

Primary Cardiac Tumors

Morphology · mechanisms · clinicopathologic correlation

Benign neoplasms, clinically consequential masses, and rare sarcomas

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Learning objectives

- Classify the principal benign and rare malignant primary cardiac tumors.
- Recognize the defining gross and microscopic patterns of myxoma, papillary fibroelastoma, and rhabdomyoma.
- Link tumor location and architecture to obstruction, embolization, and other clinical effects.
- Explain the syndromic signaling abnormalities described for myxoma and rhabdomyoma.

Primary cardiac tumors are uncommon

Frequency

Primary cardiac tumors are uncommon in routine clinical practice.

Biology

The majority are benign rather than malignant.

Why they matter

A benign mass may still impair intracardiac blood flow or embolize.

Five common benign entities

Order	Primary cardiac tumor	Teaching discriminator
1	Myxoma	Most frequent in the combined population
2	Fibroma	Benign; Resembles extracardiac counterpart
3	Lipoma	Benign; Resembles extracardiac counterpart
4	Papillary fibroelastoma	Endocardial, often valvular
5	Rhabdomyoma	Leading pediatric primary tumor

Almost 90% belong to these five types

Five frequent benign entities



No malignant potential



**≈90% of primary cardiac
tumors**

Adult and pediatric leading entities differ

Adults: Myxoma

- Myxoma predominates in adult hearts.
- Commonly attached to the atrial septum.
- Atrial obstruction, embolization, and constitutional effects.

Children: Rhabdomyoma

- Rhabdomyoma is the leading pediatric type.
- Often multiple in ventricular myocardium.
- Associated with tuberous sclerosis in about half.

The compartment predicts the mechanism

Atrial cavity

A mobile intracavitary myxoma may obstruct an atrioventricular orifice.

Valve surface

Papillary fibroelastoma is often silent but can embolize.

Ventricular myocardium

Rhabdomyoma may project into the lumen and obstruct flow.

Benign histology does not ensure benign physiology

Benign intracardiac mass



Mechanical or embolic
complication



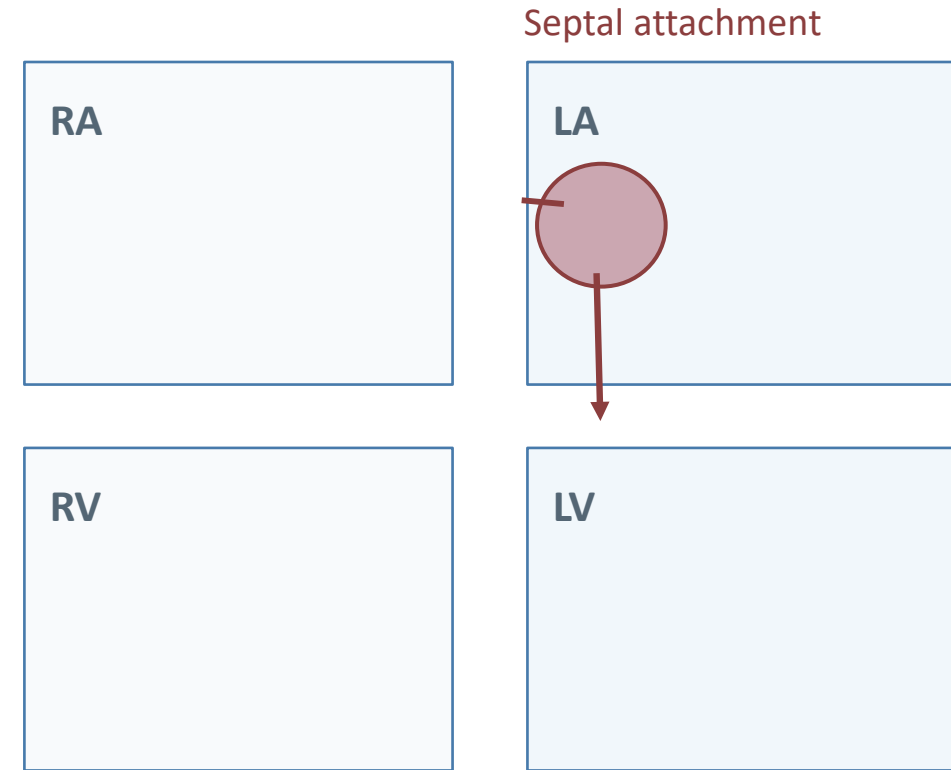
Clinically significant disease

Primary malignant cardiac tumors are exceptionally rare

- Angiosarcoma among the usual malignant histologic types.
- It also identifies undifferentiated sarcomas with MDM2 oncogene amplification.
- Cardiac sarcomas are not described as clinically or morphologically unique relative to sarcomas elsewhere.

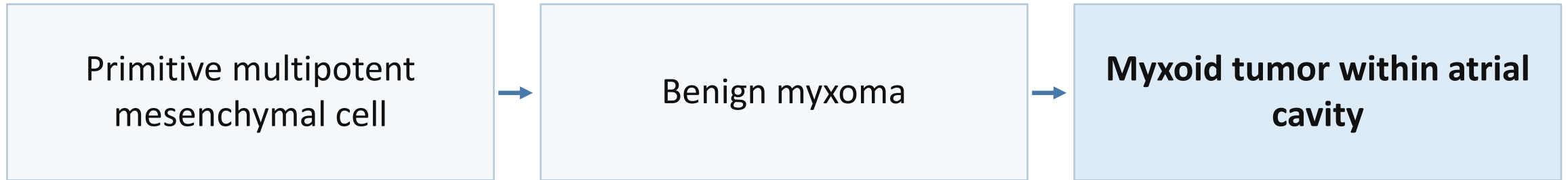
Myxoma is the most common adult primary tumor

- Benign neoplasm arising from primitive multipotent mesenchymal cells.
- Most often an atrial rather than ventricular lesion.
- Clinical significance reflects obstruction, embolization, or cytokine-associated effects.



Schematic · cardiac chambers

Myxoma arises from multipotent mesenchymal cells



Sporadic myxoma lacks a consistent genetic signature

- Sporadic myxomas have no consistent genetic alteration in the supplied Robbins section.
- This statement differs from the defined gene defects of recognized syndromic associations.
- Do not assume that every myxoma is syndromic from its histology alone.

Two recognized syndromic associations

McCune–Albright

- McCune–Albright syndrome
- Activating GNAS1 mutation
- Enhanced G-protein signaling

Carney complex

- Carney complex
- Loss-of-function PRKAR1A mutation
- Increased cAMP-dependent signaling

GNAS1: Gain of function enhances signaling



PRKAR1A: Loss of regulatory restraint

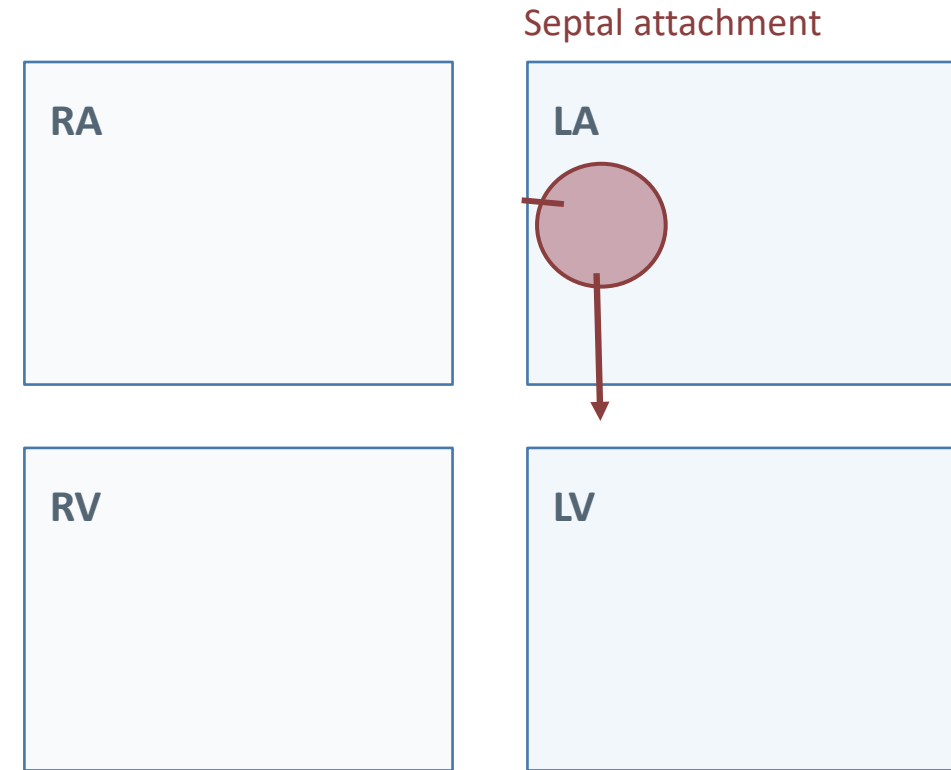


Syndromic myxoma: Contrast the direction

Syndrome	Gene	Alteration	Signaling consequence
McCune–Albright	GNAS1	Gain of function	Enhanced G-protein signaling
Carney complex	PRKAR1A	Loss of function	Increased cAMP-dependent signaling

Approximately 90% of myxomas are atrial

- Atria are overwhelmingly favored over ventricles.
- The location helps explain why an intracavitary lesion can involve a valve opening.
- The fossa ovalis region of the atrial septum is the favored origin.



Schematic · cardiac chambers

Left atrial myxoma is more frequent

Left atrial site

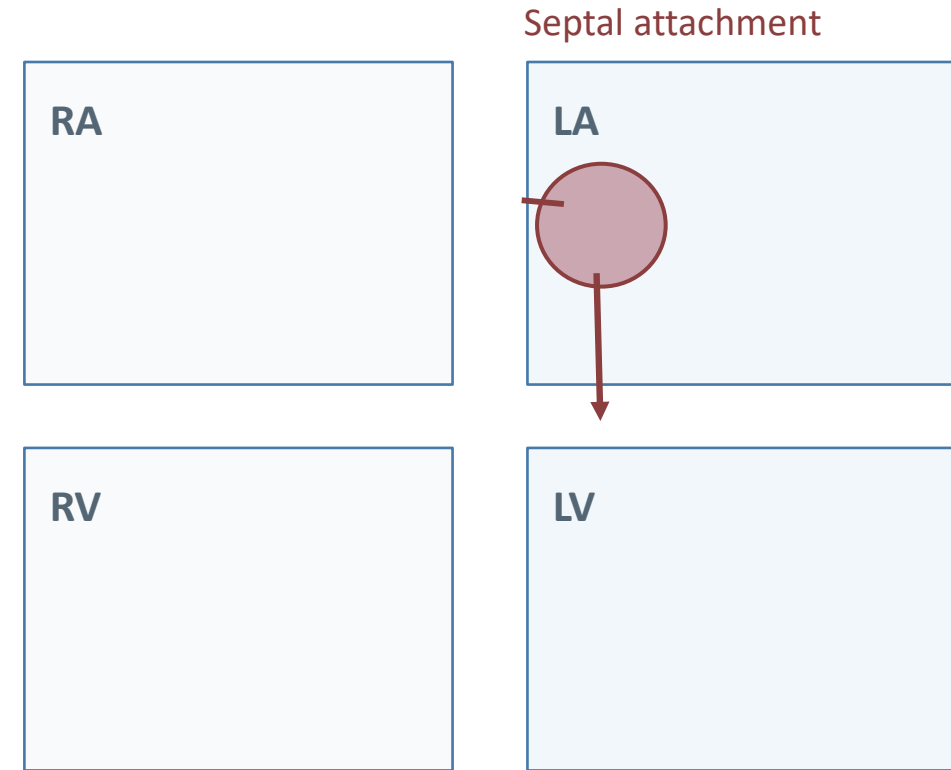
- Left atrium is the predominant atrial site.
- Often near the interatrial septum.
- May interfere with the mitral orifice.

Right atrial site

- Right atrium is less frequently involved.
- The approximate left:right atrial ratio is 4:1.
- Both sites fit the atrial predominance.

Fossa ovalis: The preferred attachment site

- Myxomas usually arise from the region of the fossa ovalis.
- This site lies in the interatrial septum.
- A lesion can remain septally attached while projecting into the atrial cavity.



Schematic · cardiac chambers

Myxomas are usually single

Usual pattern

- Most myxomas are solitary.
- A single endocardial attachment may anchor a relatively large mass.

Less common pattern

- Multiple tumors can occur, but are rare.
- The provided source does not assign a numerical multiplicity rate.

Myxoma size spans a very broad range

Small lesions

Some are under 1 cm in diameter.

Large lesions

Some reach 10 cm or more.

Clinical implication

Size and mobility together determine how a mass encounters an orifice.

Sessile versus pedunculated myxoma

Sessile

- Broad attachment at the origin.
- May form a large intracavitary mass.
- Robbins figure depicts a large sessile myxoma.

Pedunculated

- Attached by a stalk.
- Can move appreciably with the cardiac cycle.
- May intermittently enter an atrioventricular opening.

Gross myxomas have variable texture

Firm or globular

Some masses are relatively hard and mottled by hemorrhage.

Soft or gelatinous

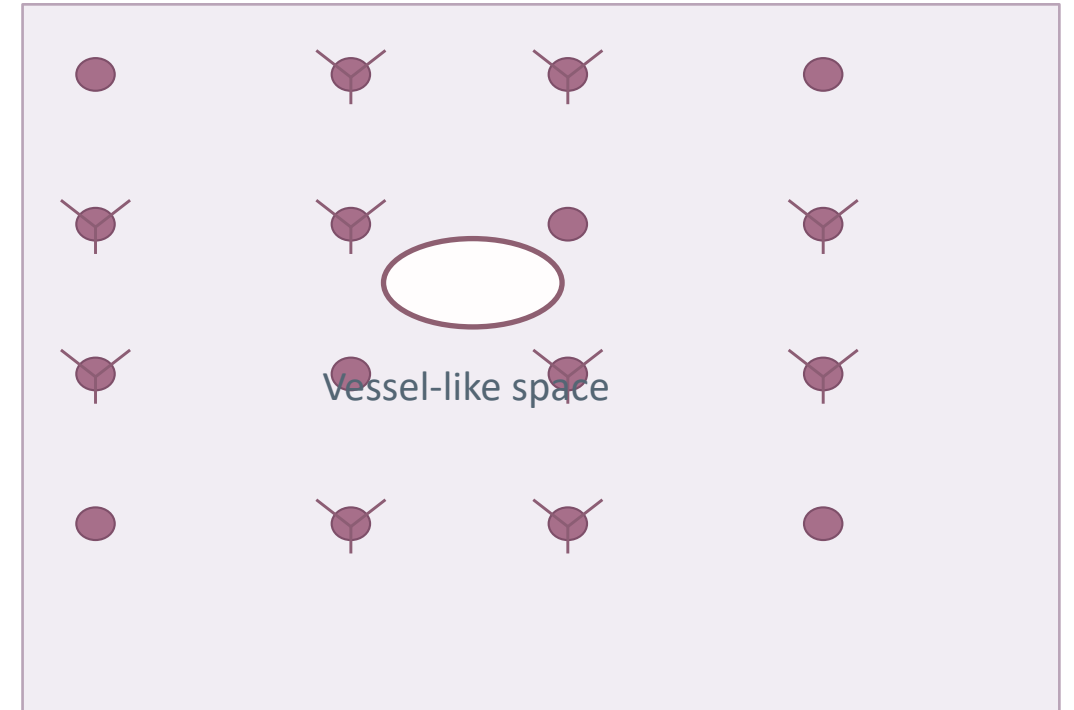
Other masses are translucent with a gelatinous quality.

Papillary or villous

Some lesions show surface projections rather than a smooth mass.

A gelatinous mass reflects abundant matrix

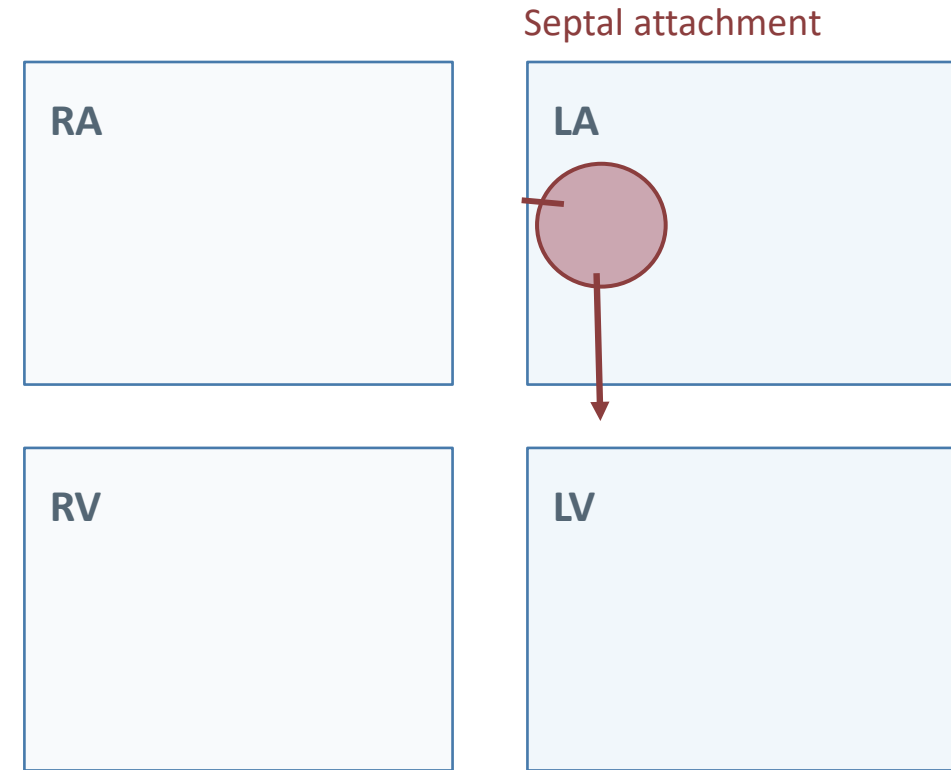
- The microscopic tumor contains abundant acid mucopolysaccharide-rich extracellular ground substance.
- The large matrix compartment accounts for much of the myxoid or gelatinous appearance.
- Cellular elements are relatively dispersed within this matrix.



Schematic · not an H&E specimen

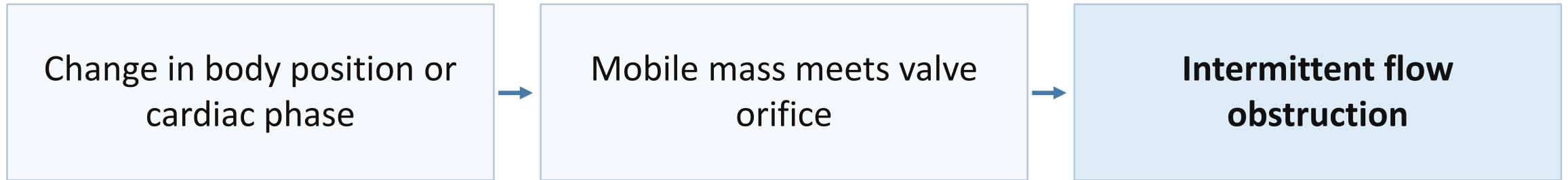
Mobile myxoma can create “ball-valve” obstruction

- A pedunculated mass may move into an atrioventricular valve opening.
- The orifice is transiently occupied by tumor tissue.
- Obstruction can vary from moment to moment rather than remaining fixed.



Schematic · cardiac chambers

Position can change the severity of obstruction

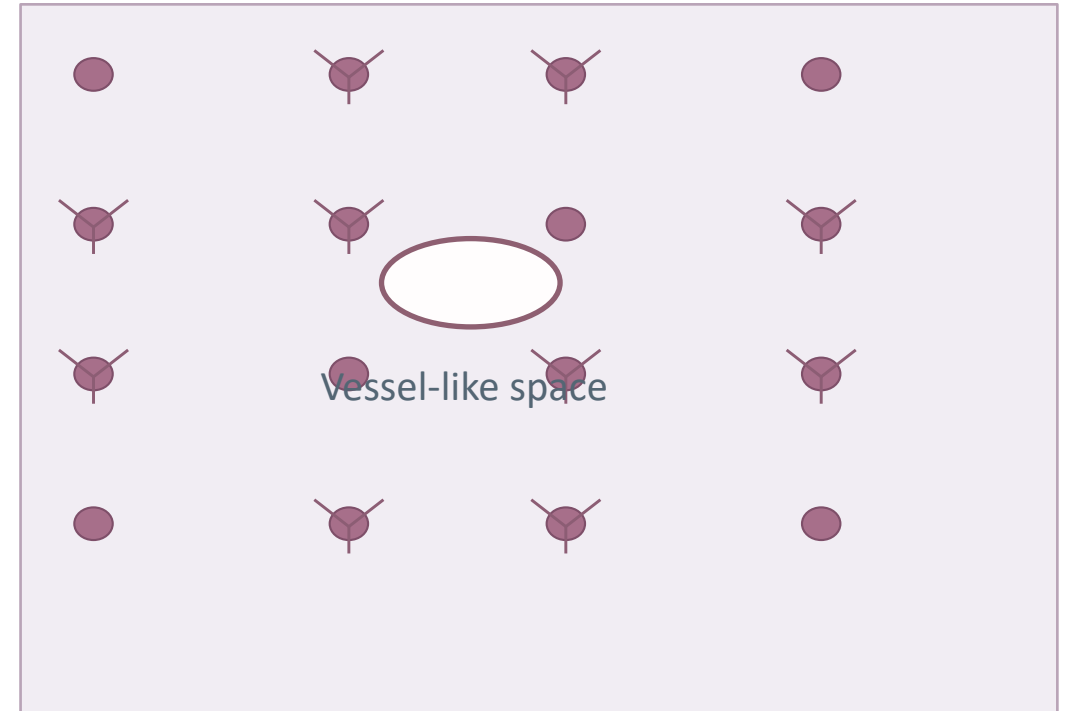


The obstruction is anatomical, not malignant

- A benign myxoma can restrict valve-orifice flow without invading the valve.
- Physical mobility and the attachment site explain episodic symptoms.
- Histologic benignity and hemodynamic significance should be assessed separately.

Myxoma histology: Abundant myxoid matrix

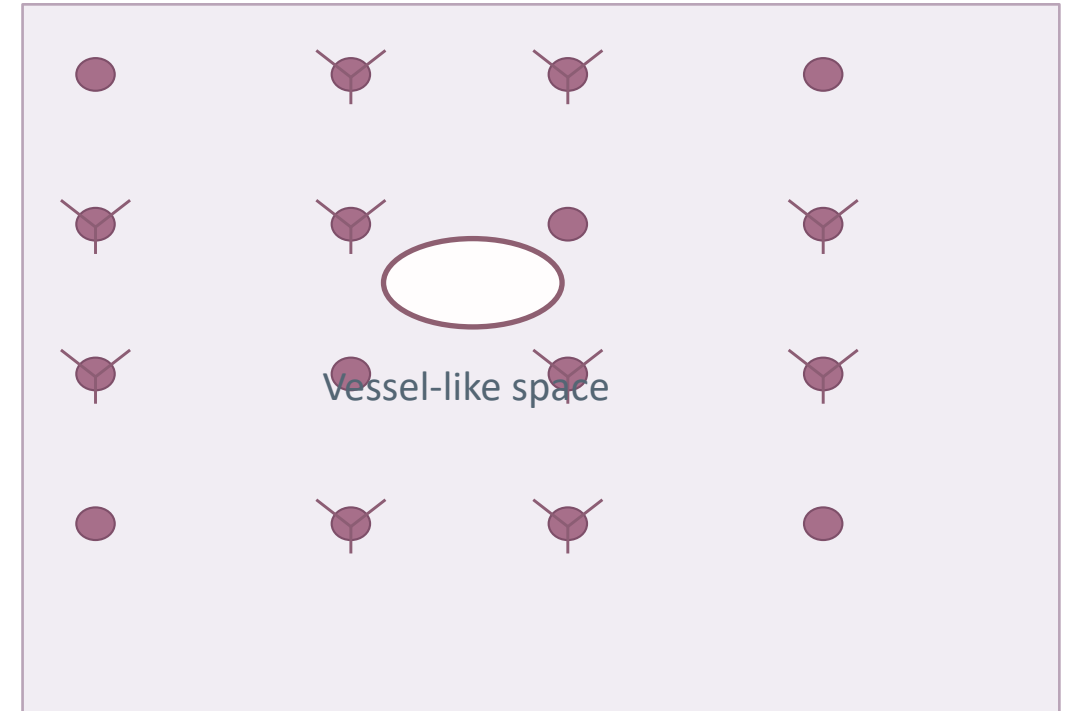
- Extracellular ground substance is rich in acid mucopolysaccharides.
- The matrix separates scattered tumor cells.
- The dominant microscopic pattern is myxoid rather than densely cellular.



Schematic · not an H&E specimen

Myxoma cells may be stellate or globular

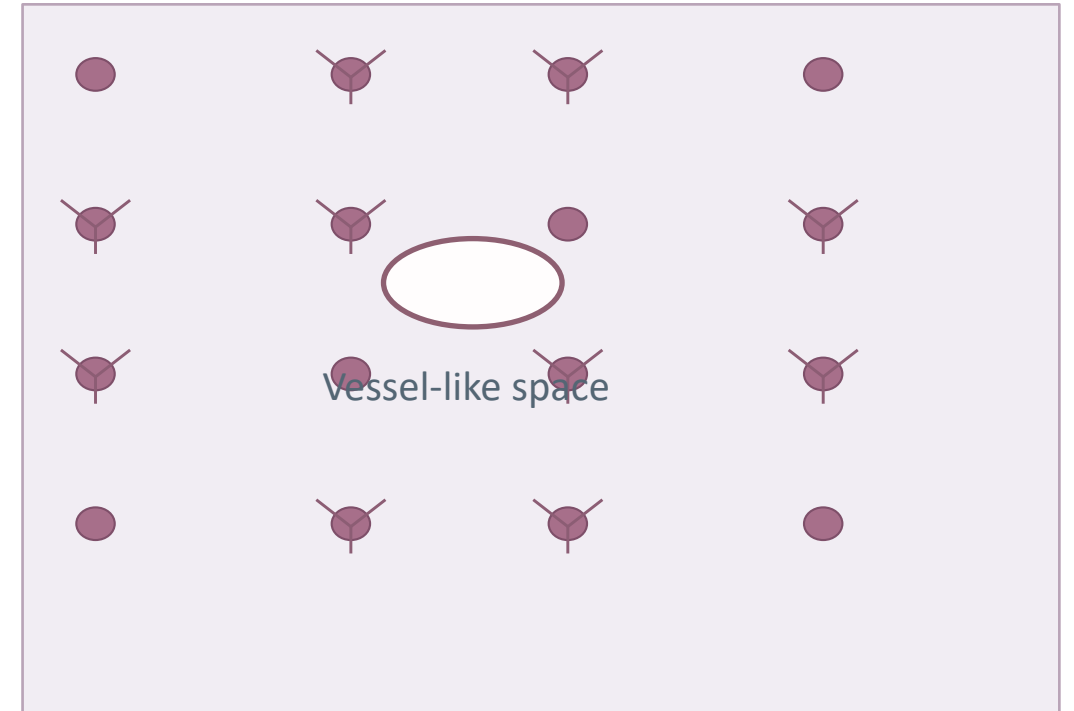
- Stellate cells extend delicate cytoplasmic processes.
- Other tumor cells appear globular within the myxoid ground substance.
- Cell shape alone is not the entire diagnosis; Architectural context matters.



Schematic · not an H&E specimen

Vessel-like and gland-like formations

- Unusual vessel-like or gland-like structures are characteristic morphologic features.
- These patterns sit within the abundant extracellular matrix.
- The schematic represents architecture, not a patient photomicrograph.



Schematic · not an H&E specimen

Hemorrhage and inflammation are usually present

- Hemorrhage frequently accompanies the tumor matrix and cells.
- Mononuclear inflammatory infiltrates are usually encountered.
- A hemorrhagic gross surface and inflammatory microscopic background may coexist.

Morphology integrates matrix, cells, and structures

Component	Histologic description
Extracellular compartment	Abundant acid mucopolysaccharide ground substance
Tumor-cell appearance	Stellate or globular myxoma cells
Distinctive arrangements	Unusual vessel-like or gland-like structures
Associated findings	Hemorrhage and mononuclear inflammation

Three pathways produce myxoma symptoms

Obstruction

Intermittent ball-valve interference with intracardiac flow.

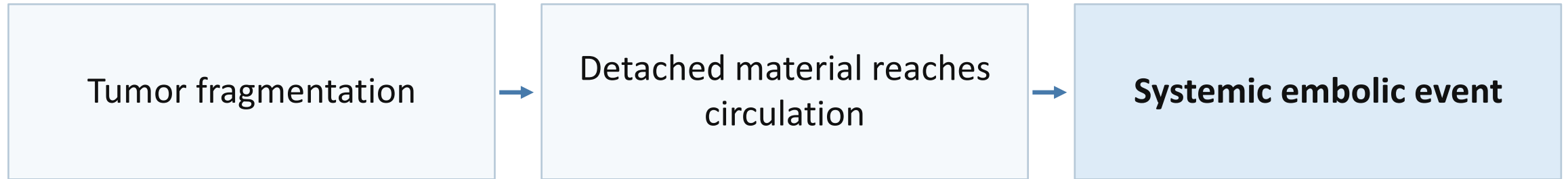
Embolization

Fragments may enter the systemic circulation.

Constitutional

Fever and malaise may reflect tumor-derived cytokines.

Fragmentation produces systemic embolization



Fever and malaise can be tumor related

- A constitutional syndrome including fever and malaise.
- These manifestations may accompany an otherwise benign neoplasm.
- They should be interpreted alongside obstruction and embolization.

Interleukin-6 links tumor and acute-phase response



Echocardiography identifies the intracardiac mass

- Echocardiography provides a noninvasive way to identify the myxoma.
- Its structural utility follows from the lesion projecting into a cardiac chamber.
- The source does not specify modality-specific performance metrics or universal imaging protocols.

Resection is usually curative

Usual outcome

- Surgical removal is typically curative.
- The physical mass and its associated complications are addressed.

Rare recurrence

- Rare recurrence occurs months to years later.
- Incomplete excision is suggested as a likely explanation in the source.

Apply the pattern: Obstructing atrial mass

An adult has intermittent symptoms that vary with position; Imaging shows a mobile atrial mass attached to the septum.

Discriminating clues

- Adult age group
- Atrial septal attachment
- Position-dependent obstruction

Most compatible tumor?

Atrial myxoma — the attachment, mobility, and clinical physiology match the textbook pattern.

Papillary fibroelastoma: Endocardial tumor

- A distinctive, sea-anemone-shaped endocardial lesion.
- Commonly found incidentally rather than through symptoms.
- Its ability to embolize makes an apparently small benign tumor clinically significant.

More than 80% occur on valves

Common location

Valvular surfaces account for more than 80% of lesions.

Gross identity

Delicate hair-like projections arise from a small endocardial base.

Clinical meaning

Valvular attachment does not guarantee functional insignificance.

Valve surface orientation is characteristic

Semilunar valves

- Semilunar valves: Ventricular surfaces are especially favored.
- These include the aortic and pulmonary valve family.

Atrioventricular valves

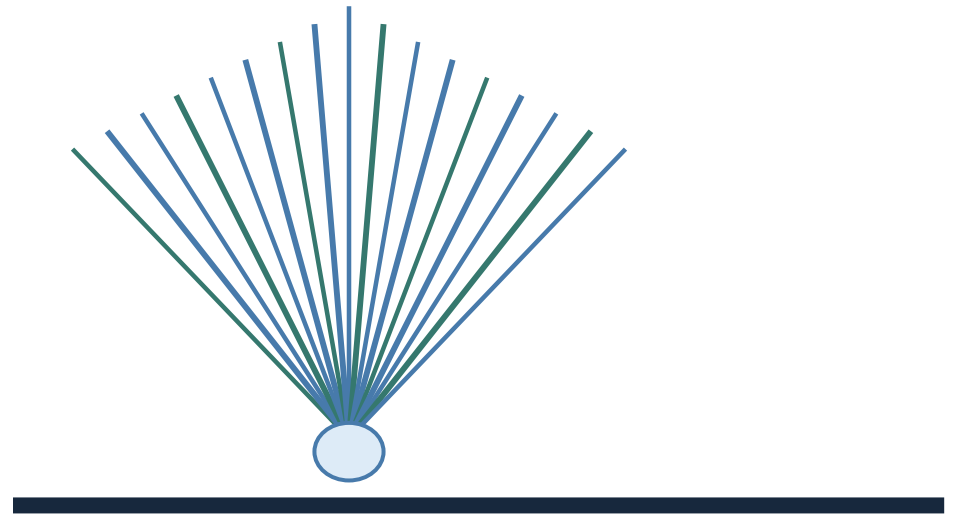
- Atrioventricular valves: Atrial surfaces are especially favored.
- These include the mitral and tricuspid valve family.

Typical fibroelastoma diameter: 0.5–2 cm

- The principal mass is typically 0.5–2 cm in diameter.
- The projecting filaments may reach 1 cm in length.
- A limited mass diameter does not eliminate embolic significance.

Papillary fibroelastoma has hair-like fronds

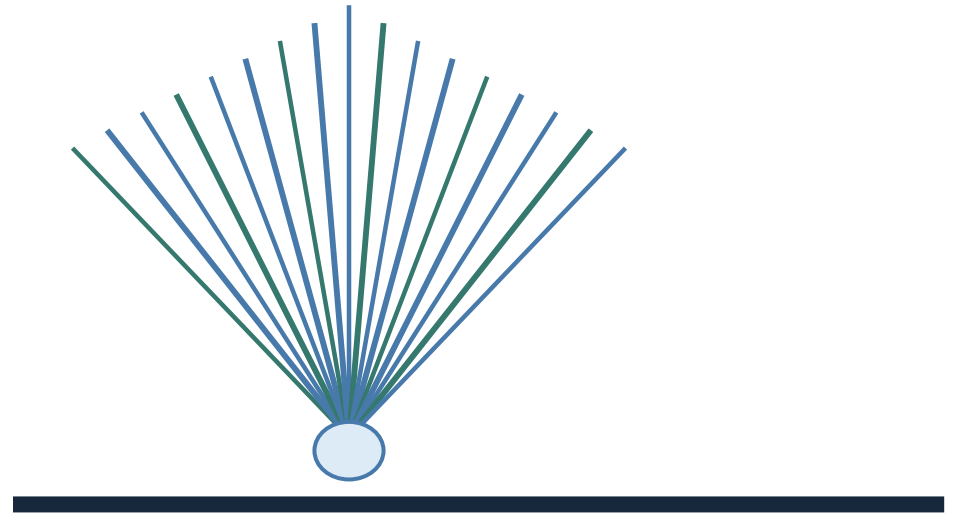
- Numerous delicate projections create a sea-anemone-like outline.
- Hair-like fronds arise from a compact valvular or endocardial attachment.
- This gross architecture is distinct from the gelatinous mass of a myxoma.



Schematic · endothelial-covered fronds

Each papillary projection has an endothelial cover

- An endothelial layer lines the outside of each projection.
- The central portion is connective tissue rather than densely packed neoplastic cells.
- The overall growth pattern is a branching papillary structure.



Schematic · endothelial-covered fronds

The fibroelastoma core contains elastic fibers

Layer / component	Textbook description
Surface	Endothelium
Central stroma	Myxoid connective tissue
Matrix	Abundant mucopolysaccharide ground substance
Other core component	Elastic fibers

Clonal abnormalities support neoplastic status

- Clonal cytogenetic abnormalities have been reported in papillary fibroelastomas.
- These findings support their interpretation as benign neoplasms.
- The source does not provide a specific recurrent chromosomal change or diagnostic molecular test.

Lambl excrescences are the main named look-alike

Papillary fibroelastoma

- Small, papillary, endothelial-covered fibroelastic tumor.
- Often on valves; Can cause embolic consequences.
- Clonal cytogenetic abnormalities have been observed.

Lambl excrescence

- Much smaller projections from sites of valve apposition.
- Usually regarded as trivial comparison.
- Morphologic resemblance does not make them identical lesions.

Fibroelastoma may first matter through embolization

Incidental endocardial lesion



Potential embolization



Clinically important event

Apply the pattern: Valvular fronded lesion

A small incidental valvular tumor has many hair-like projections, and its stalk is attached to the valve surface.

Discriminating clues

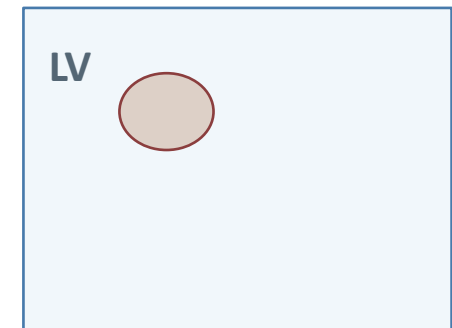
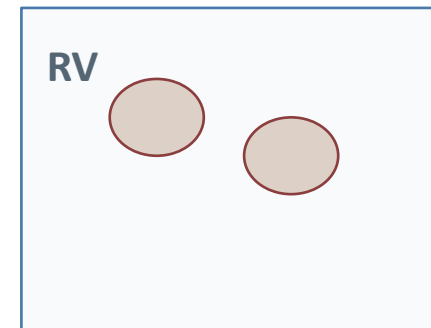
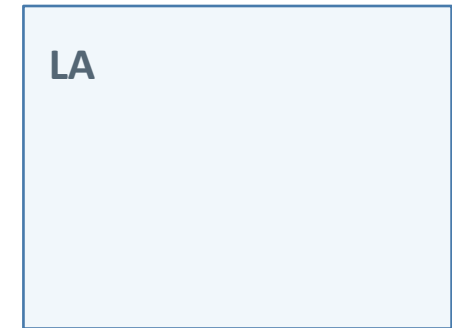
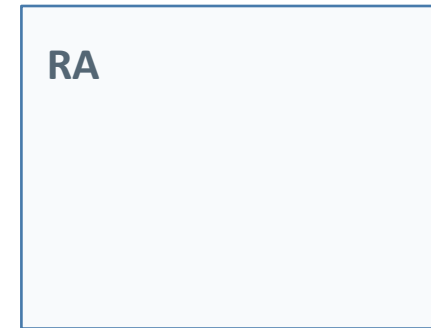
- Often valvular location
- Sea-anemone-like contour
- Fibroelastic papillary core

Most compatible tumor?

Papillary fibroelastoma; Distinguish it from the much smaller Lambl excrescence.

Rhabdomyoma predominates in pediatric hearts

- The most frequent primary tumor of the pediatric heart.
- Often detected during the first years of life.
- Evaluation may be prompted by valve dysfunction or flow obstruction.



Schematic · cardiac chambers

About half are associated with tuberous sclerosis

Syndromic group

- Approximately 50% of cardiac rhabdomyomas are associated with tuberous sclerosis.
- The syndrome involves TSC1 or TSC2 tumor suppressor gene mutations.

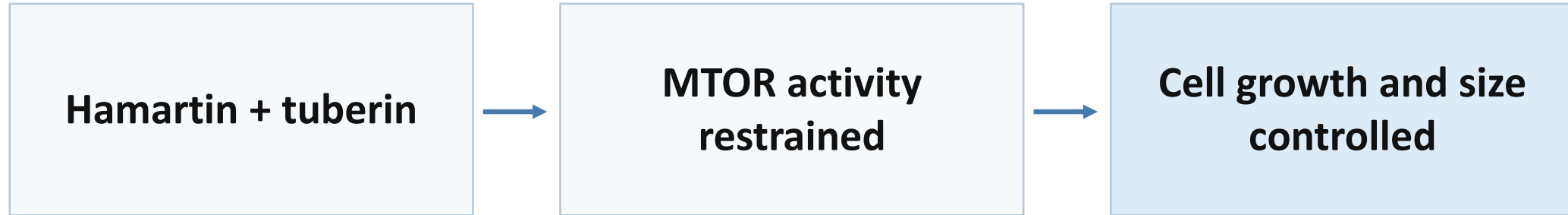
Sporadic group

- Approximately half are attributed to sporadic mutations.
- Do not label every cardiac rhabdomyoma as tuberous-sclerosis-associated.

TSC1 and TSC2 encode hamartin and tuberin

Gene / complex	Product	Role in source
TSC1	Hamartin	Tumor suppressor protein
TSC2	Tuberin	Tumor suppressor protein
TSC1/TSC2 complex	Functional pair	Inhibits mTOR activity

TSC1–TSC2 complex inhibits mTOR



Loss of inhibitory control permits overgrowth



Spontaneous regression challenges strict tumor labels

Observed biological behavior

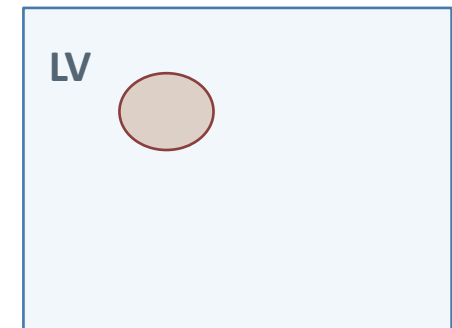
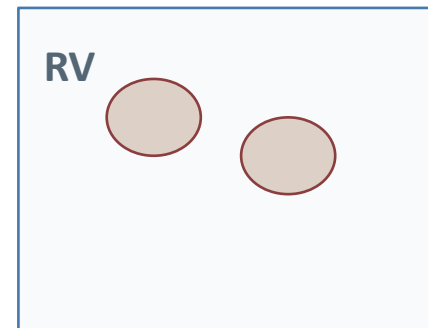
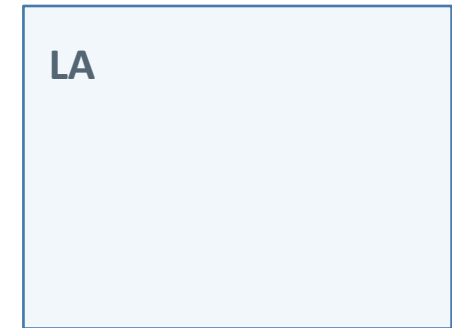
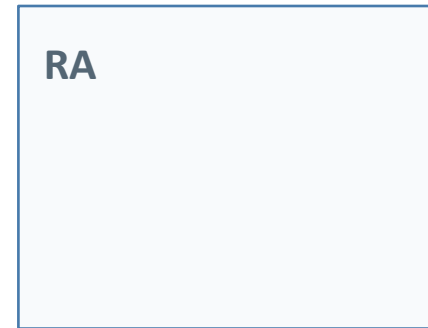
- Rhabdomyomas often regress spontaneously.
- The natural history does not resemble relentless malignant growth.

Interpretive consequence

- Some authors consider them hamartomas rather than true neoplasms.
- This is a classification interpretation, not a universal requirement.

Rhabdomyoma prefers ventricular myocardium

- Myocardial masses preferentially involve the ventricles.
- Lesions can bulge into the ventricular cavity.
- The intraluminal projection can account for flow obstruction.



Schematic · cardiac chambers

Rhabdomyoma: Multiple gray-white masses

Gross color

Typically gray-white myocardial masses.

Multiplicity

Usually multiple rather than solitary.

Size and site

From small lesions to masses several centimeters in ventricular myocardium.

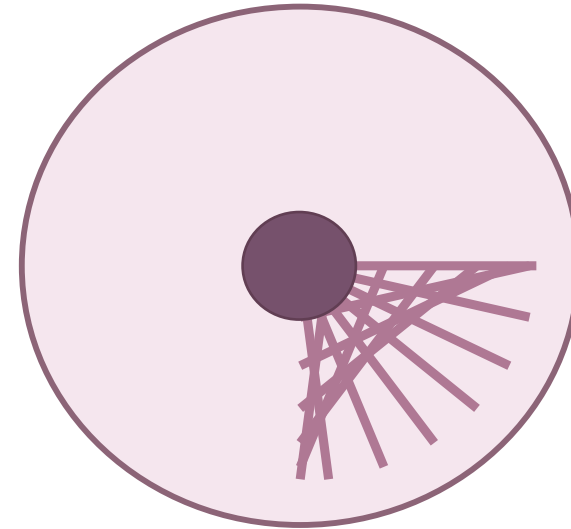
Enlarged cardiac myocytes contain glycogen

- Microscopy shows bizarre, markedly enlarged myocytes.
- Large intracellular glycogen collections are a central feature.
- Routine tissue processing removes much of the glycogen.

“Spider cells” reflect processing of glycogen

- Glycogen extraction leaves the cytoplasm appearing empty.
- Residual cytoplasm remains as thin strands from nucleus to cell membrane.
- This produces the classic spider-cell appearance.

Central nucleus



Thin residual cytoplasmic strands

Schematic · glycogen extraction artifact

Rhabdomyoma effects depend on position

Ventricular myocardial mass



Protrusion into lumen



**Valve dysfunction or flow
obstruction**

Apply the pattern: Infant ventricular masses

An infant is evaluated for cardiac flow obstruction; Imaging shows multiple ventricular myocardial masses.

Discriminating clues

- Age in first years of life
- Ventricular and multiple lesions
- Possible tuberous sclerosis association

What further mechanism fits?

Cardiac rhabdomyoma; Consider TSC1/TSC2-mediated loss of mTOR restraint.

Fibroma and lipoma: Limited textbook detail

Fibroma

- Among the five most common primary benign cardiac tumors.
- Morphologically resemble fibromas arising elsewhere.
- No site-specific mechanism is detailed here.

Lipoma

- Also among the five most common benign primary tumors.
- Resemble lipomas at extracardiac sites.
- No additional cardiac-specific morphology is supplied.

Primary cardiac sarcoma: The rare exception

Angiosarcoma

- Angiosarcoma is one of the typical malignant types.
- The section provides no special cardiac-specific diagnostic morphology.

MDM2-amplified sarcoma

- Undifferentiated sarcoma with MDM2 amplification is another named type.
- MDM2 is a molecular finding mentioned in this source.

Primary tumor versus cardiac metastatic deposit

Primary cardiac tumor

- Originates within the heart.
- Most examples are benign.
- Myxoma, fibroelastoma, and rhabdomyoma are the detailed prototypes.

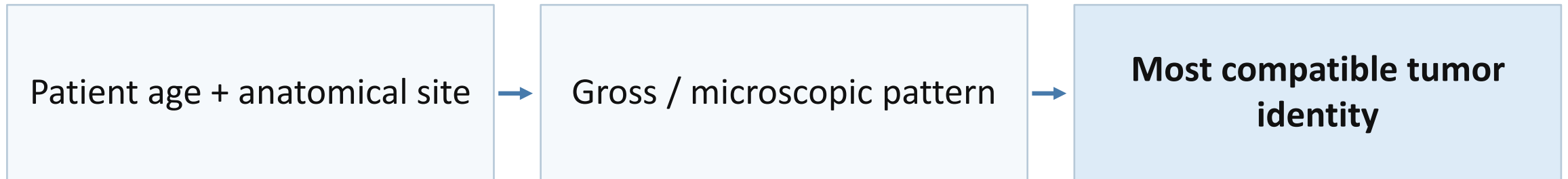
Metastatic involvement

- Reaches heart or pericardium from a noncardiac cancer.
- The adjacent section identifies several metastatic routes.
- Cardiac secondary involvement is a separate diagnostic category.

Compare the three diagnostic prototypes

Entity	Predilection	Morphology	Clinical link
Myxoma	Adult; Atrial	Gelatinous mass; Myxoid matrix	Obstruction / emboli / IL-6
Fibroelastoma	Valvular endocardium	Hair-like endothelial papillae	Potential emboli
Rhabdomyoma	Child; Ventricle	Glycogen-rich spider cells	Flow obstruction; Regression

A pattern-based approach to a cardiac mass



Three patterns worth retaining

Atrial myxoma

Myxoid matrix + septal attachment + variable obstruction.

Papillary fibroelastoma

Valvular hair-like fronds + endothelial covering + embolic risk.

Cardiac rhabdomyoma

Ventricular pediatric masses + glycogen-laden spider cells + TSC association.

Reference

- Robbins, Cotran & Kumar: Pathologic Basis of Disease. 11th edition. Elsevier; 2026.
- The Heart — Primary Cardiac Tumors.