Myxoma

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- Myxomata comprise 50% of all benign cardiac tumors in adults and 15% of such tumors in children. Occurrence during infancy is rare .
- A vast majority of myxomas occur sporadically and tend to be more common in women .
- The peak incidence is between the third and sixth decades of life, and 94% of tumors are solitary .
- Approximately 75% occur in the left atrium and 10 to 20% occur in the right atrium. The remaining proportion are equally distributed between the ventricles .
- (DNA) genotype of sporadic myxomas is normal in 80% of patients.
- Myxomas are unlikely to be associated with other abnormal conditions and have a low recurrence rate .
- About 5% of myxoma patients show a familial pattern of tumor development based on autosomal dominant inheritance .

Abnormal DNA genotype chromosomal pattern

- Younger
- More often (22%) have multicentric tumors
- Higher recurrence rate after surgical resection (21-67%)
- Approximately 20% of familial patients have associated conditions (***complex myxomas***):
  - ✓ Adrenocortical nodule hyperplasia
  - ✓ Sertoli cell tumors of the testes
  - ✓ Pituitary tumors
  - ✓ Multiple myxoid breast fibroadenomas
  - ✓ Cutaneous myomas
  - ✓ Facial or labial pigmented spots.
- A familial syndrome with autosomal X-linked inheritance characterized by primary pigmented nodular adrenocortical disease with hypercortisolism, cutaneous pigmentous lentigines, and cardiac myxoma is referred to as ***Carney's complex***

PATHOLOGY

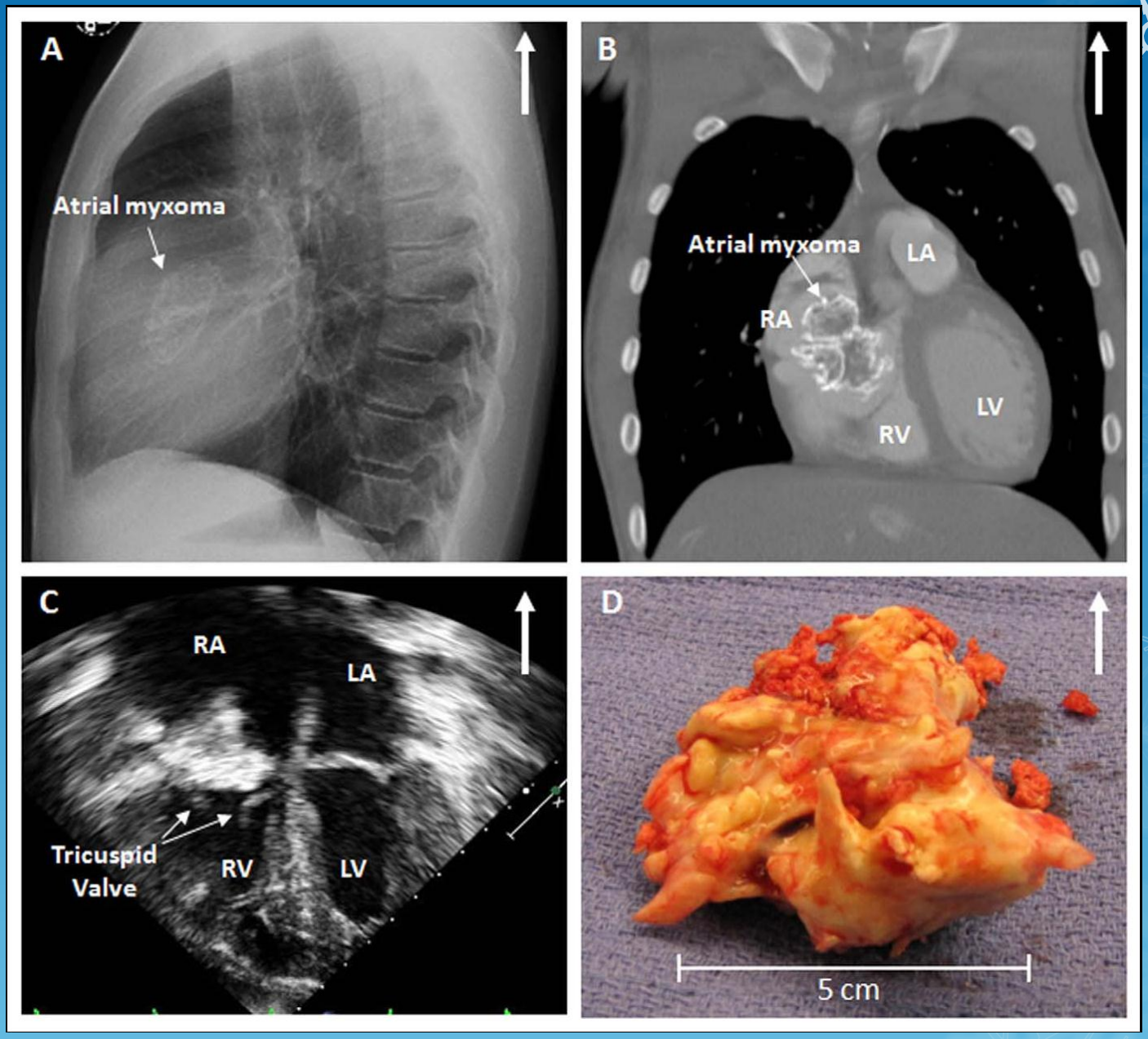
- Biatrial tumors probably arise from bidirectional growth of a tumor originating within the atrial septum.
- Atrial myxomas generally arise from the interatrial septum at the border of the fossa ovalis but can originate anywhere within the atrium
- Isolated reports confirm that myxomas can arise from the cardiac valves, pulmonary artery (PA) and vein, and vena cava.
- Right atrial myxomas are more likely to have broad-based attachments than left atrial tumors; they also are more likely to be calcified.
- Ventricular myxomas occur more often in women and children and may be multicentric.
- Right ventricular tumors typically arise from the free wall.
- Left ventricular tumors tend to originate in the proximity of the posterior papillary muscle.
- Two-thirds of myxomas are round or oval tumors with a smooth or lobulated surface .Most are polypoid, compact, pedunculated, mobile, and not likely to fragment spontaneously.
- One-third of myxomas are villous or papillary.They are gelatinous and fragile and prone to fragmentation and embolization .
- The average size is about 5 cm in diameter, but growth to 15 cm in diameter and larger has been reported.
- Weights range from 8 to 175 g, with a mean between 50 and 60 g.

CLINICAL PRESENTATION

- The classic clinical presentation of a myxoma is intracardiac obstruction with congestive heart failure (67%).
- Signs of embolization (29%).
- Systemic or constitutional symptoms of fever (19%).
- Weight loss or fatigue (17%).
- Immunologic manifestations of myalgia, weakness, and arthralgia(5%).
- Cardiac rhythm disturbances and infection occur less frequently.
- Infection increases the likelihood of systemic embolization,and an infected myxoma warrants urgent surgical resection.

DIAGNOSIS

- Clinical Examination.
- Chest Radiograph and Electrocardiogram. More specific rare findings are density within the cardiac silhouette caused by calcification within the tumor.
- Echocardiography: The sensitivity of two-dimensional (2-D) echocardiography for myxoma is 100%. (TEE) provides the best information concerning tumor size, location, mobility, and attachment.
- Computed Tomography and Magnetic Resonance Imaging: Neither CT nor MRI is needed for atrial myxomas if an adequate echocardiogram is available. The exception is the occasional right atrial myxoma that extends into one or both caval or tricuspid orifices.

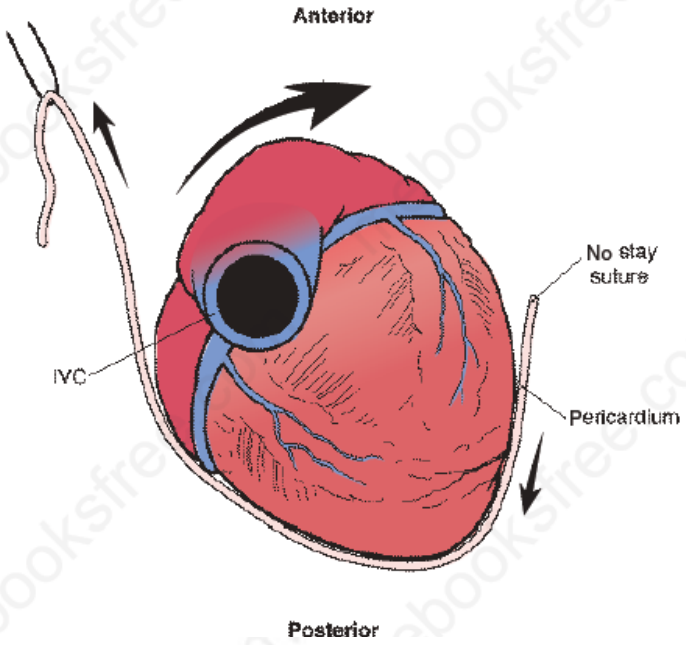

A
Atrial myxoma
B
Atrial myxoma
LA
RA
RV
LV
C
RA
LA
Tricuspid Valve
RV
LV
D
5 cm

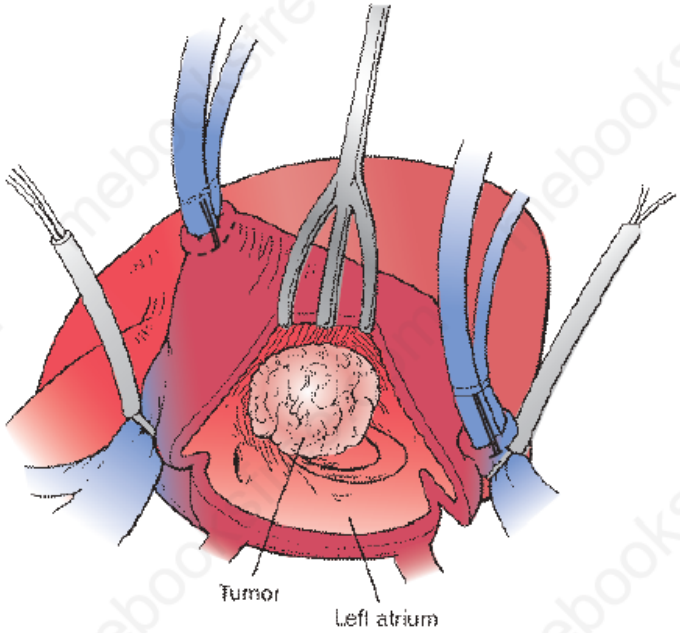
SURGICAL MANAGEMENT

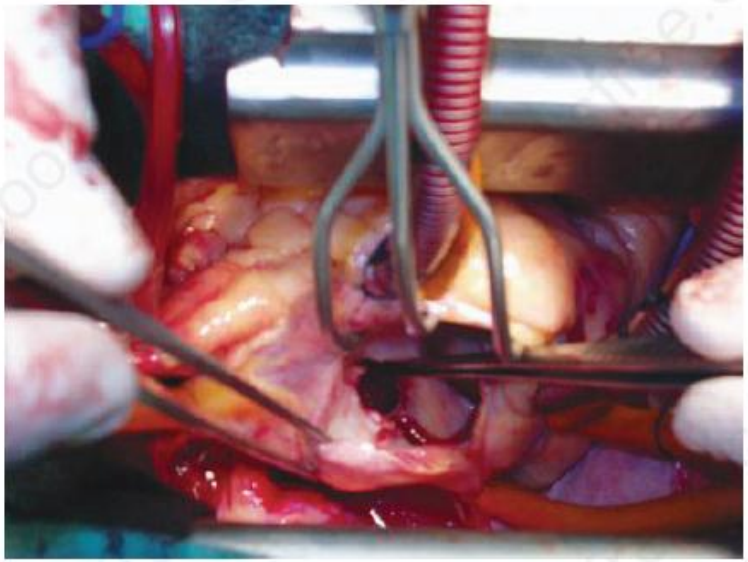
- Surgical resection is the only effective therapeutic option for patients with cardiac myxoma and should not be delayed because death from obstruction to flow within the heart or embolization may occur in as many as 8% of patients awaiting operation.
- A median sternotomy approach with ascending aortic and bicaval cannulation usually is employed.
- In the event of preoperative known cerebral embolization without hemorrhage, the tumor should be resected approximately seven days after the event to prevent further embolization and yet allow time for stabilization of the brain for CPB.

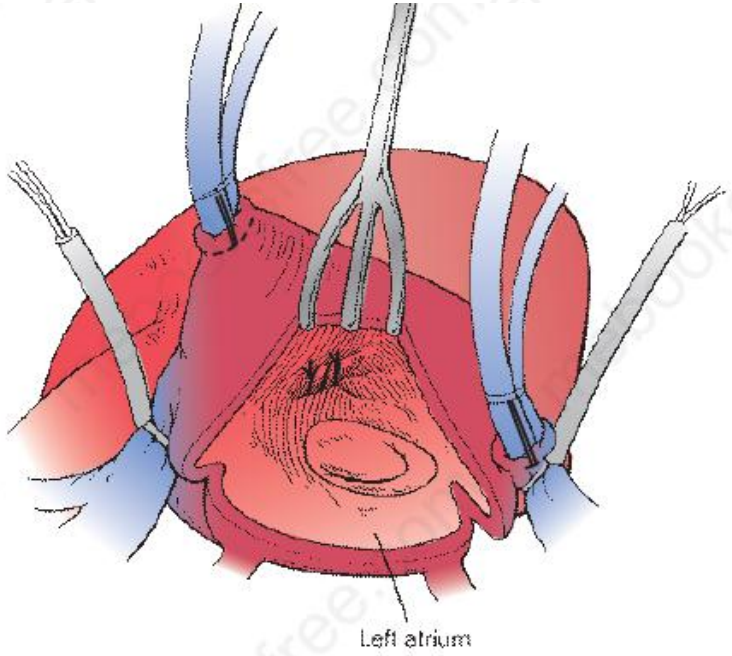
Left atrial myxomas

- Exposure of left atrial myxomas is maximized by using several principles from mitral valve repair surgery.
- Left atrial myxomas can be approached by an incision through the anterior wall of the left atrium anterior to the right pulmonary veins
- Exposure and removal of large tumors attached to the interatrial septum may be aided by a second incision parallel to the first in the right atrium. Biatrial incision allows easy removal of tumor attached to the fossa ovalis with a full-thickness Ro (margin-negative) excision at the site of attachment and easy patch closure of the atrial septum if necessary.


Anterior
No stay suture
IVC
Pericardium
Posterior


Tumor
Left atrium




Left atrium

Right atrial myxomas

- Right atrial myxomas pose special venous cannulation problems, and intraoperative echocardiography may be beneficial.
- Both venae cavae may be cannulated directly.
- When low- or high-lying tumor pedicles preclude safe transatrial cannulation, cannulation of the jugular or femoral vein can provide venous drainage of the upper or lower body.
- Resection of large or critically placed right atrial myxomas often requires careful preoperative planning, intraoperative TEE, and special extracorporeal perfusion techniques to ensure complete removal of the tumor, protection of right atrial structures, and reconstruction of the atrium.
- Because myxomas rarely extend deep in the endocardium, it is not necessary to resect deeply around the conduction tissue.

- The tricuspid valve and the right atrium, as well as the left atrium and ventricle, should be inspected carefully for multicentric tumors in patients with right atrial myxoma.
- Regardless of the surgical approach, the ideal resection encompasses the tumor and a portion of the cardiac wall or interatrial septum to which it is attached.
- Partial thickness resection of the area of tumor attachment has been performed when anatomically necessary without a noted increase in recurrence rate.

Ventricular myxomas

- Usually are approached through the AV valve or by detaching the anterior portion of the AV valve for exposure and resection and reattachment after resection.
- Occasional small tumors in either outflow tract can be removed through the outflow valve.
- If necessary, the tumor is excised through a direct incision into the ventricle, but this is unusual and the least preferred approach.
- It is not necessary to remove the full thickness of the ventricular wall because no recurrences have been reported with partial-thickness excisions.
- As with right atrial myxoma, the presence of ventricular myxoma prompts inspection for other tumors because of the high incidence of multiple tumors.
- There are rare instances of distant metastases from myxoma many years after tumor resection, and these reports raise the issue of potential intraoperative dissemination of tumor.

Results

- Removal of atrial myxomas carries an operative mortality rate of 5% or less.
- Excision of ventricular myxomas can carry a higher risk (approximately 10%).
- Recurrence of nonfamilial sporadic myxoma is approximately 1 to 4%.
- The 20% of patients with sporadic myxoma and abnormal DNA have a recurrence rate estimated at between 12 and 40%.
- The recurrence rate is highest in patients with familial complex myxomas, all of whom exhibit DNA mutation, and this is estimated to be about 22%.
- Most recurrent myxomas occur within the heart, in the same or different cardiac chambers, and may be multiple.
- Extracardiac recurrence after resection of tumor, presumably from embolization and subsequent tumor growth and local invasion, has been observed.
- Myxomas generally classified as “malignant” are often found on subsequent review to be sarcomas with myxoid degeneration.
- Symptomatic lesions of possible metastatic myxoma should be excised if feasible.

Thank You