31<sup>st</sup> Annual ECHO Hawaii

January 21, 2022 | 8:15 – 8:30 AM | 15 min

### Multimodality Assessment of Cardiac Masses

**Muhamed Sarić MD, PhD, MPA** Director of Noninvasive Cardiology | Echo Lab Professor of Medicine New York University



#### Disclosures

Speakers Bureau (Abbott, Boston Scientific, Medtronic, Philips) Advisory Board (Siemens)

Which of the following is the most common nonvalvular benign cardiac tumor seen on echocardiography in adults?

- A. Fibroma
- B. Myxoma
- C. Papillary fibroelastoma
- D. Rhabdomyoma
- E. Teratoma



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Each of the following tumors are known to invade the heart via the inferior vena cava EXCEPT?

- A. Adrenal carcinoma
- B. Hepatocellular carcinoma
- C. Leiomyoma
- D. Renal cell carcinoma
- E. Rhabdomyoma



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# **Three-of-a-Kind Game**

#### Which 1 of these 4 images does NOT belong to the set?





#### Panel A does NOT belong to the set.

Panel A is the only panel NOT showing a thrombus.







Panel B

# Large LA appendage thrombus in a patient with atrial fibrillation











## Primary & Secondary Cardiac Tumors

In general, cardiac tumors, whether benign or malignant, are rare



## Primary Cardiac Tumors

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**PRIMARY CARDIAC TUMORS** 

#### Prevalence

• Very rare (1 in 2,000)

#### **BENIGN PRIMARY CARDIAC TUMORS**

- MOST COMMON NONVALVULAR TUMORS
  - Myxoma (adults)
  - Rhabdomyoma (kids)
- MOST COMMON VALVULAR TUMOR
  - Papillary fibroelastoma
- OTHER BENIGN TUMORS
  - Lipoma
  - Fibroma

MALIGNANT PRIMARY CARDIAC TUMORS

• VARIOUS SARCOMAS

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MALIGNANT PRIMARY CARDIAC TUMORS

• VARIOUS SARCOMAS

#### Relative Prevalence of Benign Cardiac Tumors



### Manifestations of Cardiac Tumors



#### **Rhetorical Question**

What's the embolic potential of an **incidentally found** intracardiac mass?

#### Embolic Potential of Incidental Cardiac Masses

#### JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY

VOL. 73, NO. 17, 2019

		Silent Embolism on Brain Imaging
	Myxoma	63%
.etters	PFE	73%
	LV Thrombus	33%

High Prevalence of Subclinical Infarction in Asymptomatic Patients With Silent Left-Sided Cardiac Masses



of silent ischemic lesions. To exclude other possible sources of systemic embolism, we then performed 48-h Holter monitoring, duplex carotid ultrasound, and contrast enhanced transthoracic echocardiography on all patients. Coronary imaging with CT angiography or invasive angiography was subsequently performed to exclude concomitant obstructive coronary artery disease. Thereafter, all patients underwent

#### Cardiac Masses





# Myxomas



# **Embolic Potential of Myxomas**

- In at least **1/3** of cases of myxomas, there is evidence of distant embolism, including to the brain
- Embolism may be silent or cause neurologic symptoms
- Neurologic deficits may be the **first** presenting symptom of a myxoma

### Location of Cardiac Myxomas



### Most Typical LA Myxoma Location



#### **Typical Left Atrial Myxoma**



### Left Atrial Myxoma: Microbubble Contrast



#### LA Myxoma: 3D TEE



#### Myxoma Gross Specimen



Myxomas often feature thrombus formation on its surface

*J Am Soc Echocardiogr* 2011;24:110.e1-110.e4.

# Myxoma Histology



**Panel A**: Microscopic appearance under 4x power shows abundant **amorphous extracellular matrix** and scanty cellular elements

**Panel B**: Microscopic appearance under 10x power reveals scattered **stellate or myxoma cells**, endothelial and smooth muscle cells in abundant **matrix with poorly formed vessels**. No areas of hemorrhage are seen

**Panels C & D**: Lesion was **CD 34 and calretinin positive**, respectively, which is consistent with diagnosis of myxoma



Rudolph Carl Virchow (1821 – 1902) German pathologist The term 'myxoma' was coined in **1857** by Rudolph Virchow

> Greek μύξωμα : myxoma (< μύξα : myxa - mucus) 'Mucus Containing Tumor'



Rudolph Carl Virchow (1821 – 1902) German Pathologist 3.

Ein Fall von bösartigen, zum Theil in der Form des Neuroms auftretenden Fettgeschwülsten.

Von Rud. Virchow.

Am 3. Januar d. J. kam die Leiche eines 53 jährigen Schneiders zur Section, der seit etwa 11 Wochen von heftigen, dem Anscheine nach rheumatischen Schmerzen heimgesucht war, gegen welche er sowohl in Bethanien, als in der Charité vergeblich Hülfe gesucht hatte. Insbesondere hatte er über einen gürtelartigen Schmerz in der Lendengegend und über Reissen in den Hüften geklagt. Schon

Virchow, R. Ein Fall von bösartigen, zum Theil in der Form des Neuroms auftretenden Fettgeschwülsten [<mark>A case of malignant fatty tumors,</mark> occurring partly in the form of neuroma]. Archiv für pathologische Anatomie und Physiologie und für klinische Medicin **1857**;11(3):281-288.



**Thomas Hodgkin** (1798 – 1866) English Physician The first description of an apparent left atrial myxoma was mad in London by Thomas Hodgkin in **1829** 



**Thomas Hodgkin** (1798 – 1866) English Physician

To Robert Finch with the best respects of his attached find with the best respects of his attached find CATALOGUE OF THE PREPARATIONS IN THE ANATOMICAL MUSEUM GUY'S HOSPITAL. ARRANGED AND EDITED, BY DESIRE OF THE TREASURER OF THE HOSPITAL, AND OF THE TEACHERS OF THE MEDICAL AND SURGICAL SCHOOL, nv. THOMAS HODGKIN, M.D.

LICENTIATE OF THE ROVAL COLLEGE OF PHYSICIANS OF LONDON. DEMONSTRATOR OF MORBID ANATOMY AND CHATOR OF THE NUSEUM AT GUY'S HOSPITAL, MEMBER OF THE ROVAL MEDICAL SOCIETY OF CANUNEGH, AND CORRESPONDING MEMBER OF THE LYNCEAN ACADEMY OF ROME AND OF THE ORDINNIAN SOCIETY OF CATAREA, &C.

Accessed and a second second second

M DCCC XXIX.



**Thomas Hodgkin** (1798 – 1866) English Physician The case was that of a female, ætat. 55, with general dropsy and thoracic obstruction. Dr. Hodgkin's *post-mortem* history is quite satisfactory (Green, Inspn.-book, 3, p. 157):---

"In the left auricle, close to the margin of the foramen ovale, was attached a large polypiform body of about the size of a pullet's egg. The surface by which it was attached was nearly the size of a shilling. Though rather dark and discoloured in some parts, it was generally of a light-yellow, and semi-transparent, with some opaque white specks dispersed through it, having some resemblance to the opaque points in soft soap, but rarer and smaller. It was firmer than the ordinary fibrinous concretions which are found in the heart, and was especially so about its root. On fine injection being thrown into the coronary arteries, minute vessels were seen beautifully ramifying through the transparent substance. This polypiform concretion was covered by a thin membrane continuous with that of the lining membrane of the auricle. Though its substance was not disposed in layers, some appearances in it, (as, for instance, darkish points of a brownish colour) had a trace of arrangement equi-distant from the surface."-The museum catalogue is less distinct about the nature of this body. Allan Burns is amongst the authorities for the term concretion.
#### Less Typical LA Myxoma Locations

#### Case #1



#### 68-year-old man with vague neurologic symptoms



#### 68-year-old man with vague neurologic symptoms







68-year-old man with vague neurologic symptoms





#### Less Typical LA Myxoma Locations

#### Case #2



#### LA Myxoma Behind Aortic Valve

#### 71-year-old woman with transient ischemic attack



#### LA Myxoma Behind Aortic Valve



## LA Myxoma Behind Aortic Valve





#### Less Typical LA Myxoma Presentation



#### LA Myxoma Across Mitral Valve

63-year-old woman with left-sided weakness



#### LA Myxoma Across Mitral Valve



#### LA Myxoma Across Mitral Valve: MRI



#### An Intriguing Co-Existence: Atrial Myxoma and Cerebral Cavernous Malformations: Case Report and Review of Literature

Shikha Sharma, MD, Daniel Tsyvine, MD, Pierre D. Maldjian, MD, Justin T. Sambol, MD, Constantinos J. Lovoulos, MD, Gal Levy, MD, Amin Maghari, MD, Marc Klapholz, MD, and Muhamed Saric, MD, PhD, Newark, New Jersey



It is commonly postulated that neurologic complications of atrial myxomas are due to either direct tumor embolization or mycotic aneurysm of cerebral vasculature or rupture of mycotic aneurysms of cerebral arteries. However, the authors report the case of 63-year-old woman with a large left atrial myxoma whose progressive left-sided weakness was due to a different neurologic mechanism, namely, multiple bleeding cavernous malformations, which were visualized by magnetic resonance imaging of the brain. Cerebral cavernous malformations coexist with mesenchymal anomalies of other organs, including the liver, kidneys, and retinas. To the best of the authors' knowledge, this is only the second reported case of coexistent cerebral cavernous malformations and atrial myxoma. (J Am Soc Echocardiogr 2011;24:110.e1-110.e4.)

# Left Ventricular Myxoma



#### Left Ventricular Myxoma

#### 50-year-old man fevers & night sweats for 1 month



#### Left Ventricular Myxoma: Microbubble Contrast





# 63-year-old man with hypotension and suspected pulmonary embolism

- Premature puberty due to pigmented nodular adrenal hyperplasia
- He underwent excision of a benign myxoid neurofibroma of the cervical spine with radiculopathy ~ 10 years ago.
- He has plane **lentigenes** (epithelioid blue nevi)
- Prior surgical removal of 2 left atrial myxomas









#### Carney Complex



#### The Complex of Myxomas, Spotty Pigmentation, and Endocrine Overactivity

J. AIDAN CARNEY, M.D., PH.D., F.R.C.P.I., HYMIE GORDON, M.D., F.R.C.P., PAUL C. CARPENTER, M.D., B. VITTAL SHENOY, M.D., AND VAY LIANG W. GO, M.D.

*Medicine* **1985**;64(4):270-83

**J. Aidan Carney** Mayo Clinic Pathologist

### Carney Complex





#### Approximately **7% of all cardiac myxomas** are associated with Carney complex

*N Engl J Med*. **1995** Dec 14;333(24):1610-7

Spotty skin pigmentation, 65%



Cutaneous myxomas, 45%



Cardiac myxomas, 72%





PPNAD, 45%



GH-secreting pituitary tumor, 10%



Schwannomas, 5%



°0.





Testicular tumors, 56%

#### Very Unusual Form of Myxoma



### **Clinical History**

#### 64-year-old businessman living in an affluent New York City suburb

1948	Born in New York City	
1974	At age 26, treated for testicular cancer (left orchiectomy + chemo)	
Sep 2011	Sudden onset of partial vision loss in left eye >> Extensive workup but no echocardiogram >> `Nonarteritic anterior ischemic optic neuropathy (ION) of left eye'	
	<ul> <li>ION</li> <li>Loss of structure and function of a portion of the optic nerve due to obstruction of blood flow to the nerve</li> <li>No accepted treatment to reverse the damage</li> </ul>	X

#### Teaching Points Thus Far...

• **Ophthalmic artery**, the first branch of the **internal carotid artery** distal to the cavernous sinus, is part of cerebral arterial circulation

Vascular events in the eye may be stroke equivalents

Suggestive of cardioembolic source

# **Clinical History**

Sep 2011	Sudden onset of partial vision loss in left eye >> Extensive workup but no echocardiogram >> 'Nonarteritic anterior ischemic optic neuropathy (ION) of left eye'	
Dec 2012	Saw a dentist for `tooth cleaning'	
Jan 2013	New-onset malaise, fatigue and excessive sleepiness >> Given oral cephalosporin for 'respiratory infection'	
Feb 2013	Traveled to Dubai, United Arab Emirates; recurrent fevers >> Given oral azithromycin for 'respiratory infection' >> 'Mitral valve prolapse' on physical exam	
Mar 2013	<section-header><ul> <li>Positive blood culture</li> <li>&gt; Streptococcus sanguinis</li> <li>A viridans group organism</li> <li>Normal inhabitant of the human mouth, especially in dental plaque</li> </ul></section-header>	

### Transthoracic Echocardiogram

Transthoracic echocardiogram at an outside hospital reportedly revealed **a large mobile mass associated with the mitral valve** 

# **Clinical Diagnosis**

Infectious disease consult stated: Subacute bacterial endocarditis due to *Streptococcus sanguinis* 

'Large mobile mass on echo + positive blood cultures; patient meets Duke criteria for infective endocarditis'



Duke criteria first published *Am J Med*. 1994 Mar;96(3):200-9.



Mass in LA and LV (? Free floating)



MIDESOPHAGEAL 4-CHAMBER VIEW 13.0 × 1.5 cm mass tethered in LA & prolapses through MV into LV



MIDESOPHAGEAL 4-CHAMBER VIEW Mass attached to the interatrial septum near fossa ovalis

MIDESOPHAGEAL VIEW Neither significant mitral regurgitation nor mitral stenosis observed



### 3D TEE



**3D TEE: LA SIDE OF MITRAL VALVE** Mass attached to the interatrial septum **3D TEE: LV SIDE OF MITRAL VALVE** Mass plopping through mitral valve
### Intraoperative Video



#### Surgical Specimen



#### **Final Diagnosis**

#### Nonfamilial left atrial myxoma superinfected with *Streptococcus sanguinis*

Leading to subacute infective endocarditis and earlier to systemic embolism to left eye

#### JAm Coll Cardiol. 2014;63(19):2049





#### **Papillary Fibroelastoma**

#### Most Common Valvular Tumor with Significant Embolic Potential



#### Papillary Fibroelastoma



Branching **avascular papillae** containing collagen and covered with endothelium



#### PFEs commonly resemble sea anemone

ἀνεμώνη (anemṓnē), from ἄνεμος (ánemos, "wind") + matronymic suffix -ώνη (-ṓnē, "daughter of the wind")

#### Typically on the **aortic side** of the valve











#### **Images in Geriatric Cardiology**

Navin C. Nanda, MD, Section Editor The University of Alabama at Birmingham, Birmingham, AL

## **Papillary Fibroelastoma: An Uncommon Cause for a Transient Ischemic Attack**

Ather Anis, MD;<sup>1</sup> Jennifer Brady, MD;<sup>1</sup> David Sedaghat, MD;<sup>1</sup> Mark Klapholz, MD;<sup>1</sup> Barry C. Esrig, MD;<sup>2</sup> Muhamed Saric, MD, PhD<sup>1</sup> From the Departments of Medicine<sup>1</sup> and Surgery,<sup>2</sup> New Jersey Medical School, Newark, NJ Address for correspondence: Muhamed Saric, MD, PhD, Director, Echocardiography Lab, Division of Cardiovascular Diseases, New Jersey Medical School, 185 South Orange Avenue, I-538, University Heights, Newark, NJ 07103 E-mail: saricmu@umdnj.edu

*Am J Geriatr Cardiol* **2005**;14(5):269-70.

#### Mitral Valve Papillary Fibroelastoma

#### Typically on the **left atrial side** of the valve



#### Mitral Valve Papillary Fibroelastoma



#### Mitral Valve Papillary Fibroelastoma



#### Typically on the **right atrial side** of the valve











Branching **avascular papillae** containing collagen and covered with endothelium



#### Rhabdomyoma



#### Rabdomyoma

The most common primary **benign** cardiac tumor in children. Often associated with **tuberous sclerosis**, a brain anomaly that leads to seizures.



## Rhabdomyoma



4-Chamber View

Subcostal View



#### Primary Cardiac Sarcoma



#### **Right Ventricular Sarcoma**

57-year-old woman admitted for weight gain and lower extremity edema



#### **Right Ventricular Sarcoma**

57-year-old woman admitted for weight gain and lower extremity edema



#### Right Ventricular Sarcoma

57-year-old woman admitted for weight gain and lower extremity edema



**Undifferentiated pleomorphic cardiac sarcoma** (formally called malignant fibrous histiocytoma)

#### CARDIAC SARCOMAS

- Angiosarcoma
- Rhabdomyosarcoma
- Leiomyosarcoma
- Undifferentiated pleomorphic

#### **Embolism From Aortic Tumors**

#### *Typically from a malignant* tumor



56-year-old, previously healthy woman presented with a 2-week history of **headache** and **progressive right-sided weakness** 



**Fig. 1** Magnetic resonance image shows a  $4.3 \times 3.8 \times 4.3$ -cm, dural-based, left parafalx gadolinium-enhanced mass (asterisk) with surrounding edema (arrow) and mass effect.



**Fig. 4** Continuous-wave spectral Doppler image from the suprasternal notch shows abnormal coarctation-like flow in the descending thoracic aorta during systole and diastole.







#### Aortic Intimal Sarcoma: MRI





Case Reports

Alicia Mecklai, MD Barry Rosenzweig, MD Robert Applebaum, MD Leon Axel, MD, PhD Eugene Grossi, MD Alexander Chan, MD Muhamed Saric, MD, PhD

# Intimal Sarcoma in the Aortic Arch

Partially Obstructing the Aorta with Metastasis to the Brain

Primary tumors of the aorta are rare entities. We report the unusual manifestation of an aortic intimal sarcoma that presented as a brain metastasis in a 56-year-old, otherwise healthy woman. After the brain mass had been resected, multiple imaging methods revealed pseudocoarctation and the primary tumor in the aortic arch. To our knowledge, this is the first report of the diagnosis of an aortic intimal sarcoma with use of real-time, 3-dimensional transesophageal echocardiography. **(Tex Heart Inst J 2014;41(4):433-6)**
### Secondary Cardiac Malignancies



#### **Breast Cancer**



#### **Breast Cancer**

47-year-old woman with breast cancer and malignant pericardial effusion



#### **Breast Cancer**

47-year-old woman with breast cancer and malignant pericardial effusion









# Lung Cancer

59-year-old man; active tobacco smoker, with lung cancer extending into the heart through left-sided pulmonary veins



# Lung Cancer

59-year-old man; active tobacco smoker with lung cancer extending into the heart through left-sided pulmonary veins



# Lung Cancer

59-year-old man; active tobacco smoker with lung cancer extending into the heart through left-sided pulmonary veins



## Hepatocellular Carcinoma



#### **Case Presentation**

#### 61-year-old man

- : Chronic ethanol abuser
- : Brought in by his wife because of altered mental status

: TTE order to evaluate for murmur



#### **2D TEE** Apical 4-Chamber View

# RA Mass | Hepatocellular Carcinoma



TTE: Short-Axis View at AV Level Mass in RA originating in IVC **TTE: Subcostal View** Mass in the liver extending into IVC

## RA Mass | Hepatocellular Carcinoma



**Immediately Post Definity** No significant contrast uptake by RA mass **Delayed Imaging** Contrast uptake by RA mass indicative of a vascularized tumor

# Final Diagnosis

Hepatocellular carcinoma













#### Renal cell carcinoma with extension to the heart

Ather Anis MD<sup>1</sup>, Pierre Maldjian MD<sup>2</sup>, Marc Klapholz MD<sup>1</sup>, Muhamed Saric MD PhD<sup>1</sup>



**Figure 1)** Magnetic resonance imaging of the abdomen showing a 9 cm × 8 cm right renal mass and a filling defect (tumour extension) in the inferior vena cava (thin arrow) with protrusion into the right atrium (thick arrow)

**Figure 2)** Transthoracic echocardiogram showing a large mobile mass (arrow) protruding from the inferior vena cava (IVC) into the right atrium. Ao Ascending aorta; RV Right ventricle



#### Cardiac Lymphoma vs. Thrombus



#### **Case Presentation**

47-year-old man

: Presents with acute stroke

: Several months ago had acute LAD infarct; not revascularized due to late presentation



**2DTEE** Apical 2-Chamber View

#### 2 Patient with LV Mass: Microbubble Injection



**Patient #1** LV thrombus post LAD infarct **Patient #2** LV lymphoma in an AIDS patient

#### **TEACHING POINTS**

#### Differential diagnosis of an LV mass

#### LVTHROMUS

- Adjacent to akinetic/hypokinetic LV segment
- 2. Does NOT take up microbubble contrast

#### LVTUMOR

- Typically no primary LV wall motion abnormalities
- 2. Typically DOES take up microbubble contrast



## ThankYou



New York University Langone Medical Center

#### Cardiac Masses





# Short Biography



Muhamed Sarić MD, PhD, MPA

- Born in Sarajevo, Bosnia-Herzegovina
- Director of Noninvasive Cardiology and Professor of Medicine at NYU
- Primary interest is the use of 3D echocardiography in guiding percutaneous repairs of structural heart disease. At NYU my colleagues and I performed the first transseptal transcatheter mitral valve replacement in the world on June 15, 2016 using Caisson valve system.
- First to describe the tilt-up-then-left or TUPLE maneuver, which improves the diagnosis of atrial septal defects (ASDs), and facilitates its repair.
- Published numerous articles and book chapters in the field of cardiology, biochemistry and history of medicine.
- Chairman of the American Society of Echocardiography (ASE) guidelines committee for the use of echocardiography in the evaluation of a cardiac source of embolism
- Recipient of multiple teaching awards including the 2017 Richard Popp Excellence in Teaching Award from the American Society of Echocardiography