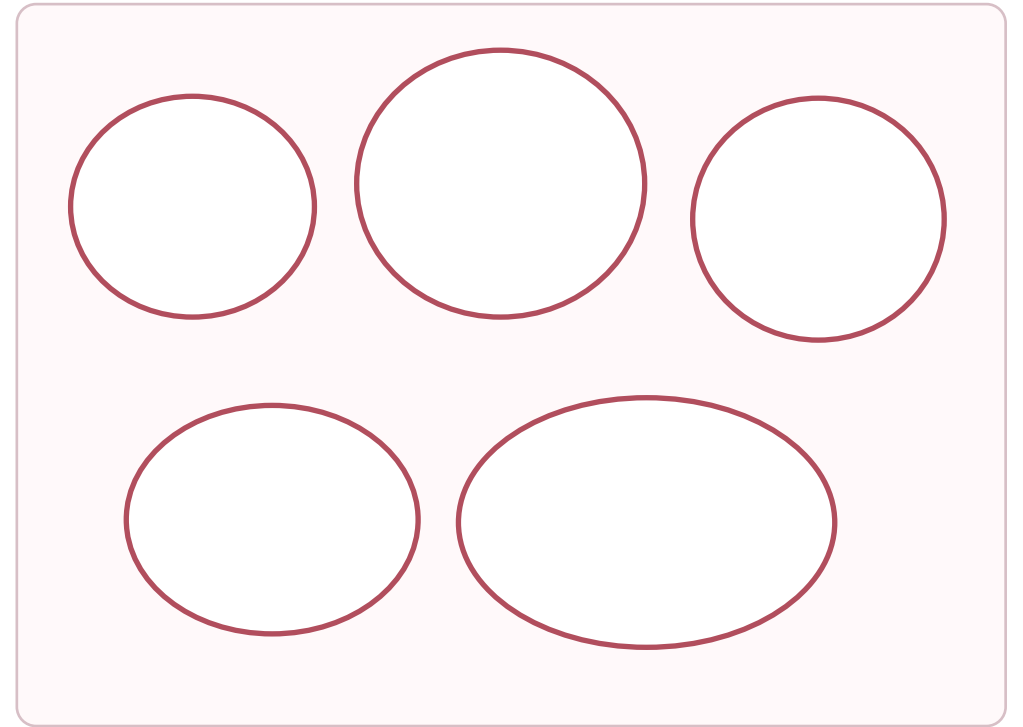
Location determines clinical significance

- Most superficial lesions are benign and primarily cosmetic.
- Ulceration or bleeding may occur with trauma.
- Deep lesions can affect nearby structures.
- CNS involvement has distinctive neurologic risks.



Cavernous hemangioma: Dilated vascular spaces

- Large, blood-filled channels dominate the architecture.
- Connective tissue separates the cavernous spaces.
- Cavernous spaces contrast with the fine capillary pattern.
- The description is morphologic; Terminology requires site context.



Large spaces separated by connective tissue

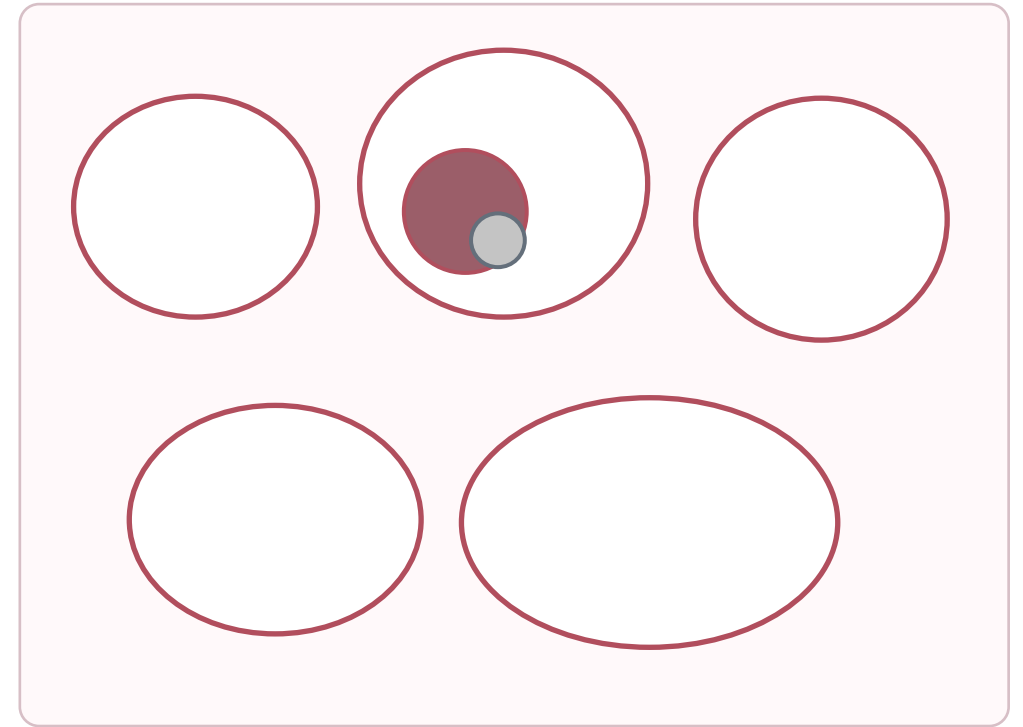
Schematic — not a histologic specimen

Cavernous hemangioma: Growth pattern

- A sharply delineated but unencapsulated lesion.
- Cavernous lesions can extend into deeper tissue.
- They are more infiltrative than capillary hemangiomas.
- Spontaneous regression is generally not expected.

Cavernous spaces favor stasis-related changes

- Dilated vascular spaces may develop intraluminal thrombosis.
- Organizing thrombi may undergo dystrophic calcification.
- These changes modify the appearance of the mass.
- Recognize secondary changes without inferring malignancy.



Thrombus and calcification can form within enlarged spaces

Schematic — not a histologic specimen

Cavernous hemangioma: Major clinical hazards

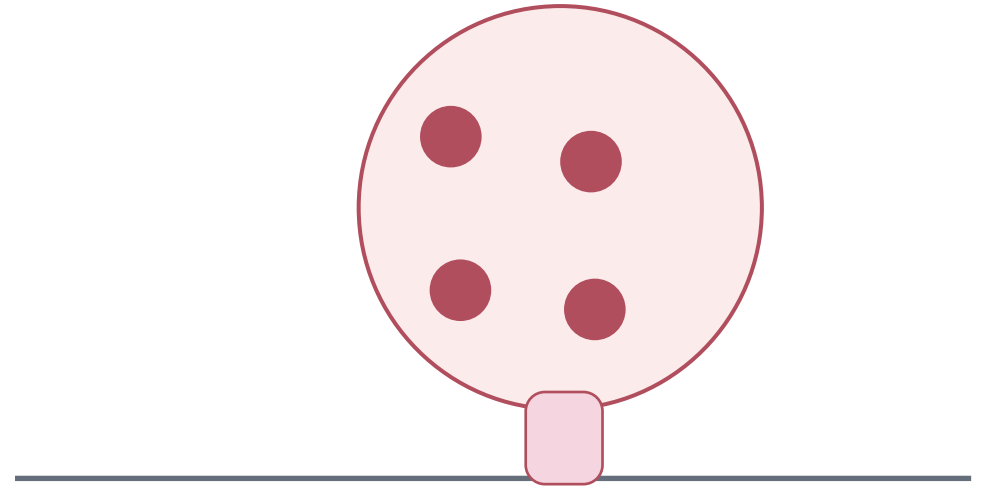
- Large lesions may produce local tissue compression.
- Surface lesions can ulcerate or bleed following trauma.
- Cerebral vascular lesions may bleed or cause mass effect.
- Clinical significance is strongly site- and size-dependent.

Capillary and cavernous hemangiomas

Attribute	Capillary	Cavernous
Dominant spaces	Small, thin-walled capillaries	Large, dilated blood-filled channels
Intervening stroma	Relatively scant	Connective tissue septa
Typical depth	Often superficial	Frequently deep
Regression	Some juvenile lesions involute	Generally not spontaneous
Secondary finding	Proliferative capillary pattern	Thrombosis / calcification possible

Pyogenic granuloma is a vascular proliferation

- Despite the name, this lesion is not a pyogenic infection.
- It with capillary hemangiomas.
- It grows rapidly as a red, often pedunculated nodule.
- Skin, gingiva, and oral mucosa are characteristic sites.



Friable pedunculated vascular nodule

Schematic — not a histologic specimen

Why pyogenic granulomas bleed easily

- The lesion is highly vascular and close to the surface.
- Surface ulceration is common.
- Minor mechanical contact may precipitate bleeding.
- A rapidly growing bleeding nodule warrants careful morphologic review.

Pyogenic granuloma: Associated settings

- Some lesions follow local trauma.
- A gingival lesion in pregnancy is termed granuloma gravidarum.
- Rapid growth can occur over only a few weeks.
- Context helps recognition but does not replace tissue diagnosis.

Pyogenic granuloma: Possible outcomes

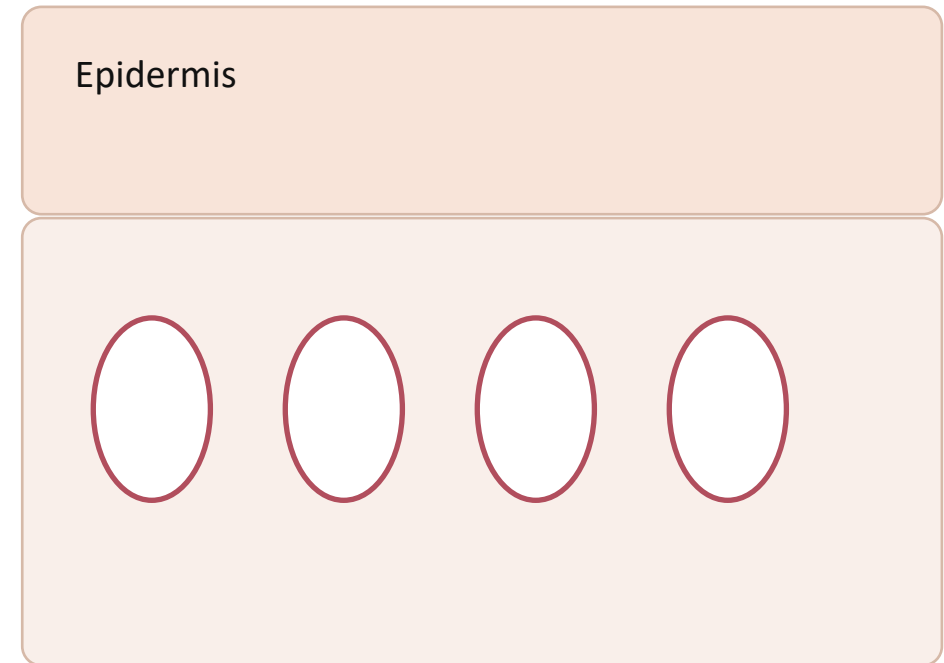
- Curettage and cautery are often effective.
- Some pregnancy-associated lesions regress after delivery.
- Other lesions may become fibrotic.
- Occasional cases require surgical excision.

Three patterns within the hemangioma family

Pattern	Defining feature	Characteristic course / symptom
Capillary	Fine vascular network	Often superficial
Juvenile / strawberry	Early proliferative cutaneous lesion	Common eventual involution
Cavernous	Broad, dilated vascular spaces	May persist and involve deep tissue
Pyogenic granuloma	Rapidly growing red pedunculated nodule	Bleeding and surface ulceration

Vascular ectasia means dilation, not neoplasia

- Ectasia signifies localized dilation of a tubular structure.
- Telangiectasia is permanent dilation of small preexisting vessels.
- The affected vessels may be capillaries, venules, or arterioles.
- Congenital and acquired telangiectasias are not true tumors.



Permanently dilated existing vessels

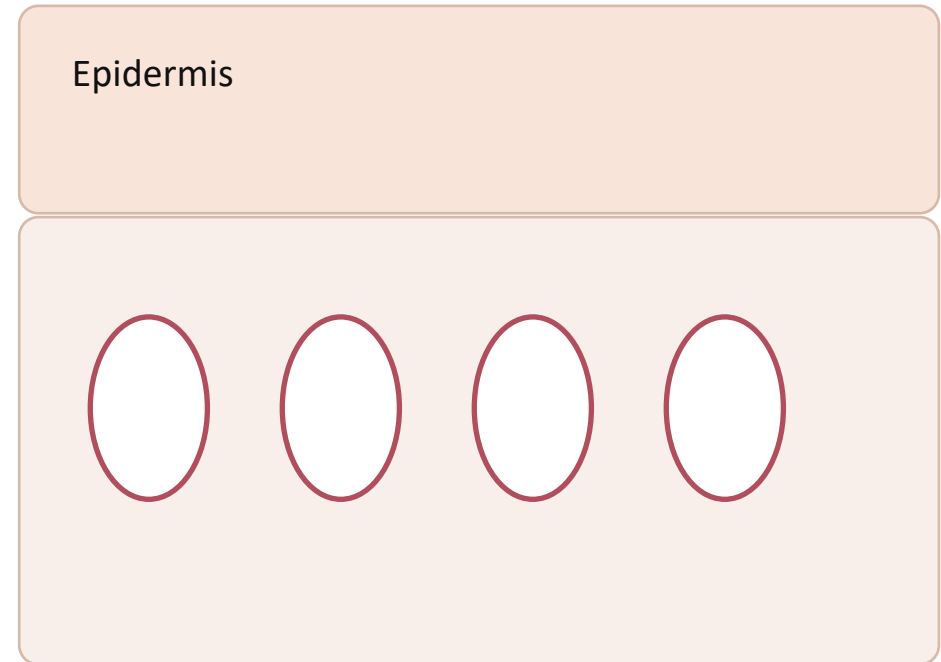
Schematic — not a histologic specimen

Nevus flammeus: A flat congenital vascular mark

- Nevus flammeus is described as a common vascular ectasia.
- It forms a light-pink to purple flat head or neck patch.
- The underlying process is dilated cutaneous vessels.
- Its clinical course distinguishes subtypes.

Port-wine stain: Persistence is diagnostic context

- A port-wine stain is a subtype of nevus flammeus.
- It typically persists instead of regressing.
- It can enlarge and thicken the overlying skin during childhood.
- An otherwise similar flat lesion may have a different time course.



Persistent ectatic vessels beneath flat patch

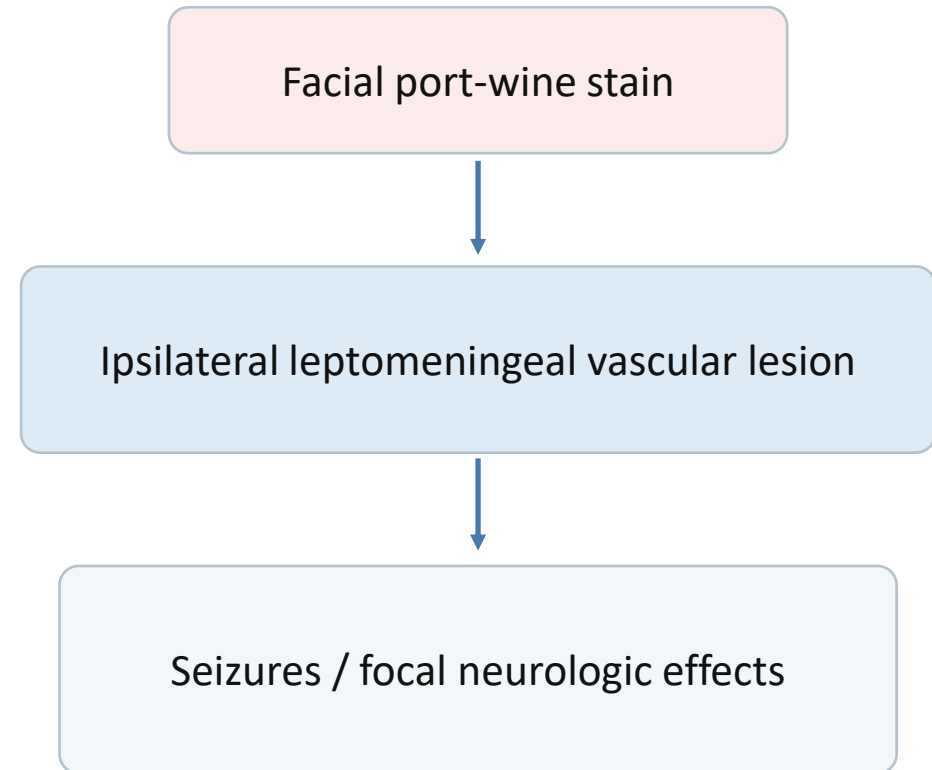
Schematic — not a histologic specimen

Port-wine stain: Anatomic pattern can be informative

- Facial port-wine stains may follow trigeminal territories.
- Some are associated with Sturge–Weber syndrome.
- An extensive facial lesion warrants attention to neurologic context.
- Anatomic association is not equivalent to syndrome diagnosis.

Sturge–Weber: Cutaneous and CNS vascular findings

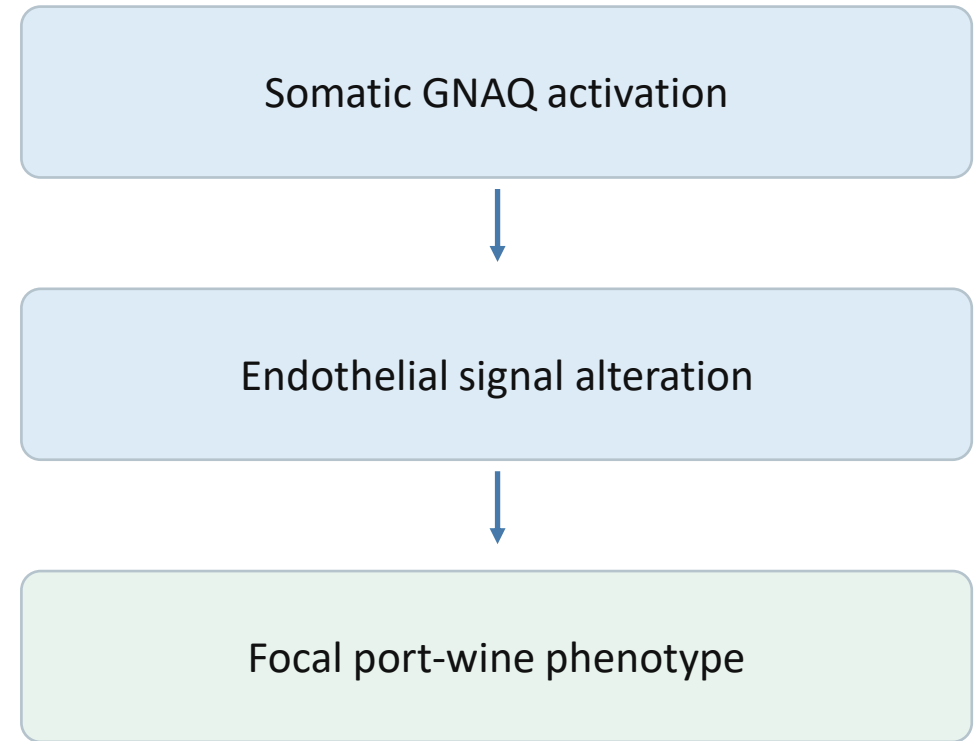
- Facial port-wine nevi can coexist with ipsilateral leptomeningeal lesions.
- The cortical venous abnormalities can cause neurologic disease.
- Seizures, weakness, and developmental impairment are described.
- Dermatologic and neurologic findings must be integrated.



Syndromic association; Not inevitable

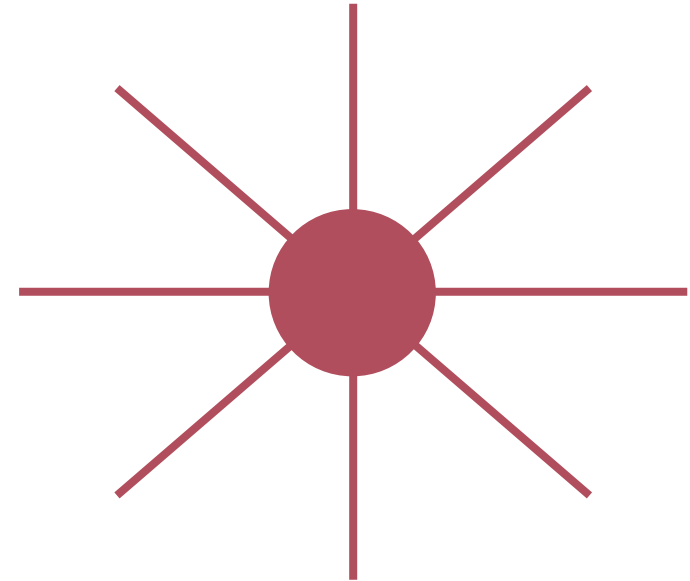
GNAQ links some port-wine stains to somatic signaling

- Most port-wine stains with activating GNAQ variants.
- GNAQ encodes a heterotrimeric G-protein α subunit.
- The alteration is somatic rather than necessarily inherited.
- A focal genetic alteration can produce a localized vascular phenotype.



Spider telangiectasia: Central core and radial vessels

- A central vascular focus gives rise to radiating small vessels.
- The lesion often appears pulsatile and blanchable.
- Face, neck, and upper chest are common locations.
- Its architecture differs from a solid vascular mass.



Radial telangiectatic vessels

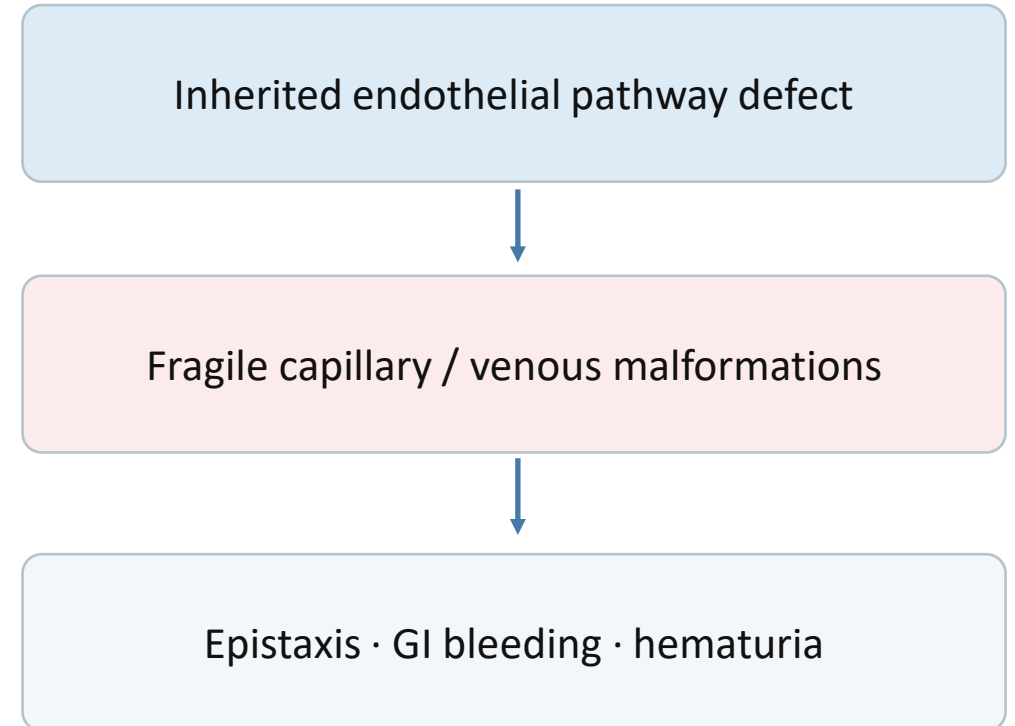
Schematic — not a histologic specimen

Spider telangiectasia and hormonal states

- Spider telangiectasias are nonneoplastic dilations.
- They occur frequently in hyperestrogenic settings.
- Pregnancy and cirrhosis are textbook associations.
- The lesion is a clinical clue, not evidence of a specific tumor.

HHT is an inherited vascular-malformation syndrome

- Hereditary hemorrhagic telangiectasia is autosomal dominant.
- Defects affect endothelial components of TGF- β signaling.
- Dilated capillaries and veins form widespread telangiectasias.
- Vascular fragility produces a characteristic bleeding phenotype.



HHT: Mucosal and systemic distribution

- Cutaneous and oral mucosal sites are common.
- Lesions can involve the respiratory tract.
- Gastrointestinal and urinary tracts may also be affected.
- Distribution helps distinguish systemic disease from a local patch.

HHT: Clinical consequences of fragile vessels

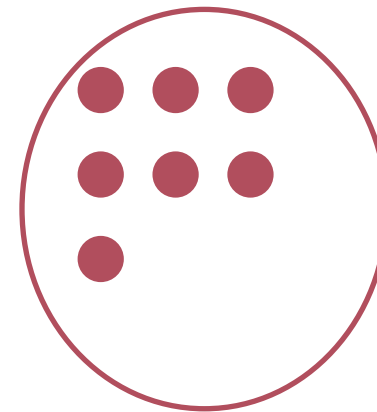
- Spontaneous epistaxis may be prominent.
- Gastrointestinal hemorrhage can be clinically significant.
- Urinary-tract lesions may cause hematuria.
- Repeated bleeding reflects vessel fragility, not tissue invasion.

Three vascular ectasia phenotypes

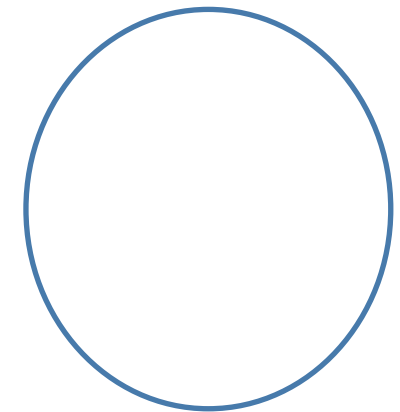
Phenotype	Morphologic clue	Clinical association
Port-wine stain	Persistent flat red-purple patch	Occasional Sturge–Weber link
Spider telangiectasia	Radial vessels around central core	Hyperestrogenic states
HHT	Widespread capillary/venous dilations	Inherited bleeding disorder

Lymphangioma involves lymphatic rather than blood channels

- Lymphangiomas are benign lymphatic counterparts of hemangiomas.
- Lymphatic lumina typically lack erythrocytes.
- Endothelial lining is still present.
- Lymphatic differentiation may be highlighted by specialized markers.



Blood: Erythrocytes



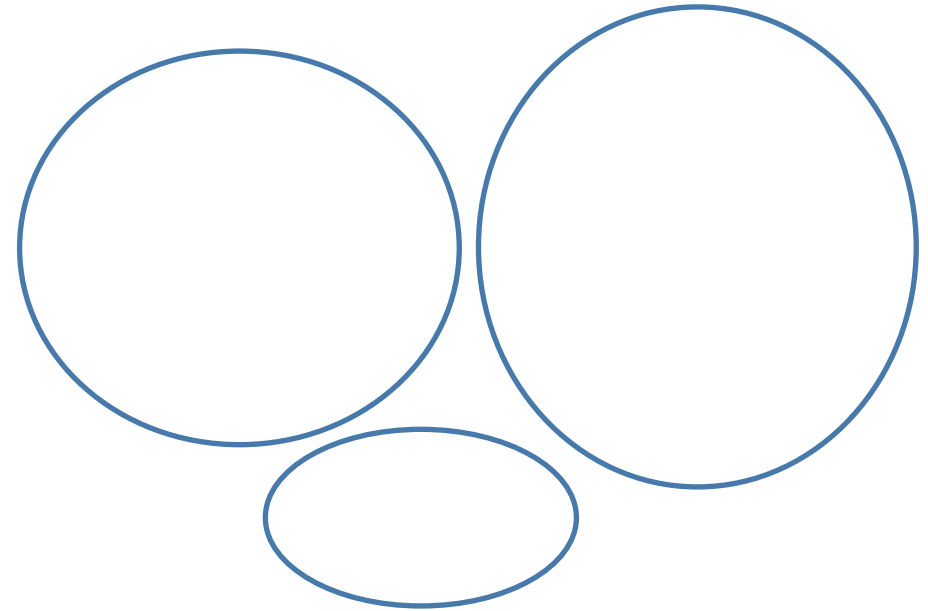
Lymph: Often no RBCs

Simple lymphangioma: Superficial pattern

- Simple capillary lymphangiomas can be slightly elevated.
- Small pedunculated lesions may also occur.
- They preferentially involve head, neck, and axillary regions.
- Histology reveals networks of endothelial-lined lymphatic spaces.

Cystic hygroma: Markedly expanded lymphatics

- Cavernous lymphangioma is also called cystic hygroma.
- The neck and axilla are characteristic childhood sites.
- Large lesions can cause visible regional deformity.
- Massively dilated lymphatic spaces define their architecture.



Enlarged endothelial-lined lymphatic spaces

Schematic — not a histologic specimen

Cystic hygroma: Supporting tissue and margins

- Cysts are separated by connective tissue stroma.
- Lymphoid aggregates can occur within the intervening tissue.
- The lesion is often unencapsulated with indistinct margins.
- These boundaries make complete resection challenging.

A clinicopathologic association: Turner syndrome

- Cervical cystic hygromas in Turner syndrome.
- The association reflects abnormal lymphatic development.
- A neck mass alone cannot establish a chromosomal disorder.
- Integrate morphology with the full clinical setting.

Lymphatic immunophenotype: Interpret the purpose

- VEGFR-3 and LYVE-1 are cited lymphatic endothelial markers.
- Their purpose is to support lymphatic lineage.
- Blood-filled versus lymph-filled spaces provide another clue.
- A marker never substitutes for lesion architecture and context.

Glomus tumor is vessel-associated but not an hemangioma

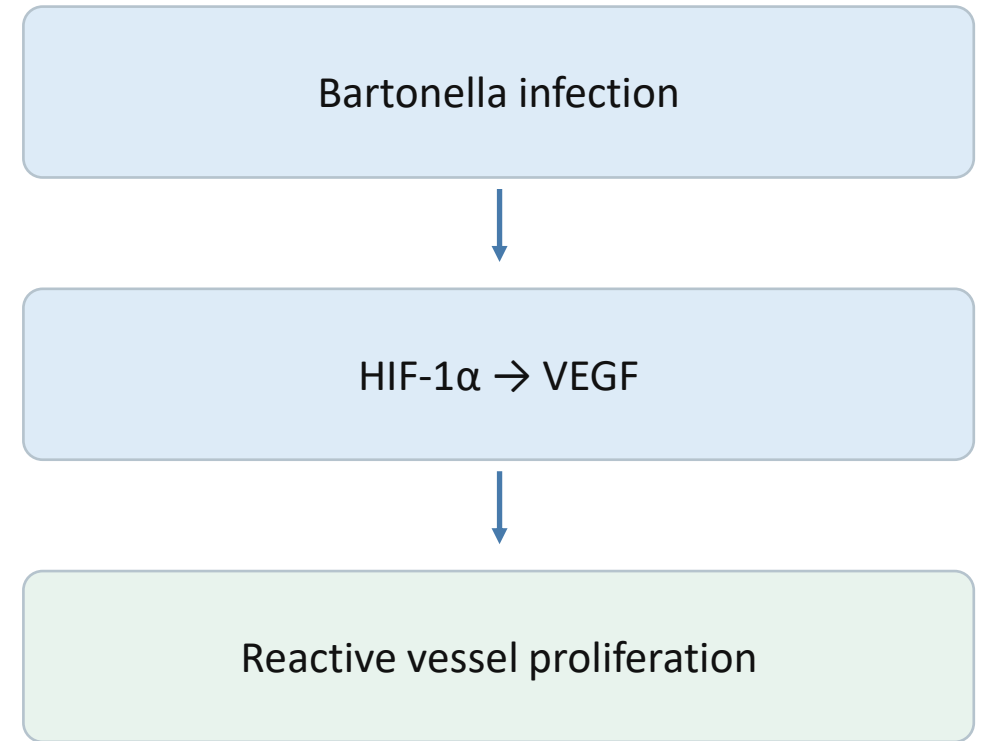
- Glomus tumors arise from modified smooth muscle cells.
- They are related to thermoregulatory arteriovenous structures.
- Subungual location and marked pain are characteristic clues.
- Cell of origin, not vascular appearance alone, is decisive.

Bacillary angiomatosis: Infectious vascular mimic

- This reactive proliferation occurs in immunocompromised hosts.
- Bartonella species are the causative pathogens.
- Skin, bone, brain, and other sites can be involved.
- Recognition redirects interpretation from neoplasia to infection.

Bartonella-driven angiogenesis has a defined sequence

- Bartonella infection can increase tissue HIF-1 α activity.
- HIF-1 α promotes vascular endothelial growth factor production.
- VEGF stimulates the local vascular proliferative response.
- The process resolves with appropriate antimicrobial treatment.



Endothelial markers solve a specific diagnostic problem

- CD31 and von Willebrand factor support endothelial lineage.
- Such stains are useful when malignant lesions lose vascular channels.
- They do not by themselves distinguish all tumor subtypes.
- Morphology and clinical context remain indispensable.

Vascular lesions span biologic categories

Category	Textbook examples	Key discriminator
Benign endothelial tumors	Capillary / cavernous hemangioma	Organized blood-filled vessels
Nonneoplastic malformations	Nevus flammeus; cerebral AVM	Abnormal established channels
Lymphatic lesions	Simple / cavernous lymphangioma	Lymphatic endothelial-lined spaces
Reactive proliferation	Bacillary angiomatosis	Infectious context
Malignant vascular tumor	Angiosarcoma	Atypical, poorly formed vessels

Four groups of cerebral vascular malformations

- Arteriovenous malformations form one major category.
- Cavernous malformations are a distinct low-flow category.
- Capillary telangiectasias are a third group.
- Venous angiomas form the fourth group.

Arteriovenous

High flow

Cavernous

Low flow

Capillary telangiectasia

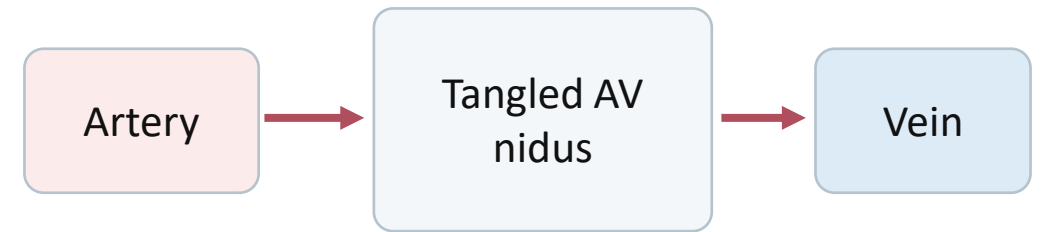
Small vessels

Venous angioma

Venous drainage

Cerebral AVM: High-flow arteriovenous shunting

- Abnormal arterial channels connect to the venous side.
- Capillary resistance is bypassed within the shunting network.
- Blood flow is relatively high and often pulsatile.
- The malformation can affect brain or subarachnoid tissues.



Direct abnormal AV connection; No normal capillary bed

Cerebral AVM: Microscopic organization

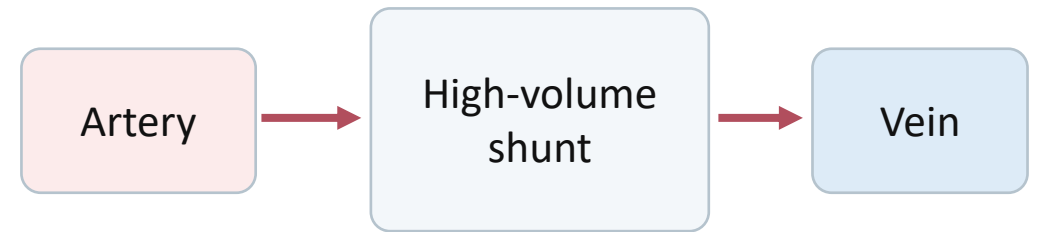
- Tangled, enlarged vessels form a wormlike network.
- Gliotic brain tissue may separate the malformed vessels.
- Old hemorrhage can be evident.
- Arterial elastic lamina and vessel-wall architecture may be abnormal.

Cerebral AVM: Major clinical presentations

- Seizures can be the first presentation.
- Intracerebral or subarachnoid hemorrhage can occur.
- A common presentation in ages 10–30.
- The middle cerebral artery territory is frequently involved.

Vein of Galen malformation: Neonatal hemodynamic risk

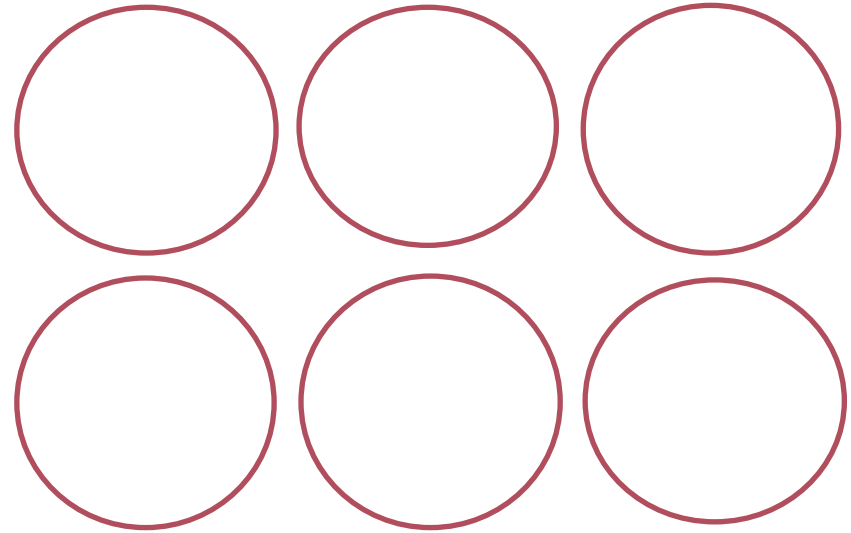
- An arteriovenous shunt can deliver high-pressure blood to veins.
- A neonatal vein of Galen lesion may produce a large shunt.
- The increased cardiac flow burden can lead to heart failure.
- This is a hemodynamic hazard distinct from cosmetic skin lesions.



Neonatal high-flow burden → cardiac failure

Cerebral cavernous malformation: Back-to-back channels

- Distended vascular spaces cluster directly against each other.
- Walls are collagenized and vary in thickness.
- Brain tissue is often sparse or absent between channels.
- Old hemorrhage, infarction, or calcification may surround the lesion.



Collagenized, adjacent low-flow channels

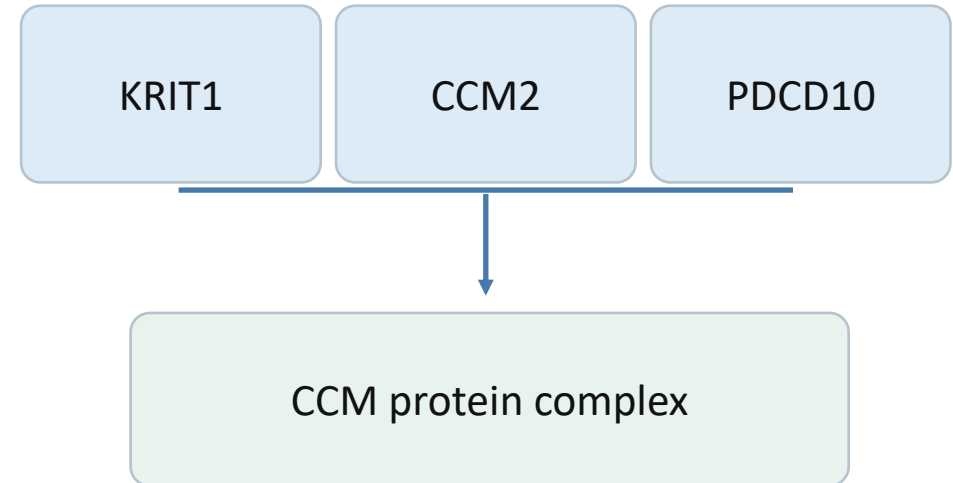
Schematic — not a histologic specimen

Cerebral cavernous malformation: Low-flow physiology

- Cavernous malformations do not create an arteriovenous shunt.
- Their channels carry relatively low-flow blood.
- Hemorrhage and focal neurologic effects remain possible.
- Physiology distinguishes them from high-flow AVMs.

Familial cerebral cavernous malformations

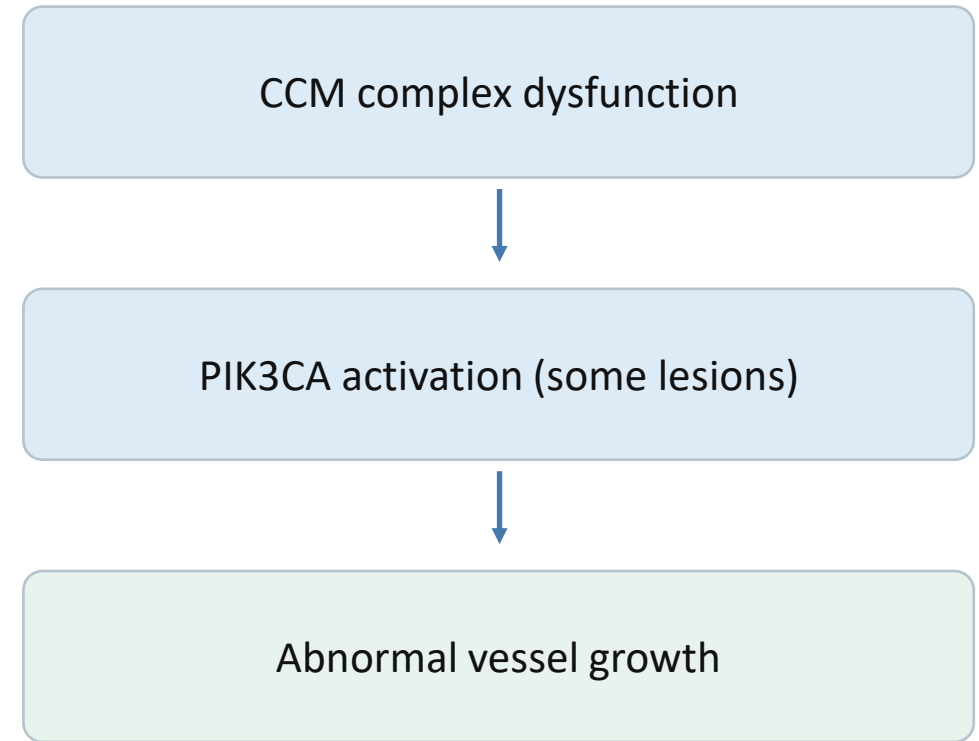
- Familial forms often feature more than one lesion.
- The inherited pattern is autosomal dominant.
- Genes include KRIT1, CCM2, and PDCD10.
- These genes encode elements of a CCM protein complex.



Inherited loss of function predisposes to lesions

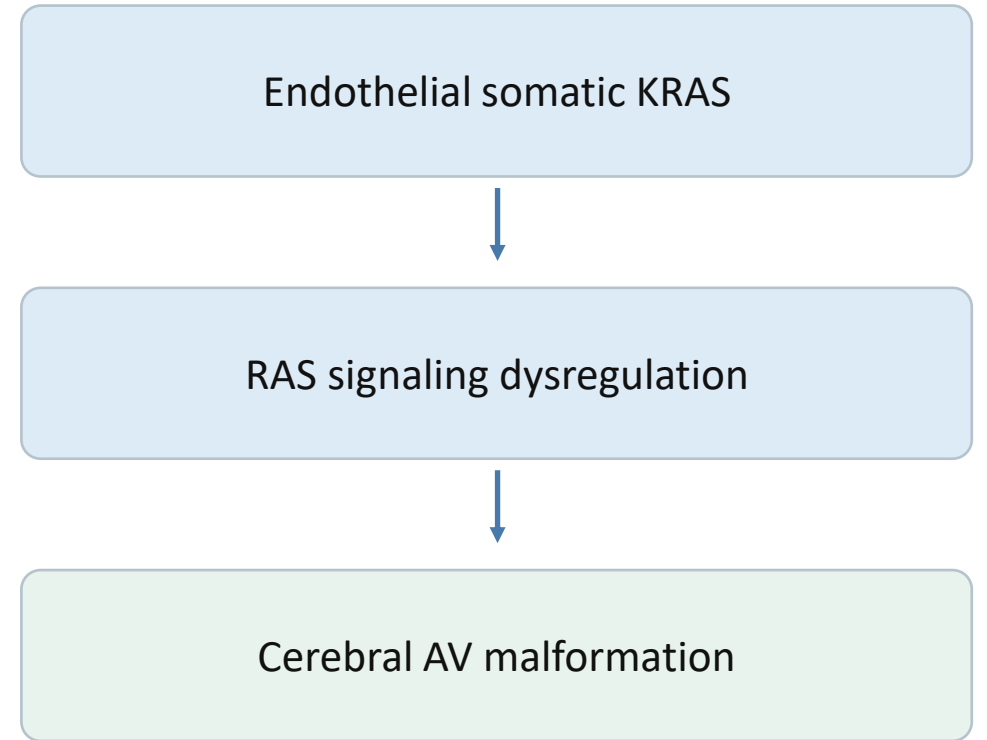
CCM signaling and acquired cooperating alterations

- Loss of CCM complex function releases restraints on vessel growth.
- PIK3CA activating changes in growing lesions.
- Both inherited susceptibility and somatic change may participate.
- This mechanism should not be generalized to every vascular mark.



Cerebral AVM: A different molecular association

- Activating somatic KRAS mutations in many AVMs.
- Endothelial RAS signaling is implicated in abnormal vessel formation.
- This differs from the familial CCM gene complex.
- Similar clinical risks do not imply the same molecular mechanism.



AVM versus cerebral cavernous malformation

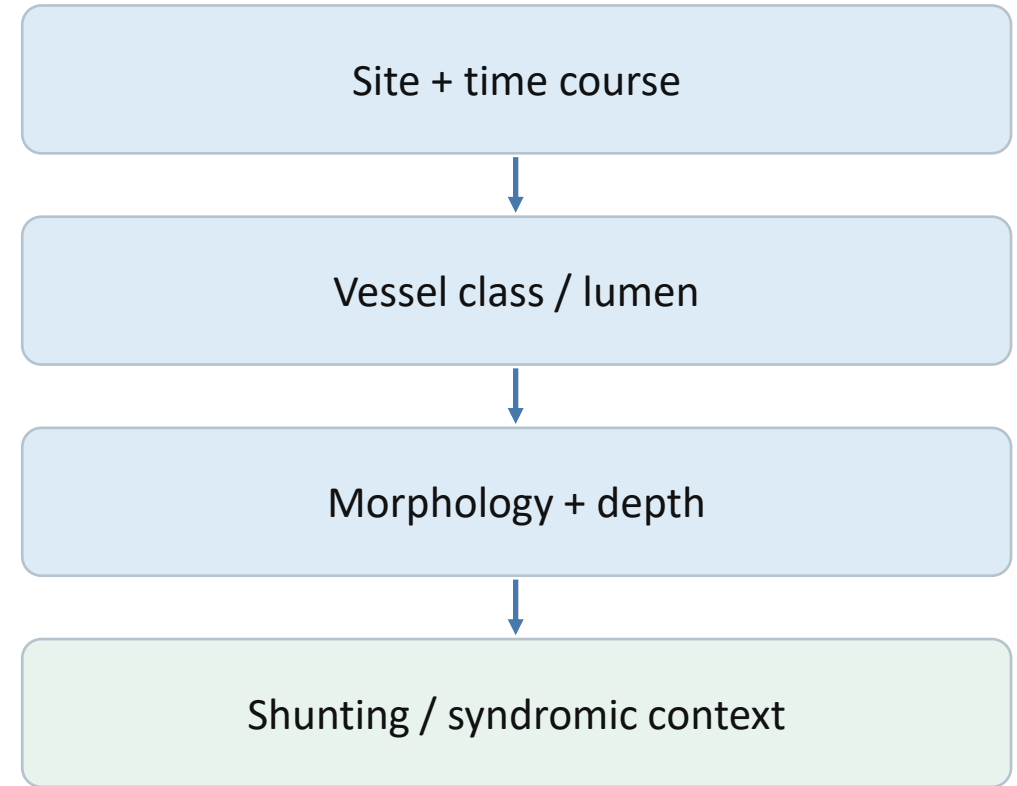
Feature	Cerebral AVM	Cerebral cavernous malformation
Hemodynamics	High-flow shunt	Low-flow; no AV shunt
Architecture	Arterial/venous tangle; gliosis	Back-to-back distended channels
Neural tissue between	Often gliotic tissue	Often absent
Molecular association	Somatic KRAS activation	CCM genes; PIK3CA cooperation
Risks	Seizures, hemorrhage, shunt complications	Hemorrhage and local neurologic dysfunction

Do not conflate VHL lesions with familial CCM

- VHL is a separate inherited tumor-predisposition syndrome.
- Its characteristic CNS vascular tumors include hemangioblastomas.
- Familial CCM instead centers on KRIT1, CCM2, and PDCD10.
- Use precise anatomic and pathologic nomenclature.

An interpretive sequence for a vascular lesion

- First establish site, depth, and observed natural history.
- Next distinguish proliferative mass from dilated vessels.
- Then classify blood-filled, lymphatic, or supportive-cell architecture.
- For cerebral lesions, distinguish shunting from low flow.



Case application: Persistent facial patch

- A child has a flat red-purple cheek lesion since infancy.
- It has not followed a proliferative-then-involutorial course.
- The lesion has become more conspicuous and thicker over time.
- Question: What vascular category best explains this history?

Case interpretation: Port-wine stain

- A persistent flat vascular mark favors nevus flammeus, port-wine type.
- Persistence separates it from the usual juvenile hemangioma course.
- Facial distribution prompts consideration of associated CNS findings.
- The vignette alone does not diagnose Sturge–Weber syndrome.

Case application: Friable gingival nodule

- A young adult develops a rapidly enlarging red gingival nodule.
- It is pedunculated and bleeds after minor contact.
- There is a possible recent local traumatic trigger.
- Question: Which benign vascular pattern is suggested?

Case interpretation: Pyogenic granuloma

- Rapid growth, pedunculation, and bleeding favor pyogenic granuloma.
- The name does not imply bacterial suppuration.
- Oral location and trauma are compatible supporting clues.
- Histologic examination remains relevant if presentation is atypical.

Case application: Repeated cerebral hemorrhage

- A patient has multiple CNS vascular lesions and recurrent focal symptoms.
- Imaging suggests low-flow vascular spaces without an AV shunt.
- Family history indicates similar lesions in relatives.
- Question: Which malformation family and genes merit consideration?

Case interpretation: Familial CCM

- Low-flow clustered channels fit cerebral cavernous malformations.
- Multiplicity and family history support a familial form.
- KRIT1, CCM2, and PDCD10 are the textbook genes.
- This is distinct from an AVM driven by arteriovenous shunting.

Five discriminators that organize the subject

- Proliferation versus malformed preexisting channels.
- Small capillaries versus broad cavernous spaces.
- Regression versus persistence or deep extension.
- Blood-filled channels versus lymphatic spaces.
- High-flow AVM versus low-flow cerebral cavernous malformation.

Source and scope

- Primary: Robbins, Cotran & Kumar, Pathologic Basis of Disease, 11th ed. (2026), Chapter 10, pp. 432–433.
- Related textbook context: Chapter 11, pp. 472–474, vascular tumors, vascular ectasias and lymphatic lesions.
- CNS extension: Chapter 28, pp. 1143–1144, cerebral vascular malformations.
- Conceptual drawings are original editable schematics, not diagnostic specimens.