

Familial Tumor Syndromes

Inherited predisposition to tumors of the nervous system

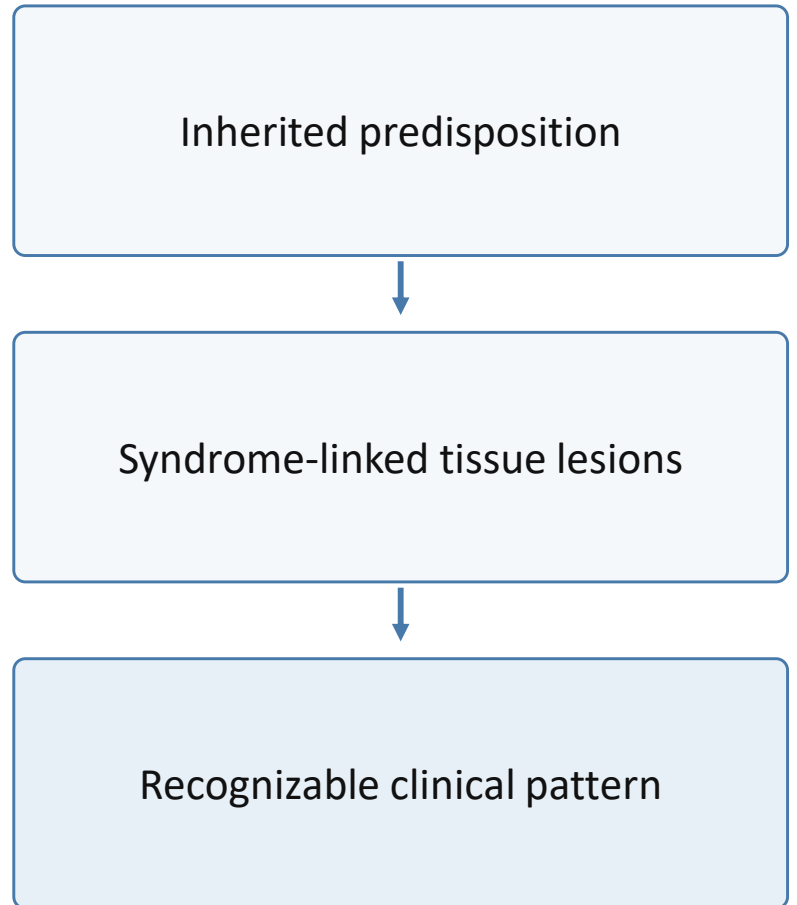
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Inherited predisposition, distinctive tumor spectra

- Inherited syndromes increase risk of characteristic neoplasms rather than one uniform “brain tumor.”
- Tumor type and anatomic location often suggest the underlying syndrome.
- Major examples in the CNS chapter: NF1, NF2, schwannomatosis, TSC and VHL.



The four major clinicopathologic patterns

- NF1: Neurofibromas, optic pathway gliomas and pigmentary findings.
- NF2 / schwannomatosis: Schwann-cell tumors with different distributions.
- TSC: Cortical and subependymal hamartomas; SEGA.
- VHL: Hemangioblastomas with systemic tumor and cyst associations.

NF1

Neurofibroma + glioma

**NF2 /
schwannomatosis**

Schwannoma pattern

TSC

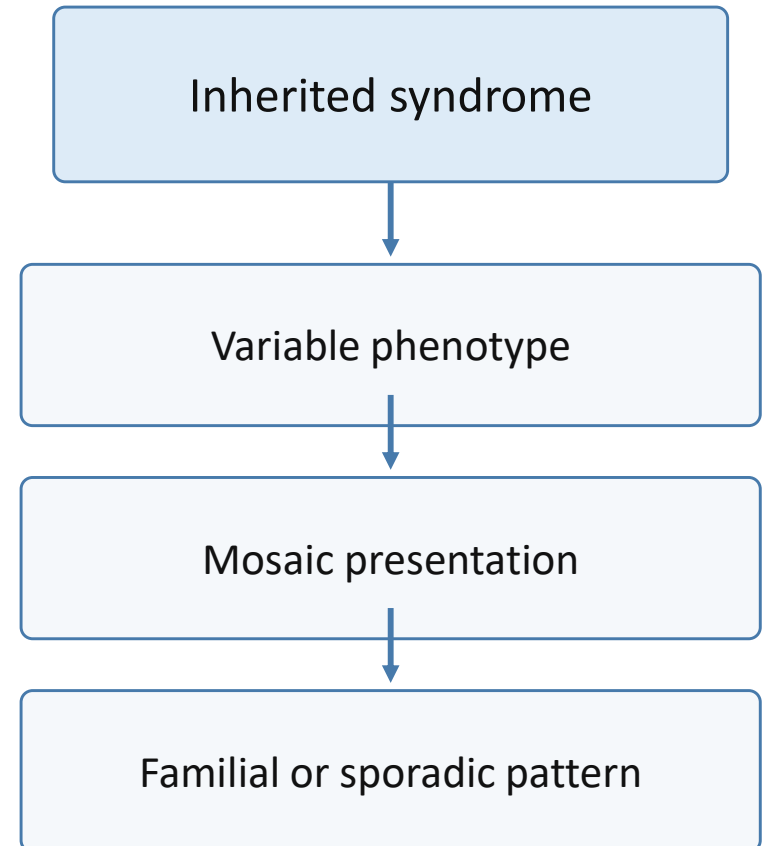
Hamartoma + SEGA

VHL

Hemangioblastoma

Why family history cannot be the only clue

- Autosomal dominant inheritance characterizes NF1, NF2, TSC and VHL.
- NF1 has high penetrance but variable expressivity; Limited distribution may reflect mosaicism.
- Schwannomatosis may be familial or sporadic description.



Genotype links to an altered cellular restraint

- NF1: Loss of neurofibromin removes a restraint on RAS activation.
- TSC1/TSC2: Loss of hamartin–tuberin control elevates mTOR activity.
- VHL: Impaired HIF down-regulation favors angiogenic signaling.
- NF2: Merlin loss affects growth, cell shape and adhesion pathways.

NF1

RAS

TSC1/TSC2

MTOR

VHL

HIF / VEGF

NF2

Merlin-regulated signaling

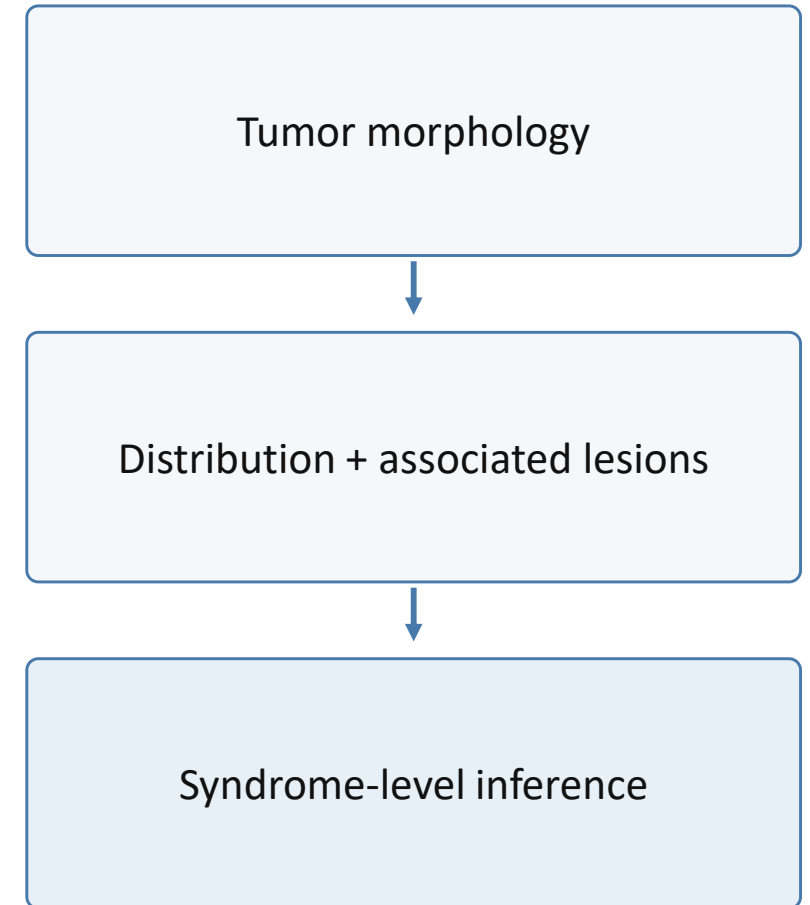
A benign mass may be neurologically dangerous

- Cortical tubers can be epileptogenic despite their hamartomatous nature.
- Subependymal giant cell astrocytoma may obstruct ventricular CSF flow.
- Vestibular schwannoma may compromise hearing or compress adjacent neural tissue.
- Hemangioblastoma may cause mass effect in the cerebellum.

- Cortex: Seizures
- Ventricle: Obstruction
- CN VIII: Hearing
- Cerebellum: Mass effect

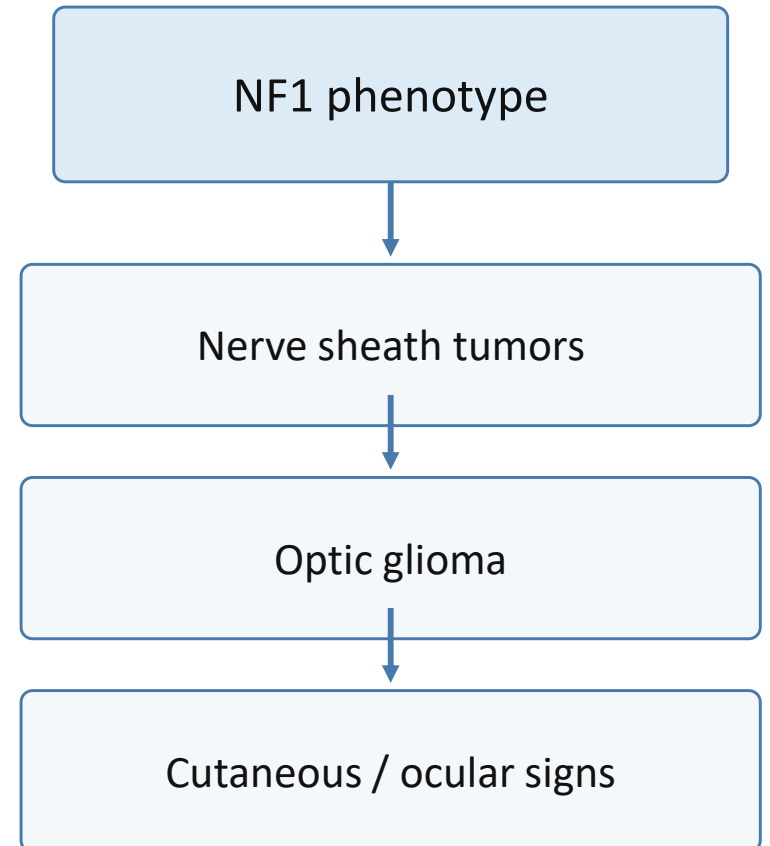
The central diagnostic task: Recognize the pattern

- Synthesize tumor histology with the number and distribution of lesions.
- Assess accompanying skin, eye, renal and other systemic manifestations.
- Separate a solitary sporadic tumor from a syndromic constellation.



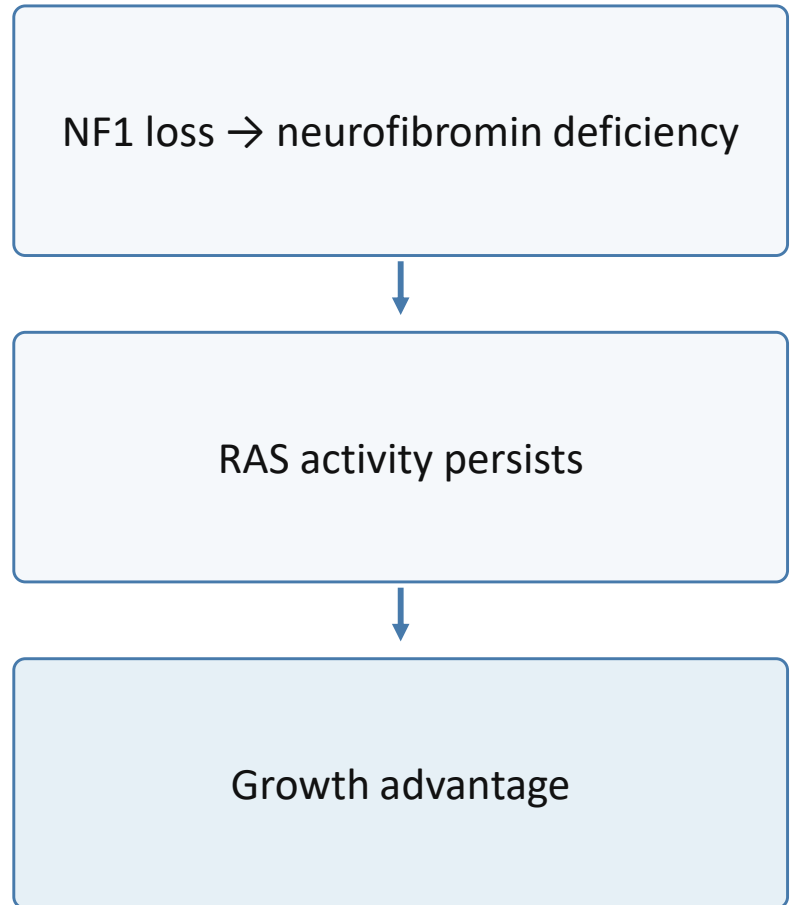
NF1 combines nerve sheath tumors and developmental signs

- Autosomal dominant syndrome occurring in approximately 1 in 3,000 individuals.
- Associated tumors include neurofibromas, optic nerve gliomas, other gliomas and MPNST.
- Pigmentary, ocular, skeletal and neurologic changes complete the phenotype.



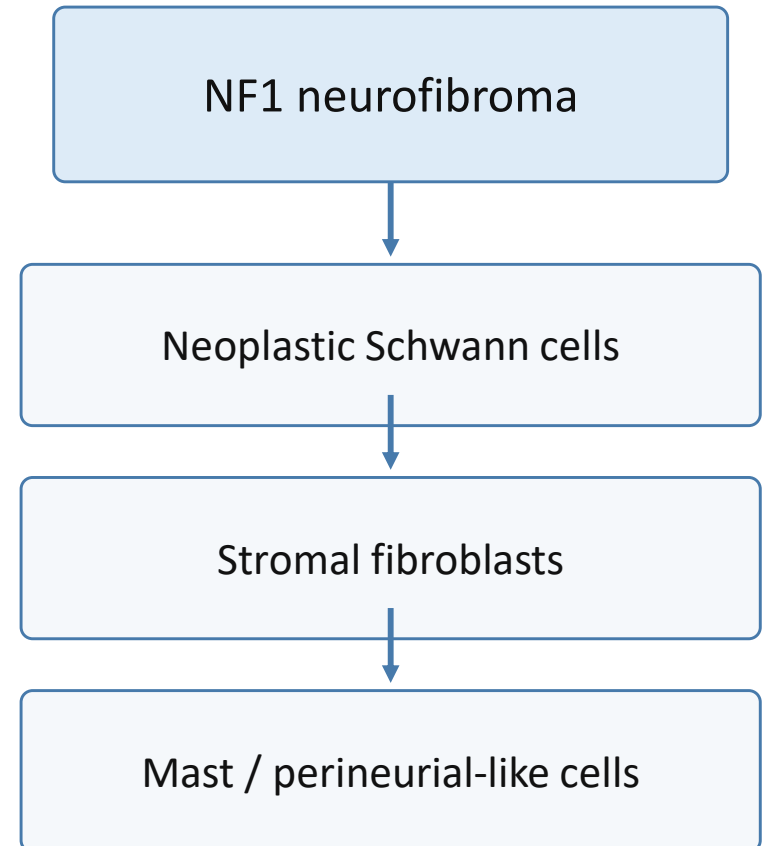
NF1 is a RAS-pathway tumor suppressor disorder

- NF1 maps to chromosome 17q11.2 and encodes neurofibromin.
- Neurofibromin limits active RAS signaling through GTPase-related regulation.
- Its loss permits prolonged RAS activation and growth signaling.



Biallelic NF1 loss identifies neoplastic Schwann cells

- The inherited NF1 defect alone does not make all tissue cells neoplastic.
- Schwann cells of NF1-related neurofibromas show complete loss of neurofibromin.
- Other tumor components can retain one functional allele and participate in the microenvironment.



Variable expressivity is part of NF1 pathology

- Some patients have subtle disease while others show extensive lesions.
- Mosaic NF1 can produce a phenotype restricted to a body region.
- Large deletions involving NF1 and adjacent genes correlate with greater severity.

Broad phenotype

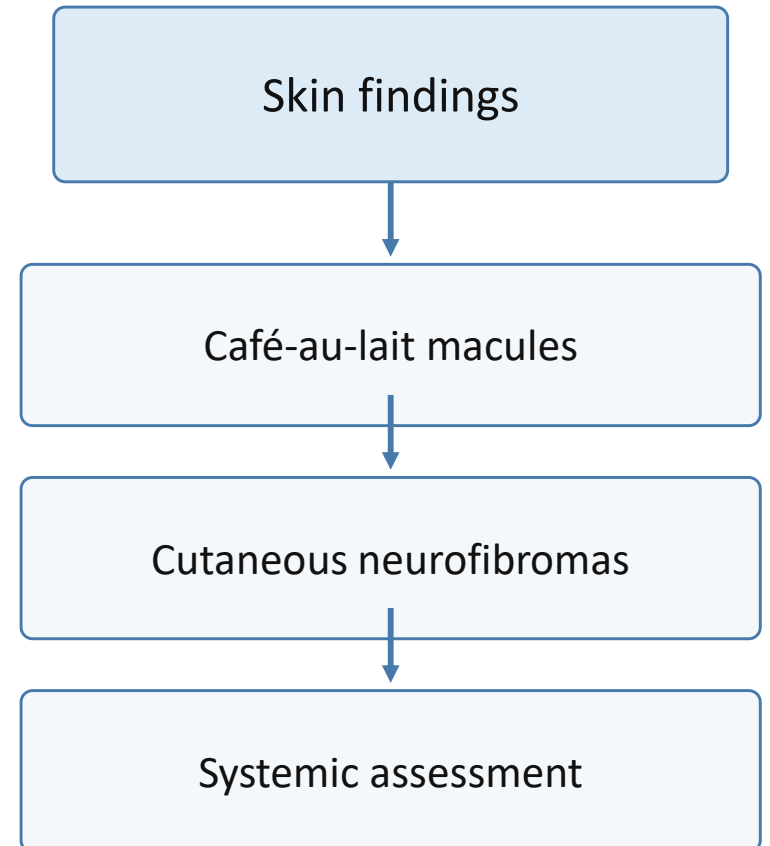
Generalized lesions

Mosaic phenotype

Region-restricted
lesions

Cutaneous signs provide clues beyond the tumor

- Café-au-lait macules are hyperpigmented cutaneous patches.
- Superficial cutaneous neurofibromas may be multiple.
- Other affected tissues should be evaluated when these findings coexist.



The eye may show both pigmentary and neoplastic lesions

- Iris Lisch nodules are pigmented nodules linked to NF1.
- Optic nerve gliomas are a prominent neoplastic manifestation.
- The combination connects a visible ocular clue with a CNS tumor predisposition.

Lisch nodules

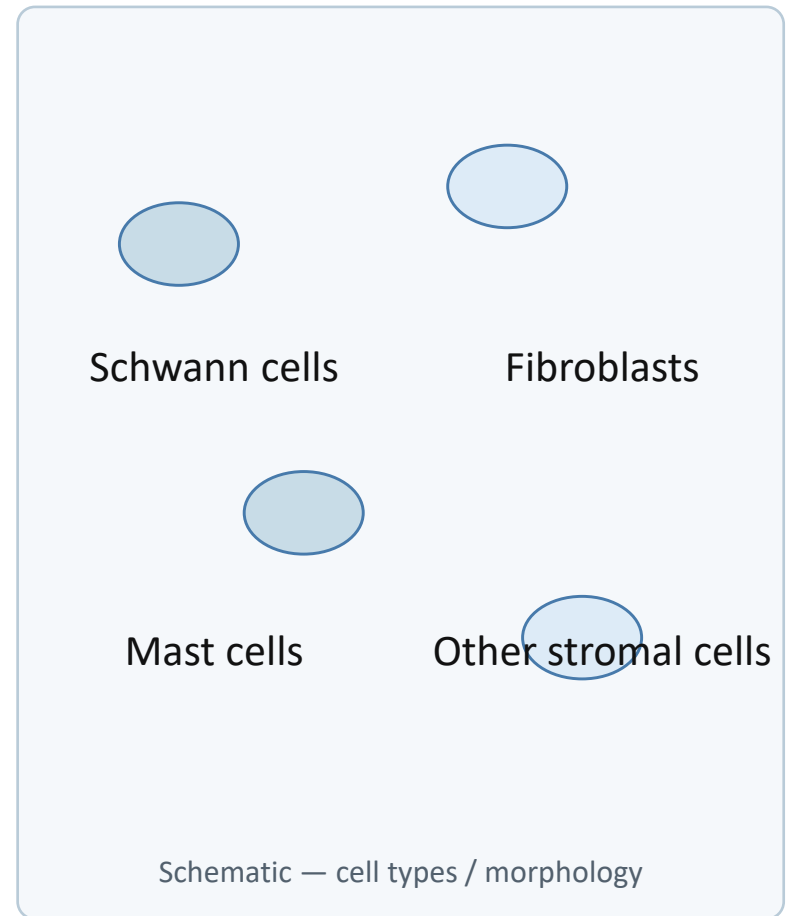
Iris pigmentation

Optic glioma

Optic pathway
tumor

Neurofibromas are heterogeneous nerve sheath tumors

- Neoplastic Schwann cells coexist with fibroblasts, perineurial-like cells and mast cells.
- CD34-positive spindle cells also contribute to the admixture.
- The variable extracellular matrix helps explain their diverse patterns.



Three growth patterns of neurofibroma

- Localized cutaneous: Well-delineated dermal/subcutaneous nodule.
- Diffuse: Plaque-like infiltration through skin and deeper tissue.
- Plexiform: Expansion of multiple nerve fascicles along a nerve.

**Localized
cutaneous**

Nodular

Diffuse

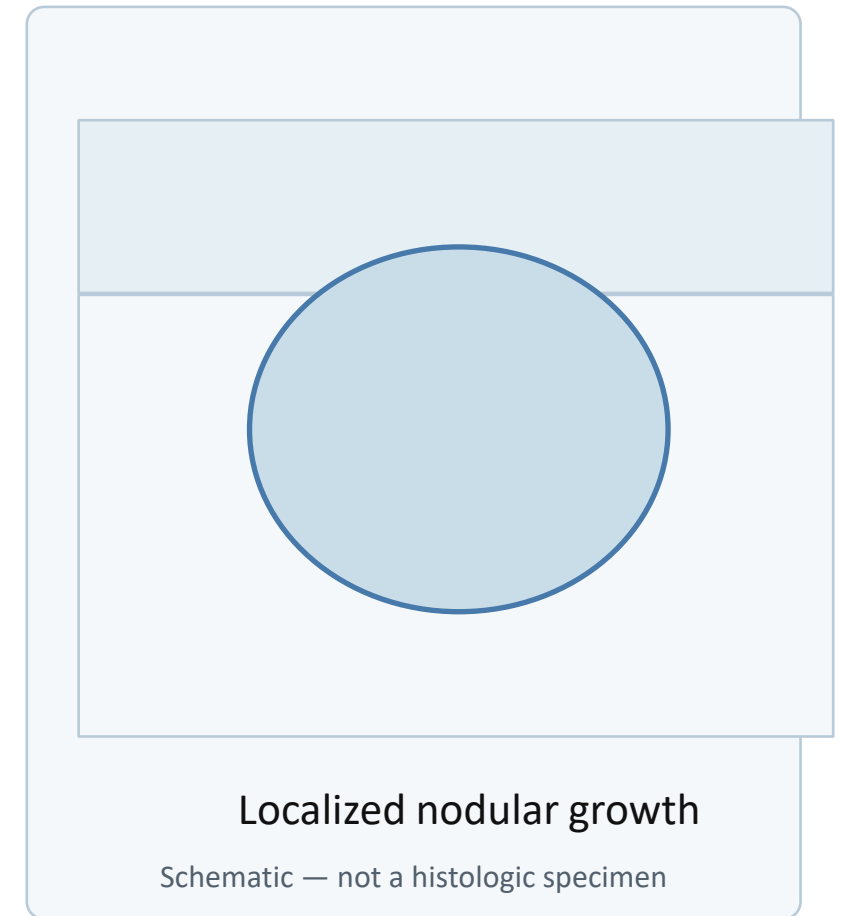
Plaque-like, infiltrative

Plexiform

Multiple enlarged
fascicles

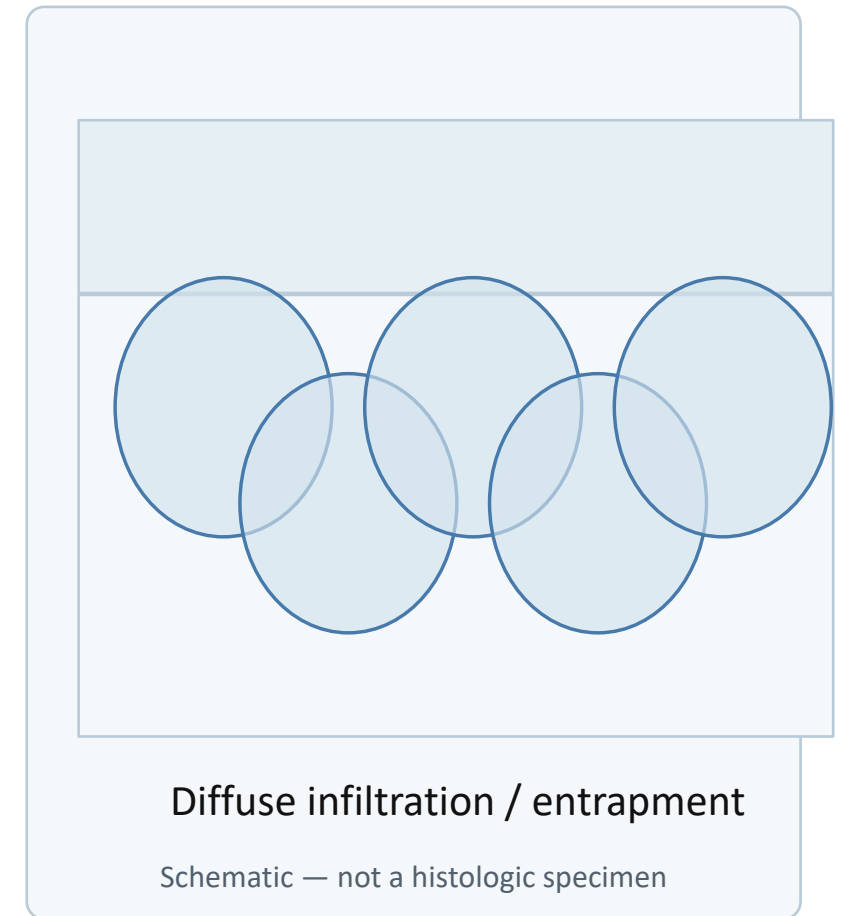
Localized cutaneous neurofibroma: Low-cellularity lesion

- Small, unencapsulated nodules in the dermis or subcutaneous fat.
- Bland Schwann cells intermingle with stromal elements and loose collagen.
- Adnexal structures may become entrapped near the lesion edge.



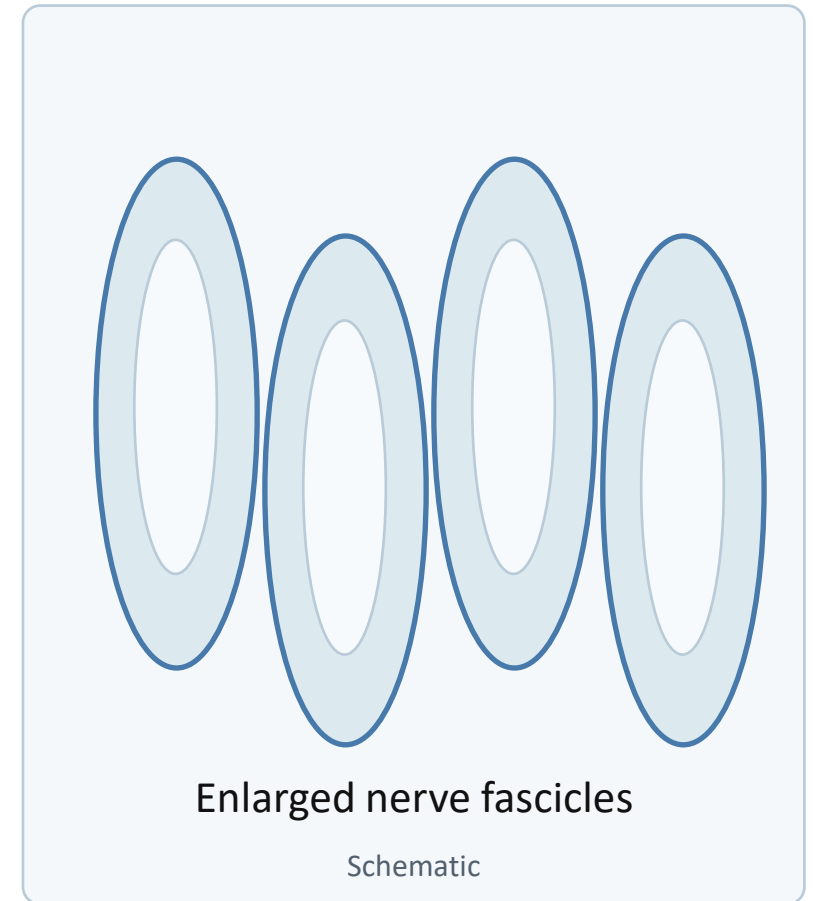
Diffuse neurofibroma spreads through tissue planes

- Infiltrates dermis, subcutaneous connective tissue or deeper soft tissue.
- Entraps adipose tissue and skin appendages as it expands.
- Pseudo-Meissner corpuscles may occur within the lesion.



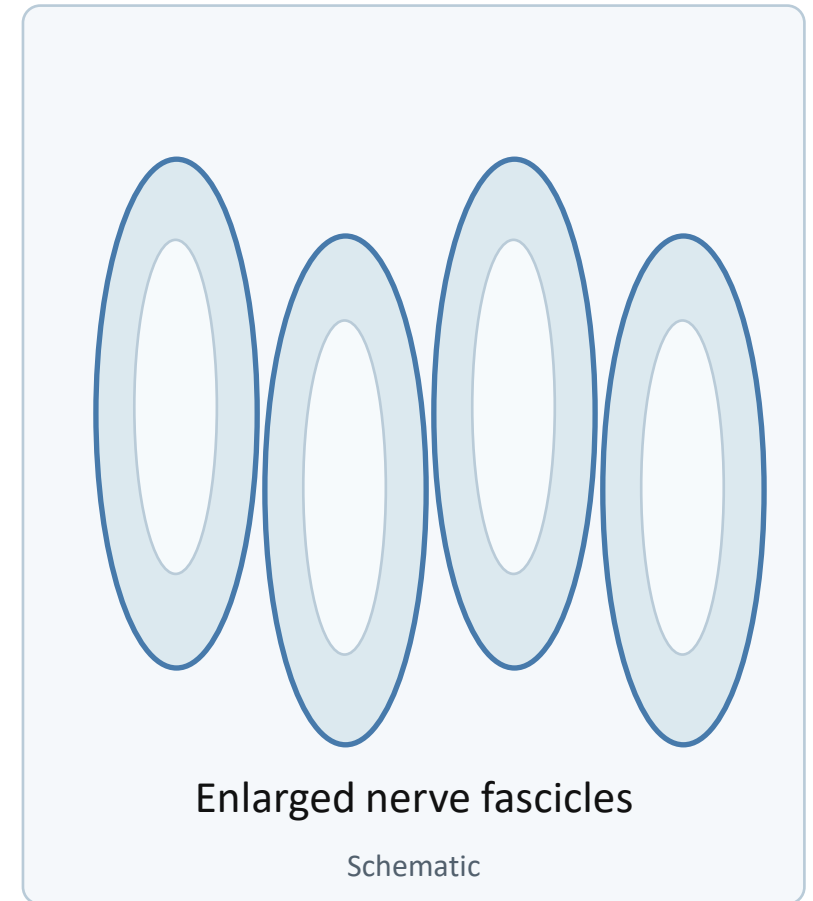
Plexiform neurofibroma expands nerve fascicles

- Arises in association with large nerves or nerve roots.
- Multiple fascicles enlarge, producing a rope-like “bag of worms” mass.
- This pattern is strongly linked to NF1.



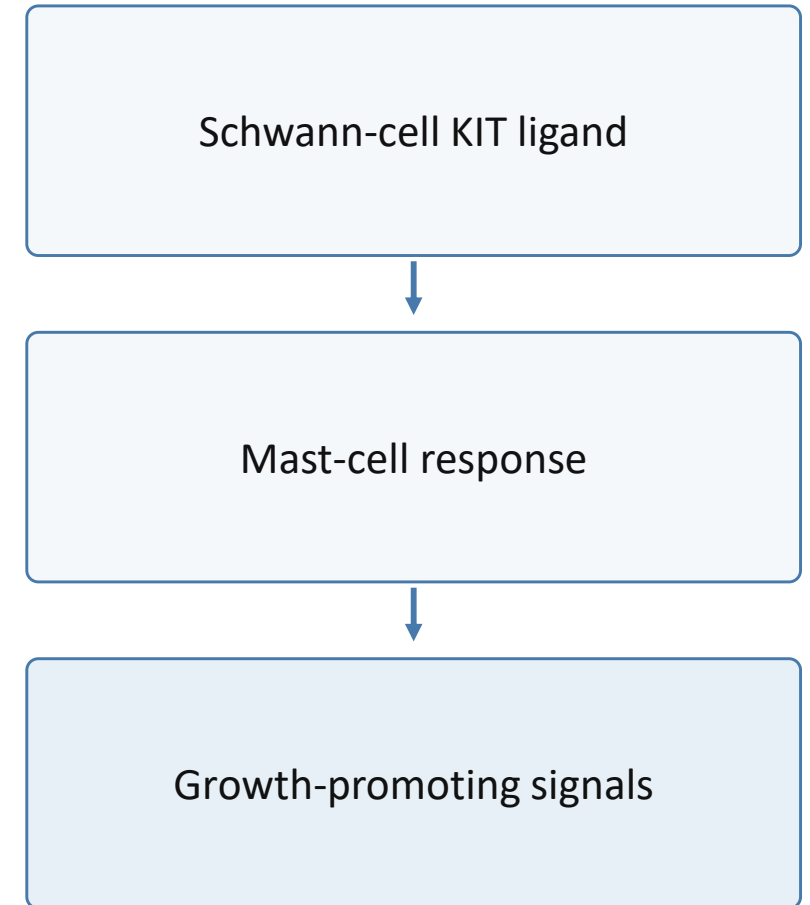
Plexiform neurofibroma: Microscopic architecture

- Tumor cells permeate and expand individual nerve fascicles while entrapping axons.
- Perineurial coverings can remain recognizable around expanded fascicles.
- Collagen bundles may appear reminiscent of shredded carrots.



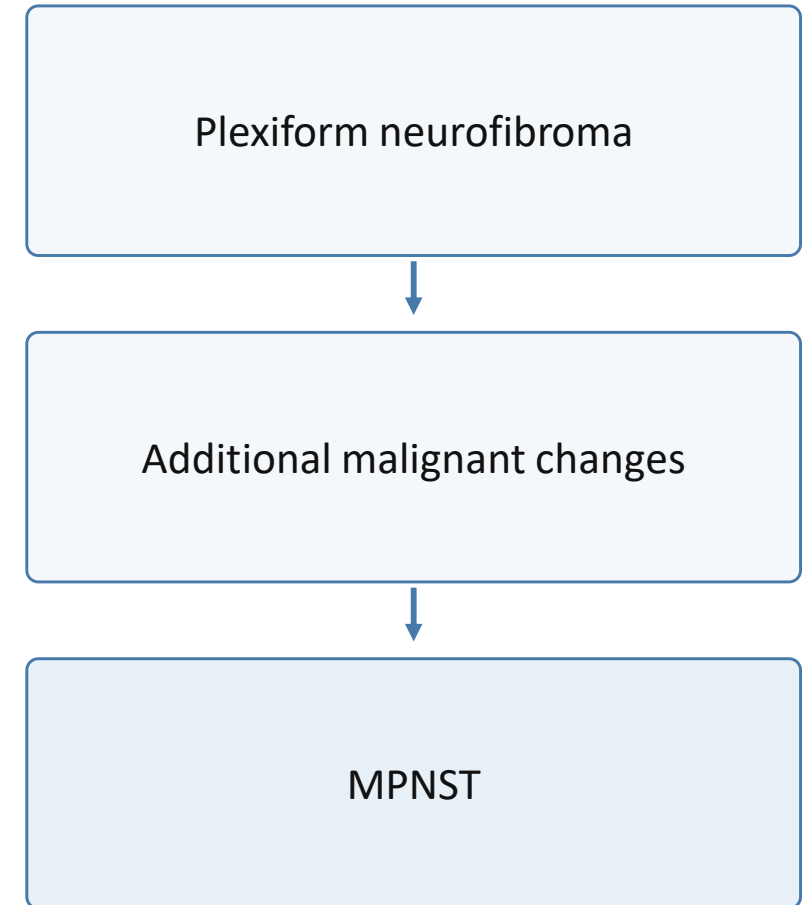
The tumor microenvironment matters in NF1

- NF1-haploinsufficient mast cells may respond strongly to KIT ligand from Schwann cells.
- Mast-cell factors can then stimulate Schwann-cell growth.
- Tumor–stroma signaling may reinforce growth of the lesion.



Plexiform lesions carry an important malignant risk

- MPNST can arise by malignant transformation of a preexisting neurofibroma.
- Transformation is most often associated with the plexiform variant.
- MPNST occurs in approximately 5%–10% of patients with NF1.



MPNST differs fundamentally from benign neurofibroma

- High-grade MPNST shows infiltrative growth into neighboring soft tissue.
- Mitoses, necrosis and nuclear anaplasia are common.
- A fascicular spindle-cell pattern may retain evidence of nerve sheath origin.

Benign neurofibroma

Bland mixed cells

Loose collagen

MPNST

Anaplasia /
mitoses

Invasion / necrosis

Histologic clues to MPNST in an NF1 patient

- A mass may appear variably cellular and “marbleized” at low power.
- Spindled tumor cells form infiltrative fascicles with malignant cytology.
- Nerve association and prior neurofibroma help distinguish it from other sarcomas.

Architecture

Infiltrative fascicles

Cytology

Anaplasia, mitoses

Context

Nerve / neurofibroma
association

MPNST progression may involve further suppressor loss

- NF1 loss is a founding feature in syndromic tumors.
- High-grade MPNST may show PRC2 pathway disruption and CDKN2A/B deletion.
- Malignancy therefore reflects additional alterations beyond the original predisposition.

Inherited NF1 susceptibility



Neurofibroma formation



Additional changes in MPNST

NF1: Link the clinical clue to the tissue lesion

- Multiple pigmentary findings plus nerve sheath nodules favor syndromic evaluation.
- A plexiform lesion requires different risk interpretation from a small cutaneous nodule.
- Pathology reports should specify neurofibroma subtype and any concerning malignant features.

Skin / ocular phenotype



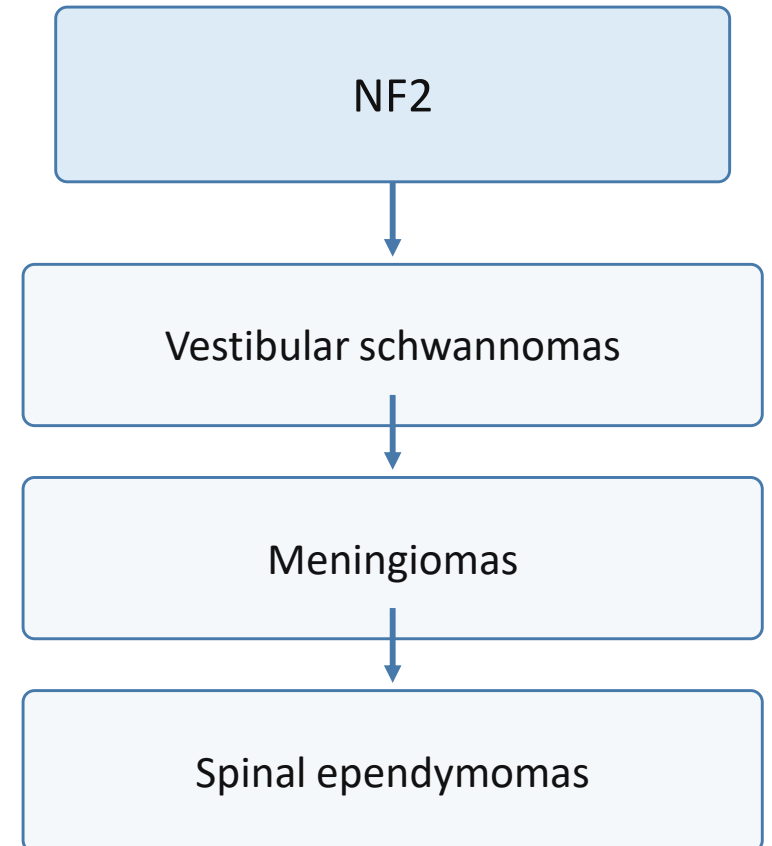
Neurofibroma subtype



Risk-oriented interpretation

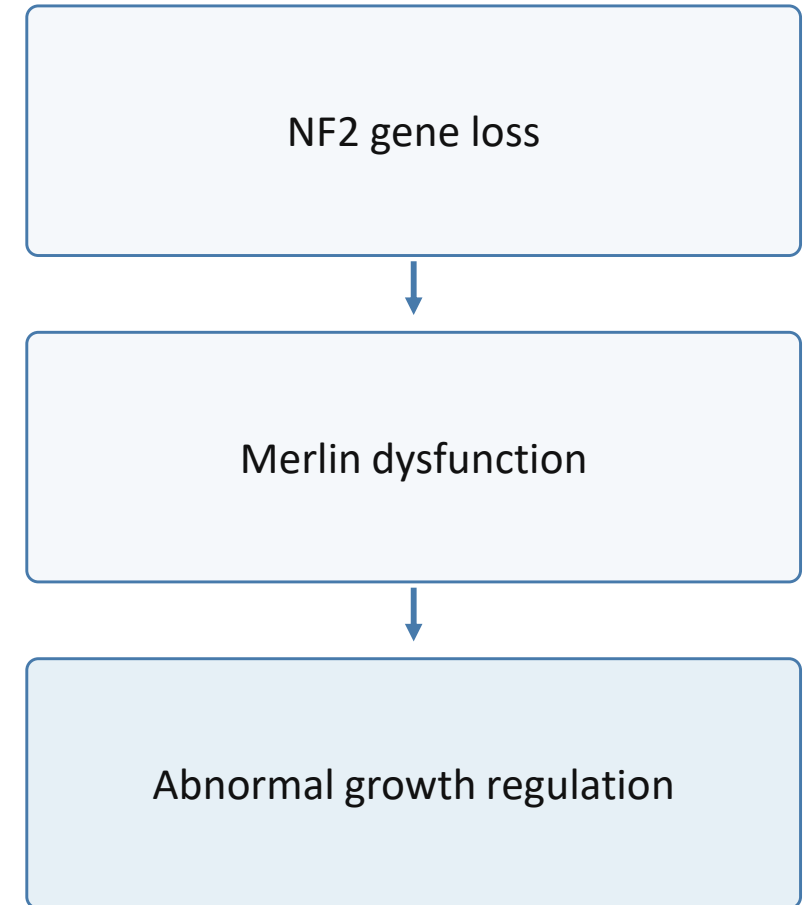
NF2 has a distinct tumor distribution

- Autosomal dominant syndrome occurring in approximately 1 in 40,000–50,000 people.
- Typical neoplasms: Vestibular schwannomas, meningiomas and spinal ependymomas.
- Neurofibromas are not the defining tumor of NF2.



NF2 encodes the cytoskeletal regulator merlin

- NF2 maps to 22q12 and encodes merlin.
- Merlin participates in signaling controlling cell growth, shape and adhesion.
- Somatic NF2 alterations also occur in sporadic schwannomas, meningiomas and spinal ependymomas.



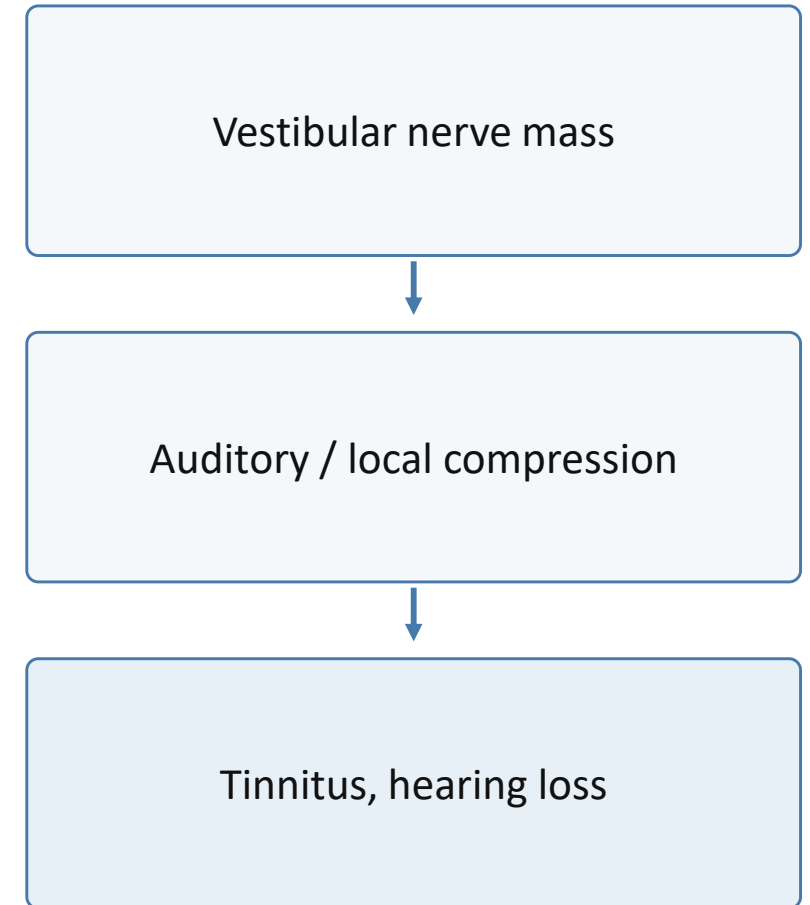
Bilateral vestibular schwannomas suggest NF2

- The vestibular division of cranial nerve VIII is a characteristic tumor origin.
- Bilateral lesions are a hallmark of the syndromic pattern.
- Associated intracranial and spinal tumors strengthen the interpretation.

- Right CN VIII
- Left CN VIII
- Other CNS tumors

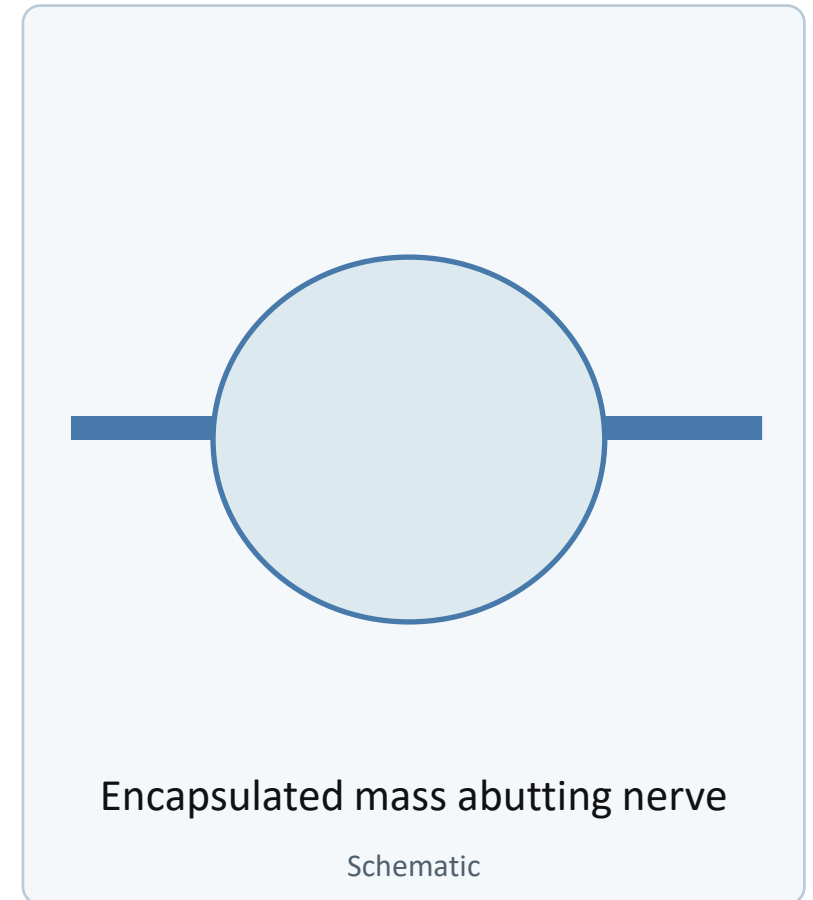
Vestibular schwannoma causes symptoms by location

- Tumors commonly arise near the cerebellopontine angle.
- Tinnitus and hearing loss may be presenting symptoms.
- Larger tumors may compress neighboring CNS structures.



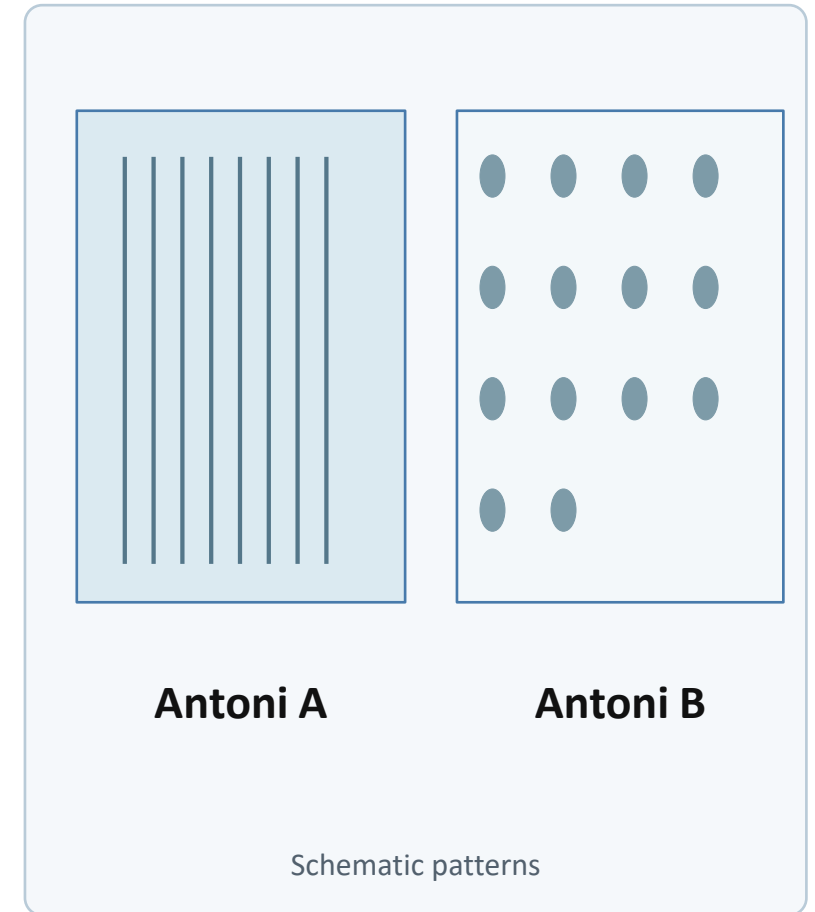
Schwannomas grow beside rather than through the nerve

- Most are circumscribed and encapsulated, attached loosely to the associated nerve.
- They often produce a firm gray mass.
- This pattern contrasts with fascicular infiltration by plexiform neurofibroma.



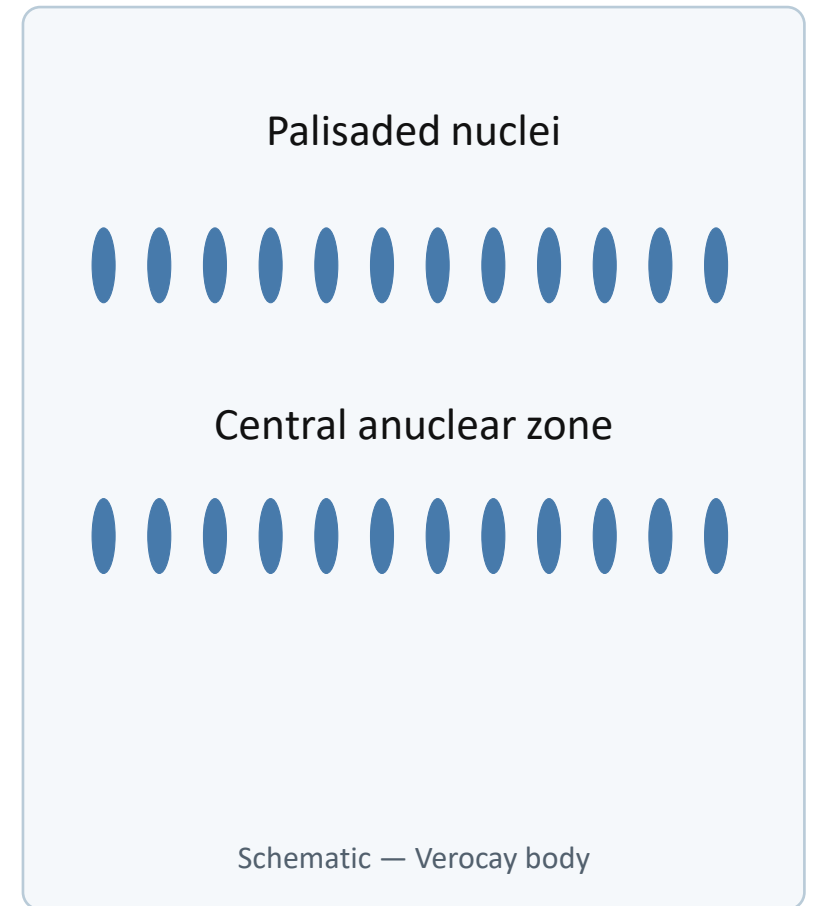
Antoni A and Antoni B are complementary patterns

- Antoni A areas are cellular and architecturally organized.
- Antoni B areas are looser, more myxoid and less structured.
- Their juxtaposition is a characteristic histologic feature of schwannoma.



Verocay bodies arise from nuclear palisading

- Aligned Schwann-cell nuclei create palisading rows in Antoni A tissue.
- A central anuclear zone separates the parallel rows.
- The pattern is named a Verocay body.



Schwann-cell lineage markers aid interpretation

- Schwannomas typically express S100 and SOX10.
- Continuous basement membrane material surrounds tumor cells.
- Marker expression supports lineage but must be interpreted with architecture.

S100 / SOX10

Schwann differentiation

**Basement
membrane**

Continuous pericellular
pattern

Architecture

Antoni A / B, palisading

Degenerative atypia does not automatically mean malignancy

- Schwannomas may show pleomorphic nuclei, hyalinized vessels and cystic change.
- Xanthomatous change and necrosis are also described.
- Interpret atypical changes alongside mitotic activity, architecture and clinical behavior.

Degenerative schwannoma

Hyalinization

Cystic / nuclear
change

Malignant concern

Invasive growth

Malignant cytology

Schwannomatosis differs from NF2 by tumor distribution

- The textbook characterizes non-NF2 schwannomatosis by multiple nonvestibular schwannomas.
- Distribution may be generalized or restricted to one body region.
- Cases may be familial or sporadic.

NF2 pattern

Vestibular
schwannomas

Meningioma /
ependymoma

Schwannomatosis pattern

Nonvestibular
schwannomas

Generalized or
localized

Three diagnoses, three nerve sheath patterns

- NF1 chiefly links neurofibromas with pigmentary and optic pathway findings.
- NF2 prominently links vestibular schwannomas to additional CNS neoplasms.
- Schwannomatosis centers on multiple nonvestibular schwannomas.

NF1

Neurofibromas + optic
glioma

NF2

Vestibular schwannomas

**Schwannomatos
is**

Nonvestibular
schwannomas

Neurofibroma versus schwannoma: Read the anatomy

- Neurofibroma: Mixed cellular composition and variable nerve infiltration.
- Schwannoma: Circumscribed, usually encapsulated, predominantly Schwann cells.
- Antoni A/B and Verocay bodies favor schwannoma over neurofibroma.

Neurofibroma

Mixed cells

Often
unencapsulated

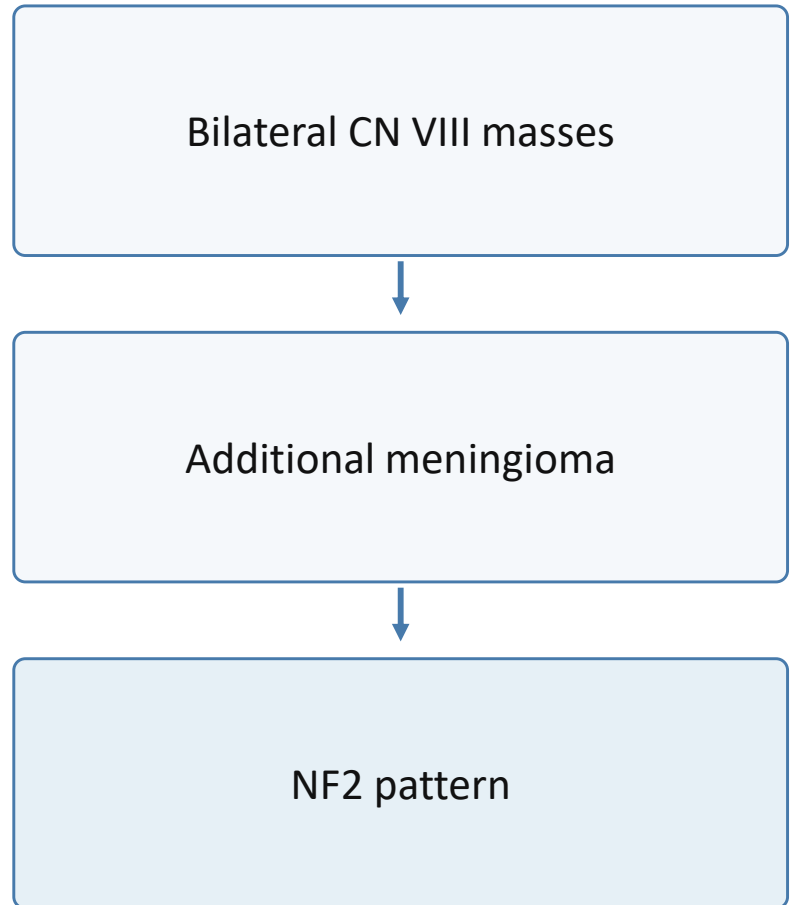
Schwannoma

Schwann cells

Antoni A/B

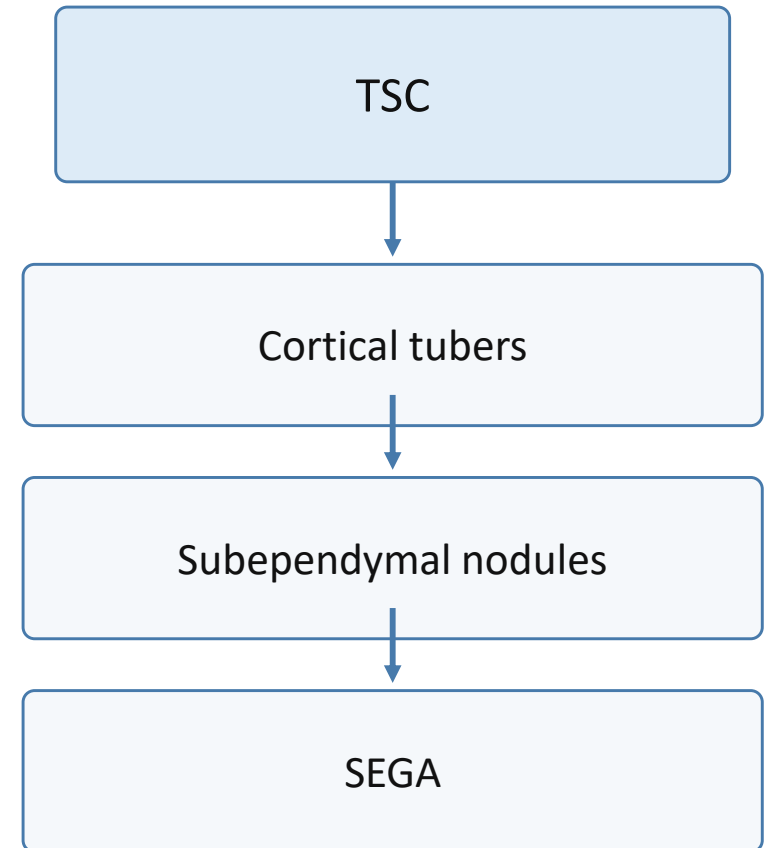
Clinical application: Bilateral VIII-nerve tumors

- A young patient has bilateral vestibular nerve masses and a meningioma.
- The lesion constellation supports NF2 rather than isolated sporadic schwannoma.
- The strongest clue is distribution, not a single slide's histology.



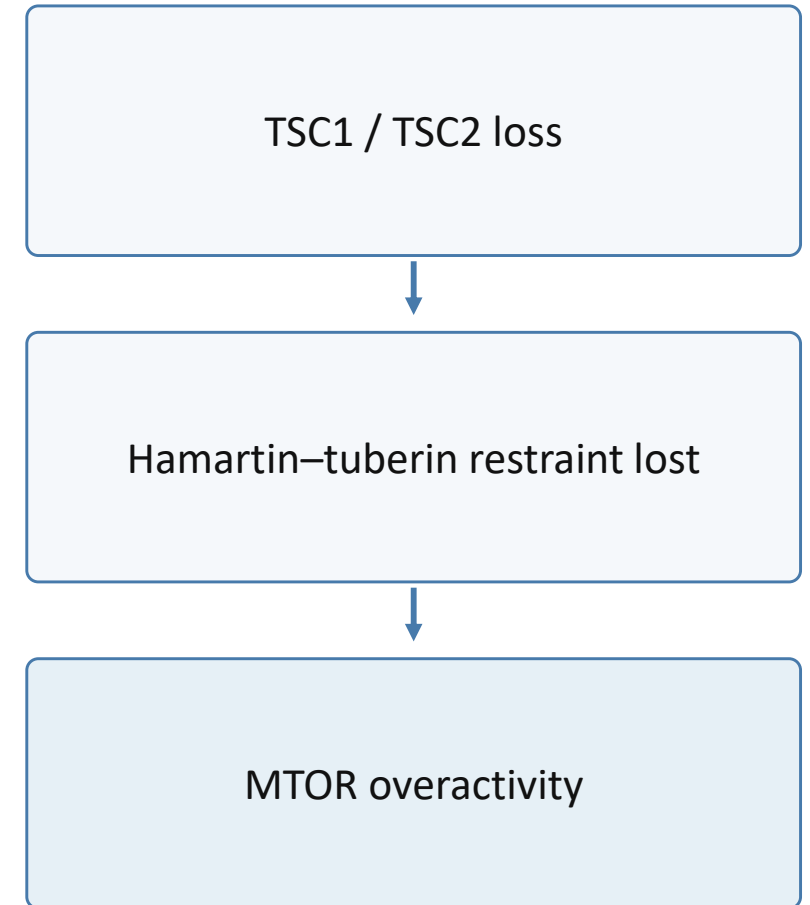
TSC unites developmental lesions and neoplasia

- Autosomal dominant disorder affecting roughly 1 in 6,000 births.
- CNS manifestations include cortical tubers, subependymal nodules and SEGA.
- Systemic hamartomas and benign neoplasms involve several other organs.



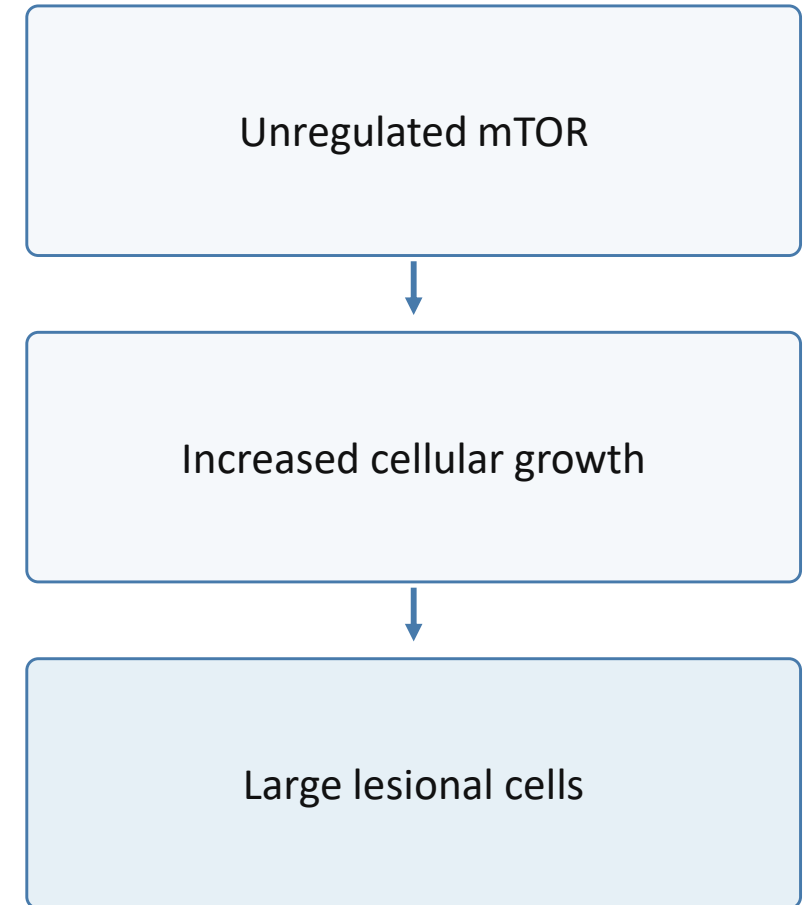
Hamartin and tuberin restrain mTOR activity

- TSC1 at 9q34 encodes hamartin; TSC2 at 16p13.3 encodes tuberin.
- Their protein complex suppresses mTOR signaling.
- Loss of complex function permits excessive anabolic and growth signaling.



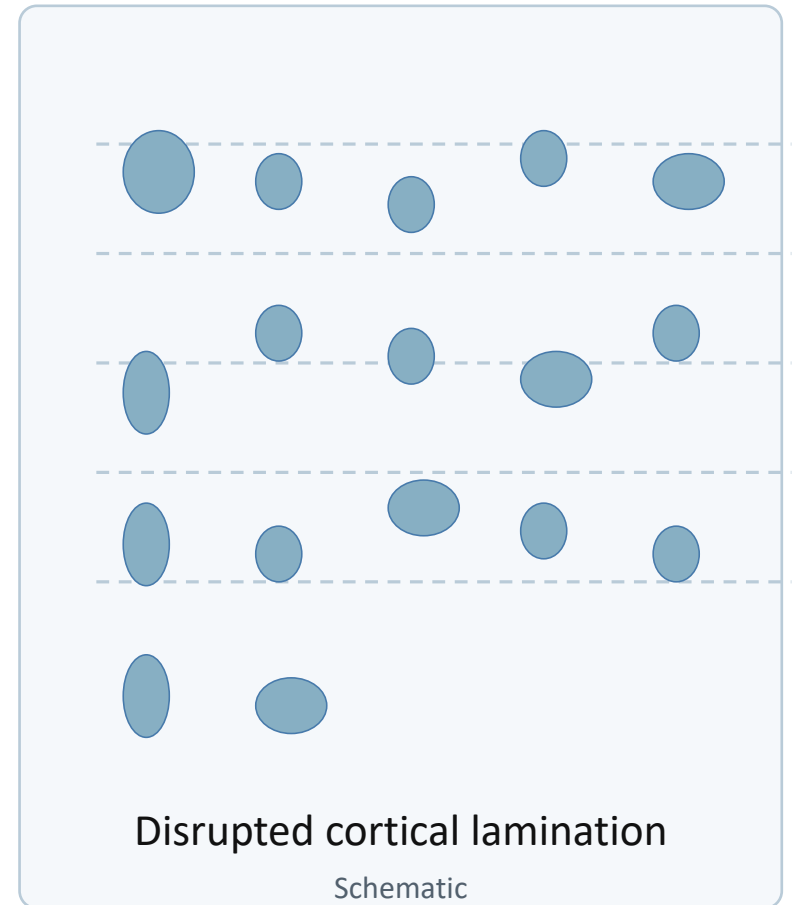
MTOR dysregulation explains enlarged lesional cells

- MTOR regulates protein synthesis and cell size.
- Increased mTOR activity contributes to cell growth and hamartomatous lesions.
- SEGA cells frequently display strikingly voluminous cytoplasm.



Cortical tubers disturb normal cortical architecture

- Cortical hamartomas form firm, disorganized regions (“tubers”).
- Neurons lose the orderly laminar arrangement of normal cortex.
- This disturbed architecture contributes to epileptogenic activity.



Cortical tubers: Histology is disorganization

- Haphazard neuronal distribution replaces normal layered cortical structure.
- Abnormal large cells can combine glial and neuronal features.
- Recognition depends on architecture as well as lesional cell cytology.

Normal cortex

Organized laminae

Ordered neurons

Cortical tuber

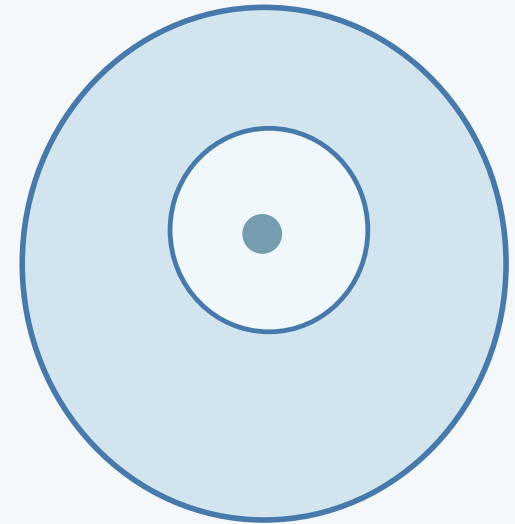
Disordered
laminae

Dysplastic giant
cells

Dysplastic cells show mixed neuronal and glial traits

- Large vesicular nuclei with nucleoli can resemble neurons.
- Abundant eosinophilic cytoplasm can resemble gemistocytic astrocytes.
- Cells may express both neurofilament and GFAP.

Large vesicular nucleus



Abundant cytoplasm

Neurofilament + GFAP expression

Schematic — not a histologic specimen

Cortical tubers explain common neurological findings

- Seizures are prominent and tubers can act as seizure foci.
- Autism and intellectual disability are frequent clinical manifestations.
- In difficult epilepsy, resection of an epileptogenic tuber may help.

Cortical structural abnormality



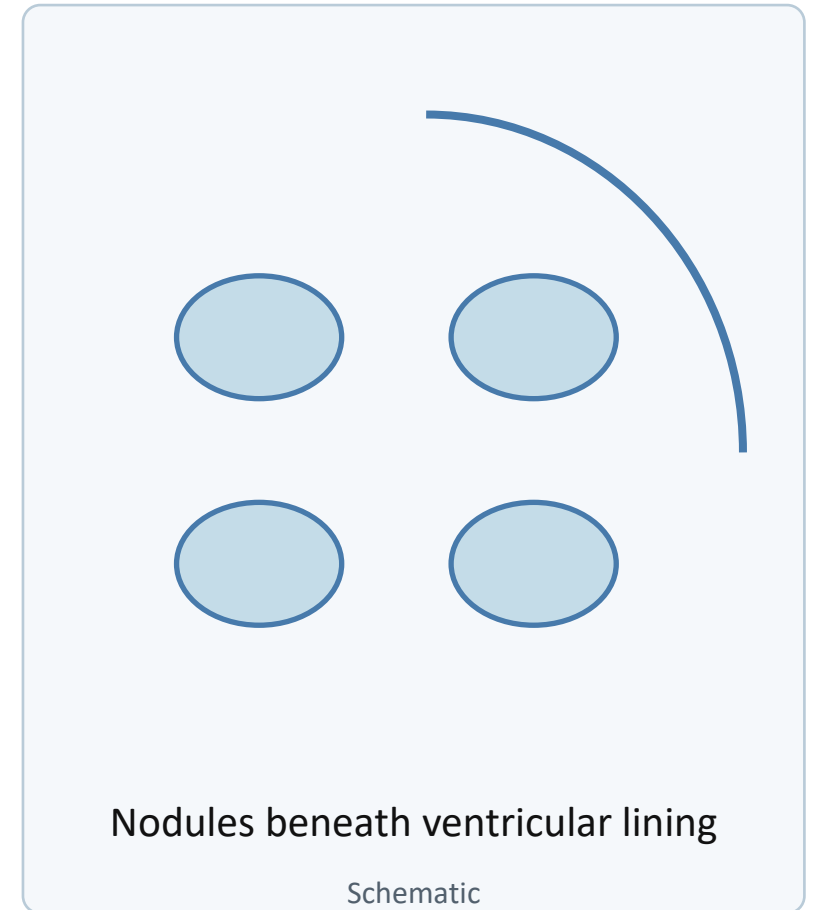
Abnormal network excitability



Seizure phenotype

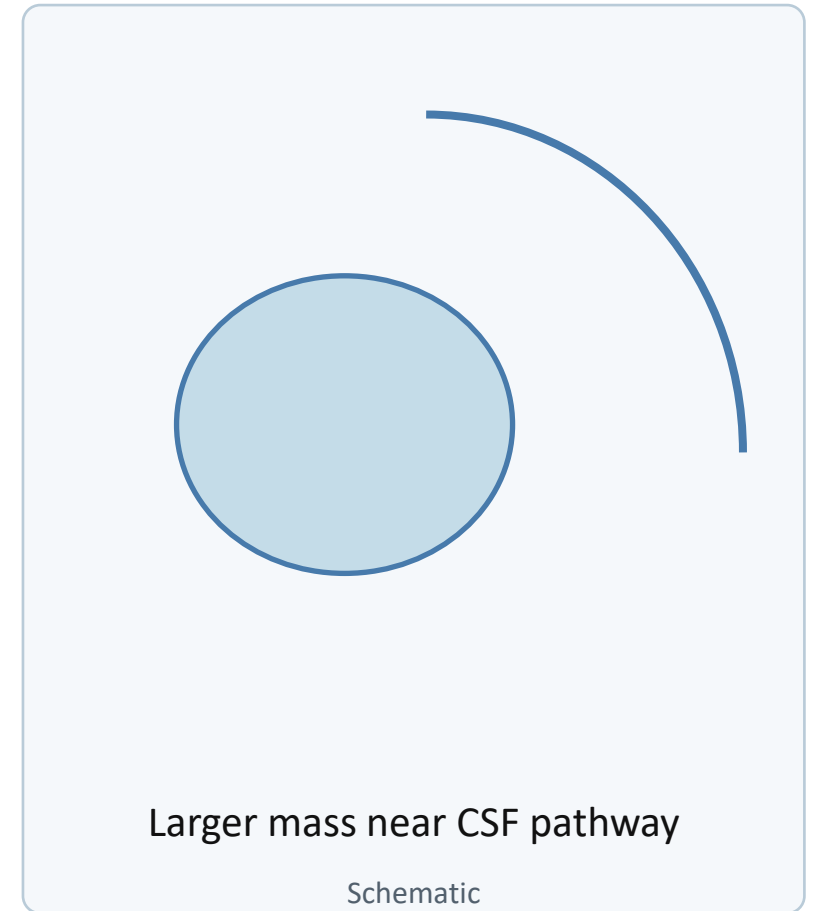
Subependymal nodules border the ventricle

- Hamartomatous nodules lie beneath the ependymal surface.
- Large astrocyte-like cells form clustered subependymal lesions.
- Protrusion into the ventricle creates the “candle gutterings” appearance.



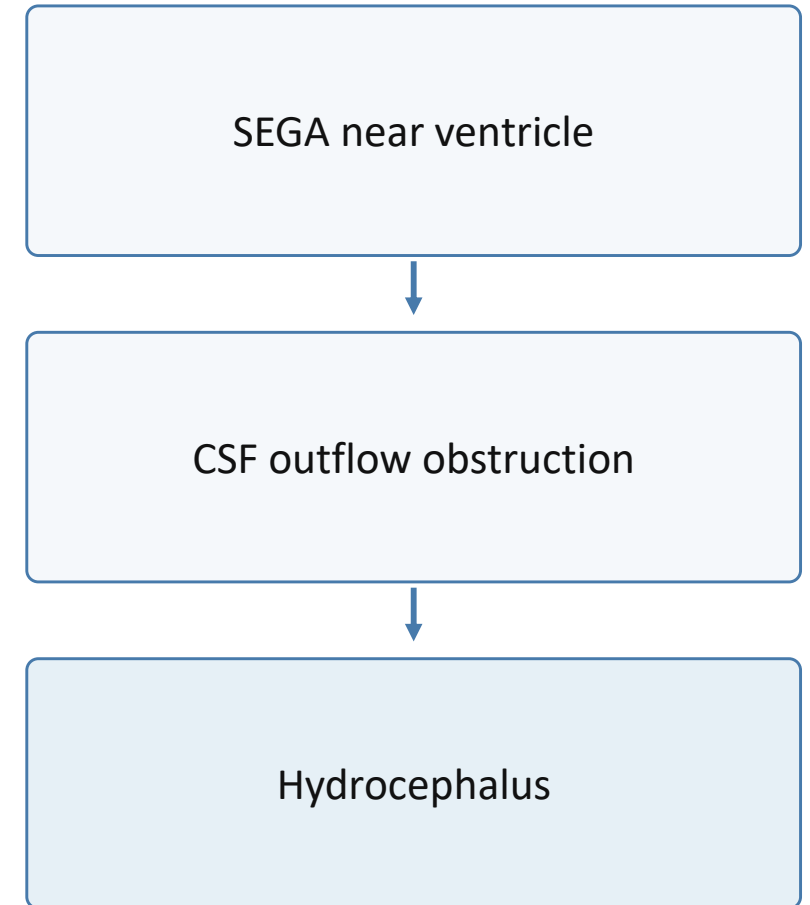
SEGA is a larger related subependymal tumor

- Subependymal giant cell astrocytoma resembles the cellular pattern of nodules.
- The mass is larger and behaves as a benign neoplasm.
- Its clinical importance lies particularly in ventricular obstruction.



How SEGA produces obstructive hydrocephalus

- A subependymal mass may narrow a ventricular CSF pathway.
- Obstruction leads to upstream ventricular enlargement.
- Clinical impact reflects location even without high-grade cytology.



Systemic hamartomas offer diagnostic corroboration

- Renal angiomyolipomas and retinal glial hamartomas are recognized associations.
- Cardiac rhabdomyomas and pulmonary lymphangioleiomyomatosis may occur.
- Cysts may involve kidney, pancreas or liver.

Kidney

Angiomyolipoma; Cysts

Retina / heart

Glial hamartoma;
Rhabdomyoma

Lung

Lymphangioleiomyomatosis

Liver / pancreas

Cysts

The skin provides high-yield syndrome clues

- Facial angiofibromas are recognized cutaneous lesions.
- Hypopigmented “ash-leaf” macules and shagreen patches are characteristic.
- Subungual fibromas may also occur.

Pigment

Ash-leaf macules

Dermis

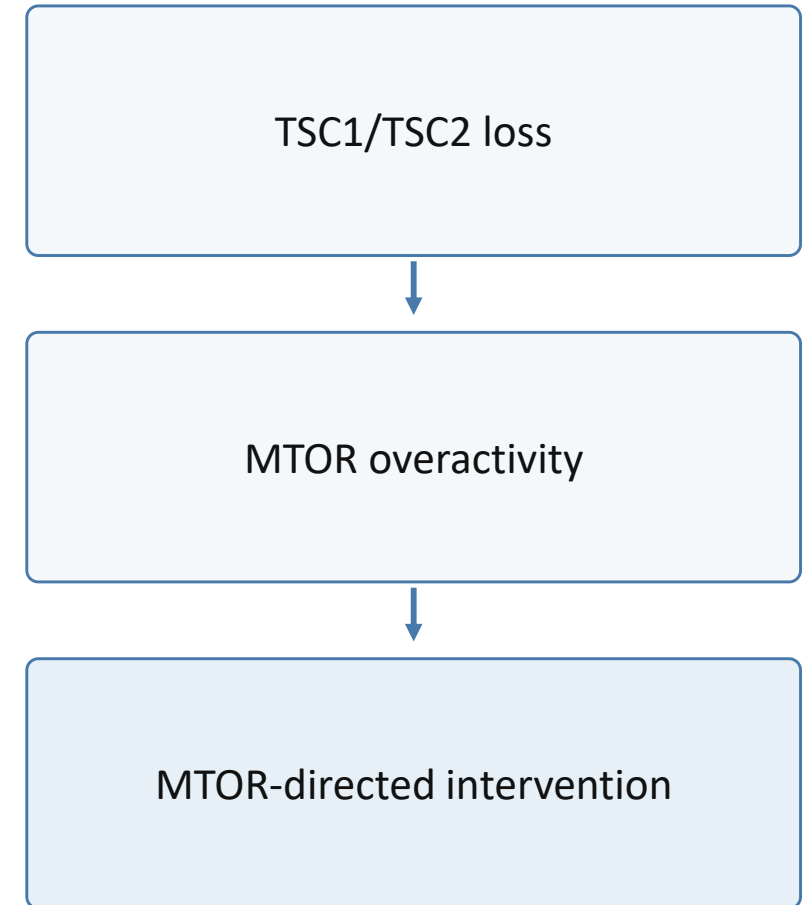
Angiofibromas; Shagreen patches

Nail region

Subungual fibromas

MTOR inhibitors target the causal signaling axis

- Conventional treatment includes anticonvulsants for seizure control.
- MTOR inhibitors can reduce the consequences of pathway hyperactivity.
- Selected patients may show marked improvement with pathway-directed therapy.



Different lesions create different clinical problems

- Cortical tuber: Network dysfunction and refractory epilepsy.
- Subependymal nodule: Often a recognizable ventricular hamartoma.
- SEGA: Larger mass with potential CSF-flow obstruction.

Cortical tuber

Epileptogenic cortex

**Subependymal
nodule**

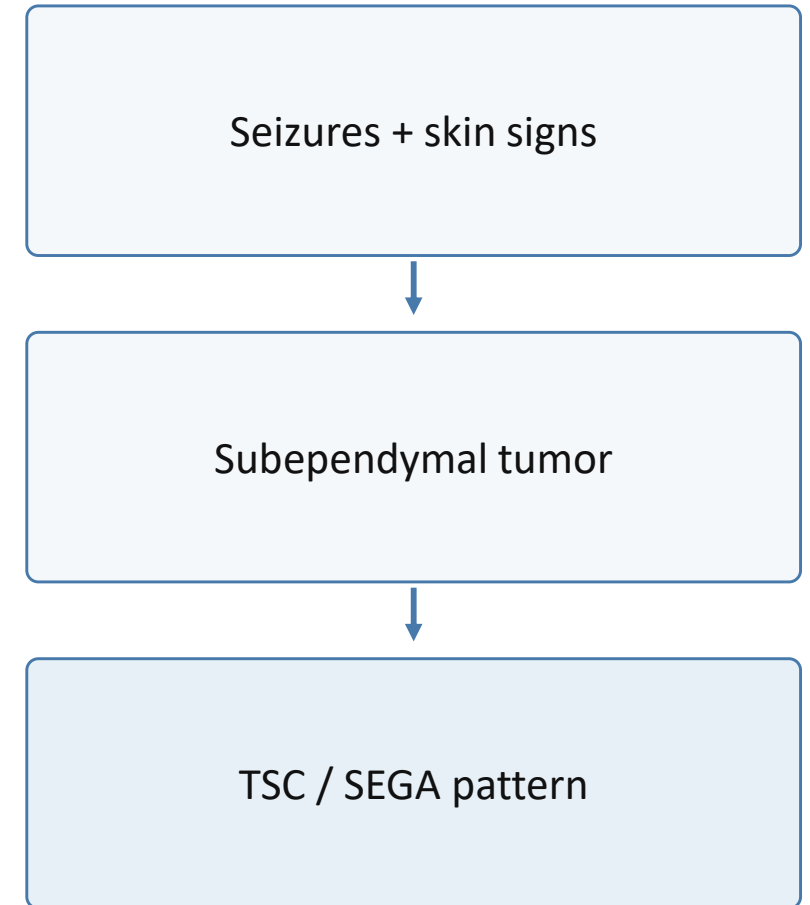
Ventricular hamartoma

SEGA

Obstructive mass effect

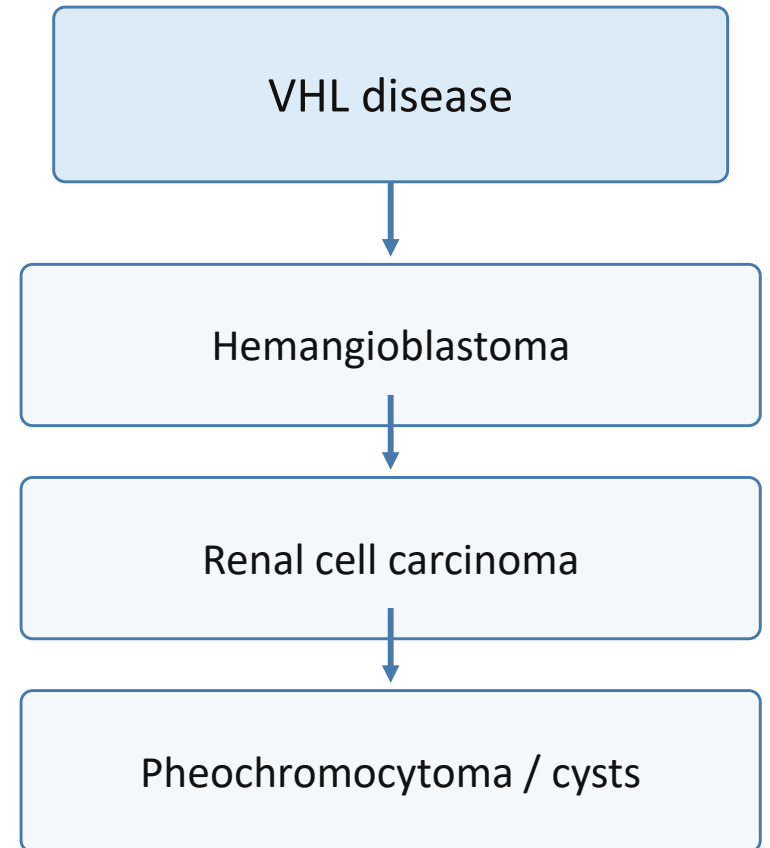
Clinical application: Epilepsy with ventricular lesion

- A patient has seizures, hypopigmented macules and a ventricular mass.
- The combined phenotype raises TSC with possible SEGA.
- Assess whether ventricular enlargement suggests an obstructive consequence.



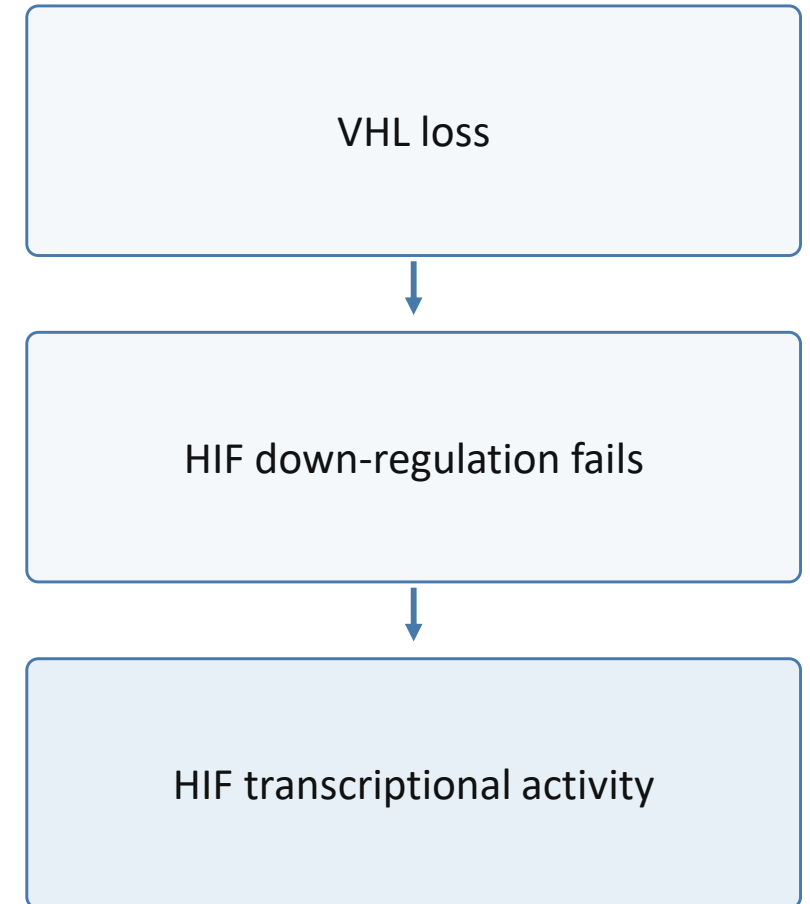
VHL links vascular CNS neoplasia to systemic tumors

- Autosomal dominant syndrome present in roughly 1 in 30,000–40,000 people.
- Hemangioblastomas favor the cerebellum and retina.
- Renal cell carcinoma, pheochromocytoma and visceral cysts also occur.



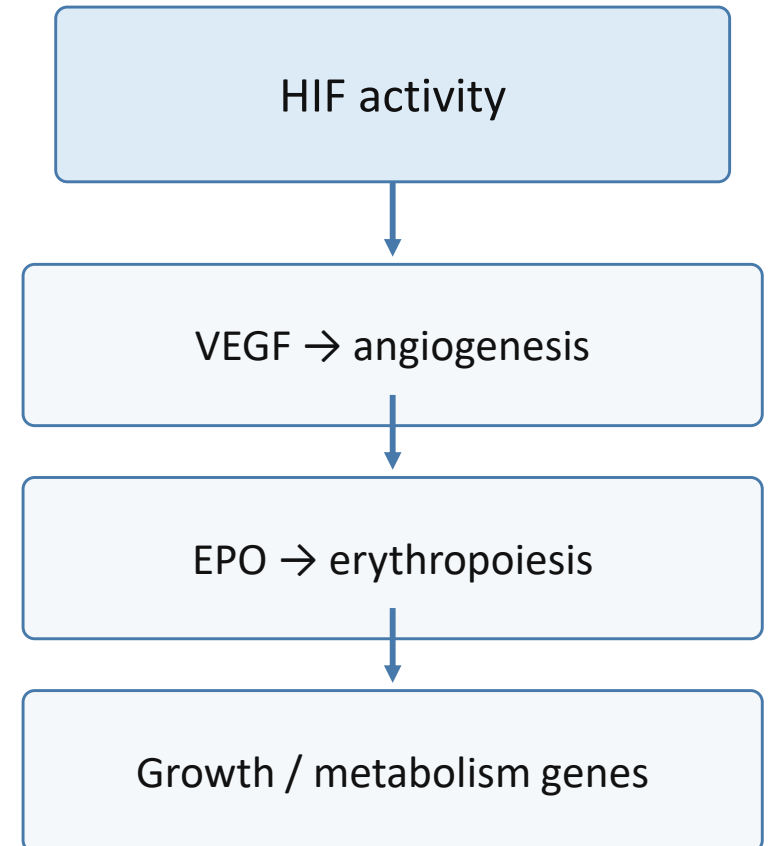
VHL is a tumor suppressor at chromosome 3p25.3

- VHL protein participates in a ubiquitin ligase complex.
- The complex normally down-regulates hypoxia-inducible factors (HIFs).
- Loss of VHL function leads to inappropriate persistence of HIF-dependent signaling.



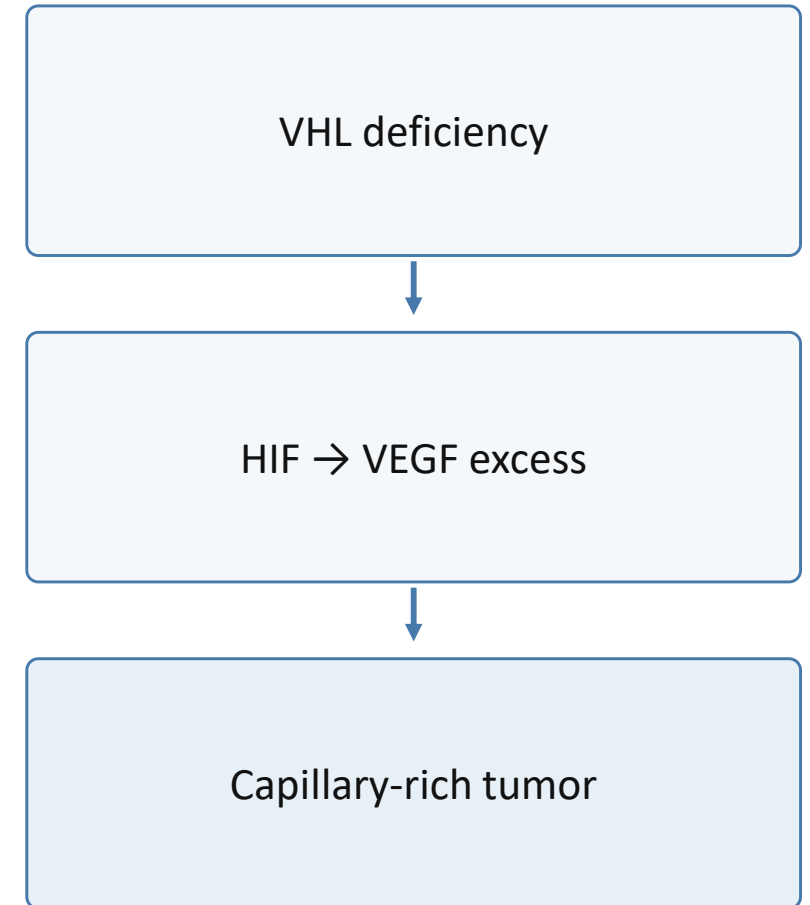
HIF drives an angiogenic and metabolic program

- HIFs control expression of VEGF, erythropoietin and other factors.
- VEGF increases vascular growth, contributing to hemangioblastomas.
- HIF also regulates genes involved in cellular metabolism and growth.



VEGF explains the dense vascularity of hemangioblastoma

- Excess VEGF promotes extensive capillary formation.
- The lesional vasculature consists of numerous thin-walled vessels.
- Highly vascular morphology is therefore mechanistically linked to VHL loss.



Erythropoietin can explain secondary polycythemia

- Hemangioblastoma stromal cells may express erythropoietin.
- Excess erythropoietin stimulates red-cell production.
- Polycythemia in about 10% of hemangioblastoma patients.

Tumor-derived erythropoietin



Increased red-cell production



Polycythemia

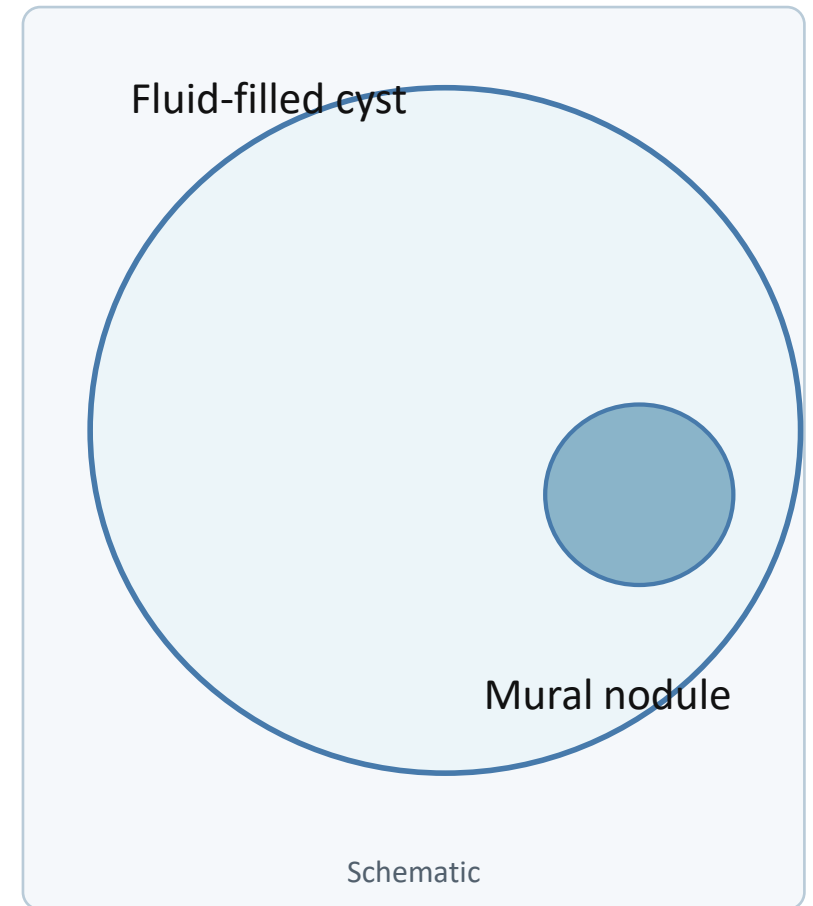
Hemangioblastoma has a characteristic distribution

- The cerebellum is a common CNS location.
- Retinal hemangioblastomas are another major syndrome manifestation.
- Other CNS locations may also be involved.

- Cerebellum
- Retina
- Other CNS sites

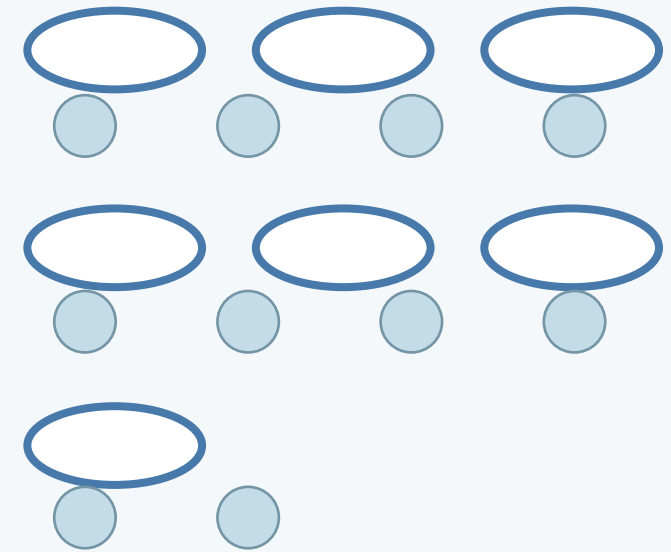
Hemangioblastoma may form a cyst with a mural nodule

- A solid vascular nodule can lie along the wall of a large fluid-filled cyst.
- The cyst can dominate gross lesion size and contribute to mass effect.
- Recognition of the cyst–nodule configuration helps connect imaging and gross pathology.



Hemangioblastoma histology: Capillaries plus stromal cells

- Numerous thin-walled capillary-sized vessels traverse the lesion.
- Intervening neoplastic stromal cells have vacuolated, lipid-rich cytoplasm.
- The mixed vascular–stromal architecture is a key diagnostic clue.



Thin vessels + lipid-rich stromal cells

Schematic — not a histologic specimen

Inhibin is a useful stromal-cell marker

- The neoplastic stromal component expresses inhibin.
- The same cells may express VEGF and erythropoietin.
- Diagnostic interpretation must distinguish stromal cells from the prominent vasculature.

Stromal cells

Lipid-rich
cytoplasm

Inhibin expression

Vascular component

Thin-walled
capillaries

Angiogenic
response

Systemic tumor spectrum distinguishes VHL

- Renal cell carcinomas and renal cysts are important extracranial lesions.
- Pheochromocytomas are part of the syndrome.
- Cysts may involve pancreas and liver as well as kidney.

CNS / retina

Hemangioblastomas

Kidneys

RCC; Cysts

Adrenal

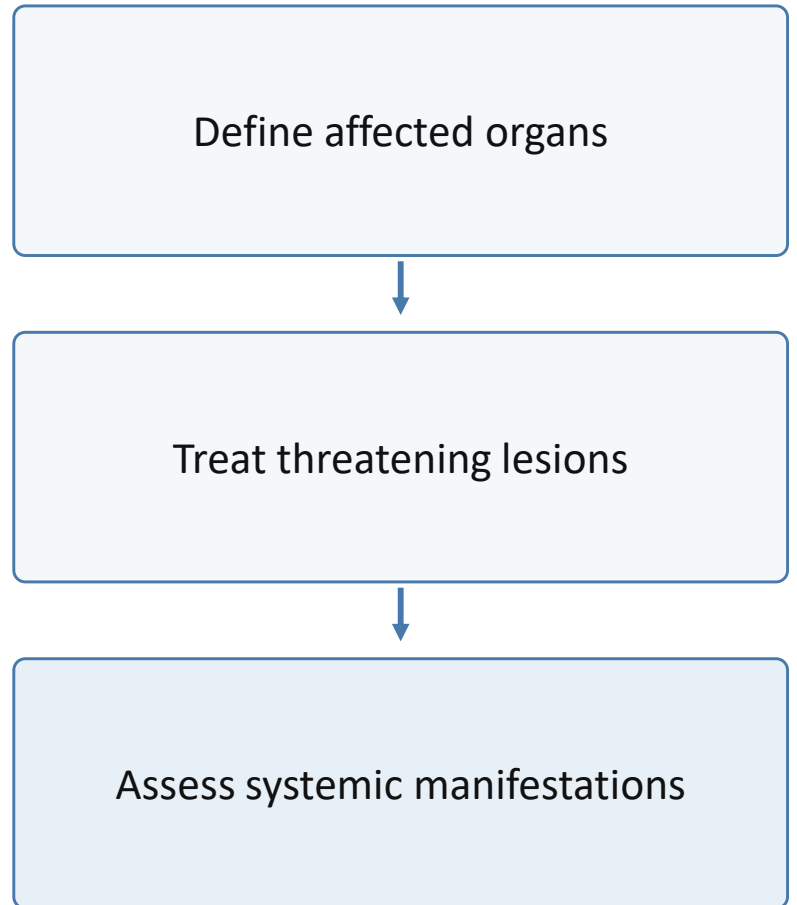
Pheochromocytoma

Viscera

Pancreatic / hepatic cysts

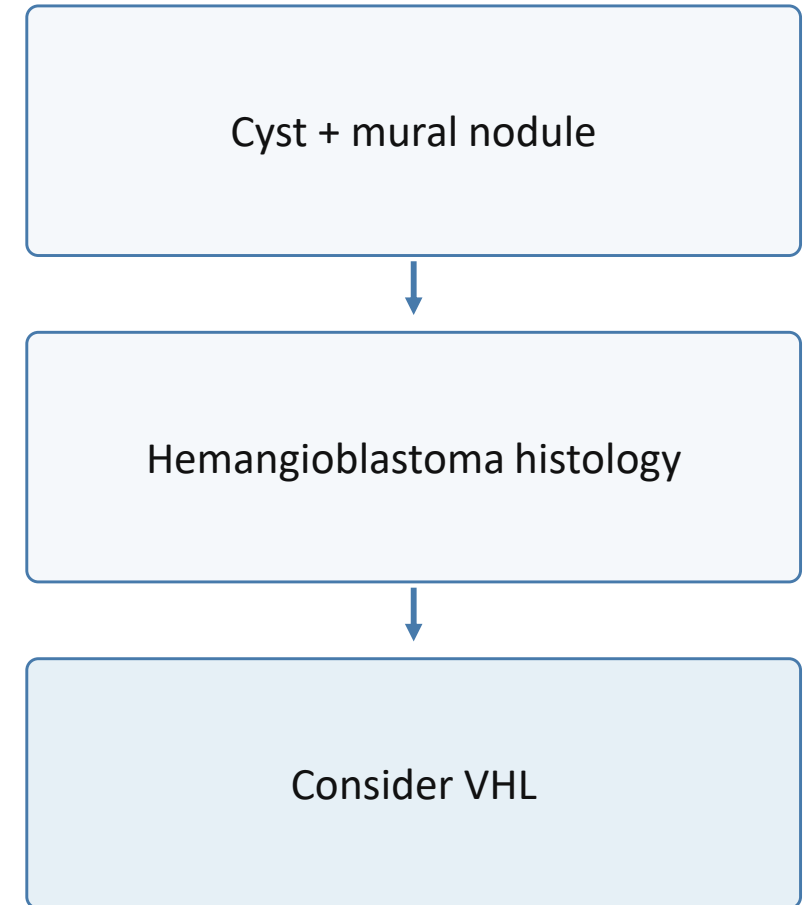
Hemangioblastoma care is lesion-directed

- Symptomatic cerebellar lesions may be surgically resected.
- Retinal hemangioblastomas can be treated with laser methods.
- Systemic evaluation is essential because the predisposition affects several organs.



Clinical application: Vascular cerebellar mural nodule

- A cystic cerebellar mass contains a mural nodule rich in capillaries.
- Vacuolated stromal cells and inhibin expression support hemangioblastoma.
- The associated syndrome to consider is VHL, especially with visceral lesions.



Each syndrome disrupts a different cell-growth control

- NF1: RAS activation persists without neurofibromin restraint.
- NF2: Merlin-dependent growth and adhesion control is disturbed.
- TSC: MTOR signaling is overactive.
- VHL: HIF-driven angiogenic signaling is excessive.

NF1

Neurofibromin → RAS

NF2

Merlin →
growth/adhesion

TSC

Hamartin/tuberin →
mTOR

VHL

VHL → HIF/VEGF

Tumor phenotype provides a practical syndrome index

- Plexiform neurofibroma strongly favors NF1.
- Bilateral vestibular schwannomas strongly favor NF2.
- SEGA with cortical tubers favors TSC.
- Cystic vascular cerebellar tumor with mural nodule favors VHL.

**Plexiform
neurofibroma**

NF1

**Bilateral
vestibular
schwannoma**

NF2

**SEGA + cortical
tubers**

TSC

**Hemangioblasto
ma**

VHL

Location can matter as much as cell phenotype

- A benign nerve sheath tumor can impair cranial nerve VIII function.
- A benign SEGA can obstruct a ventricle.
- A cerebellar vascular lesion can produce clinically important compression.

- CN VIII / hearing
- Ventricle / CSF
- Cerebellum / compression

Morphologic differentials rely on architecture

- Schwannoma: Circumscription, Antoni A/B architecture and Verocay bodies.
- Neurofibroma: Mixed cells and variable infiltration through nerve structures.
- Hemangioblastoma: Capillary-rich stroma with vacuolated neoplastic cells.

Schwannoma

Antoni A/B

Neurofibroma

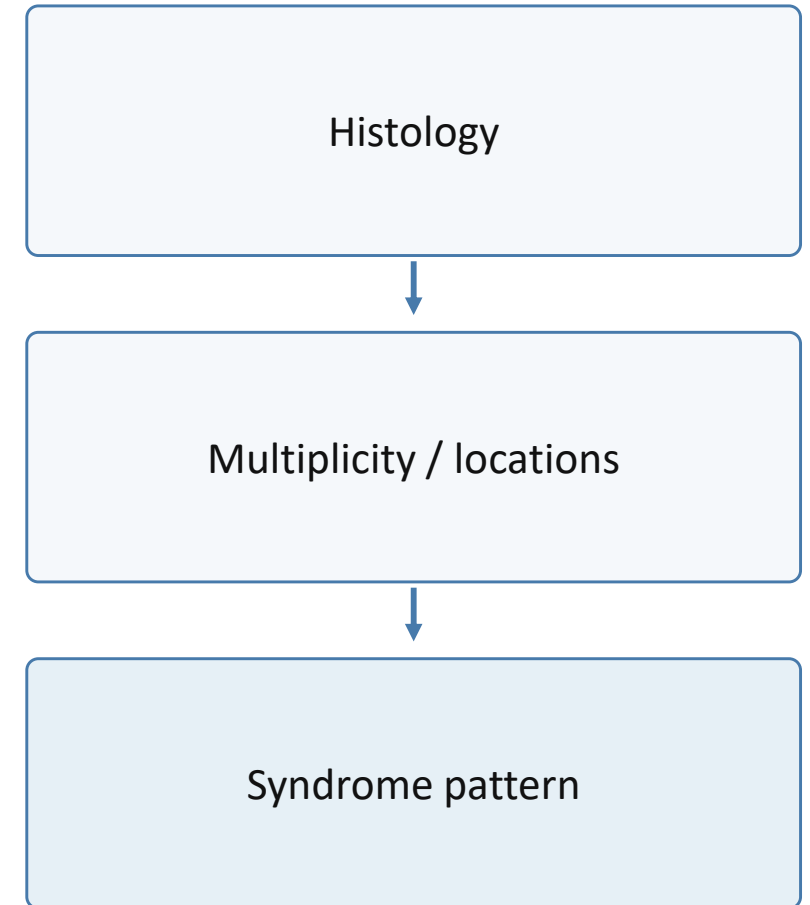
Mixed nerve sheath cells

Hemangioblastoma

Vessels + lipid-rich stromal cells

A diagnostic sequence for multifocal nervous-system tumors

- First identify a dominant tumor type and its histologic architecture.
- Next map its anatomic distribution and count synchronous lesions.
- Then seek the characteristic skin, eye and systemic clues.



Case comparison: Pigmentary signs or bilateral vestibular masses?

- Patient A: Café-au-lait macules, Lisch nodules and a plexiform neurofibroma.
- Patient B: Bilateral vestibular schwannomas with a meningioma.
- Distinctive associated features distinguish NF1 from NF2.

Patient A

Pigmentary +
plexiform

NF1

Patient B

Bilateral vestibular

NF2

Case comparison: SEGA or hemangioblastoma?

- Patient C: Cortical tubers, seizures and a subependymal ventricular tumor.
- Patient D: Cerebellar cyst with a highly vascular mural nodule.
- Cellular and anatomic patterns favor TSC and VHL, respectively.

Patient C

Tuber + ventricular
mass

TSC

Patient D

Cystic vascular
mass

VHL

Four high-yield pathology-to-mechanism links

- NF1 neurofibromas: Biallelic neurofibromin loss in neoplastic Schwann cells.
- NF2 schwannomas: Merlin pathway dysfunction in nerve sheath tumors.
- TSC giant cells: Loss of mTOR restraint favors enlargement.
- VHL hemangioblastoma: HIF/VEGF excess explains vascularity.

NF1

Schwann-cell tumor →
RAS

NF2

Schwannoma → merlin

TSC

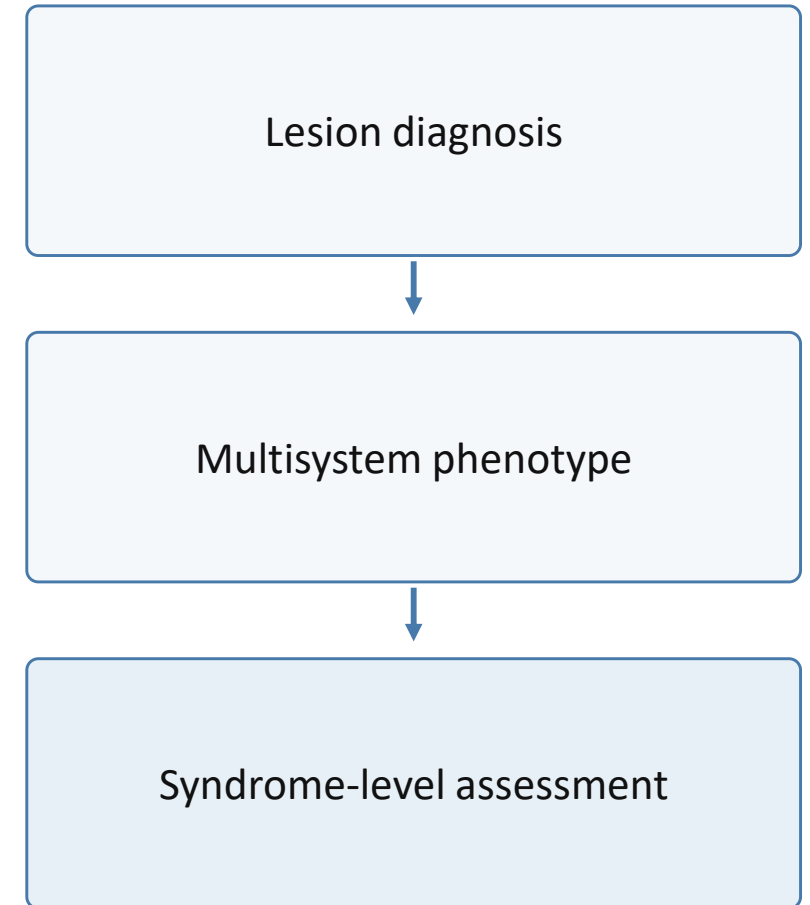
Giant cells → mTOR

VHL

Capillaries → HIF/VEGF

What pathology can and cannot establish alone

- Characteristic histology helps identify a lesion but may not prove hereditary disease.
- Multiple characteristic tumors and nonneoplastic findings raise syndrome-level confidence.
- Genetic diagnosis and clinical classification require appropriate contextual assessment.



Synthesis: Hereditary tumor syndromes of the nervous system

- Recognize each characteristic tumor distribution and its distinctive microscopy.
- Connect the lesion to NF1/RAS, NF2/merlin, TSC/mTOR or VHL/HIF signaling.
- Explain symptoms through lesion location, not grade alone.
- Use systemic manifestations to recognize a syndrome beyond a solitary tumor.

NF1

Neurofibroma; Optic glioma

NF2

Vestibular schwannoma

TSC

Cortical tuber; SEGA

VHL

Hemangioblastoma

Primary textbook source

Robbins, Cotran & Kumar: Pathologic Basis of Disease

11th edition (2026)

Chapter 28: The Central Nervous System — Familial Tumor Syndromes, pp. 1185–1186

Related context: Chapter 27, Peripheral Nerves and Skeletal Muscles, pp. 1125–1127