Interesting Case Studies
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1. Dueling Myxomas
2. RV Thrombus in Transit
3. Double Chambered LV

American Society of Echocardiography

Dueling Myxoma

Outline
• Signs & Symptoms
• History
• CT / CTA
• Echo (TTE)
• Echo (TEE)
• Treatment
• Outcome

Patient History
• 69-year-old man
• Hx ischemic CMO
• Afib (anticoagulated)
• CAD, PCI/prox LAD /2 years prior
• EF 25% - 30%
• Single-lead ICD
• Hyperlipidemia, DM, and distant history of pulmonary embolism.
Signs and Symptoms

Presents with:
- Aphasia
- Right-sided weakness
- Right facial droop
- BP 130/90 mmHg
- Irregular rhythm with soft systolic murmur
- ECG - ventricular paced rhythm with underlying Afib

Head CT / CTA

- Head CT - no pathology
- CTA - Acute occlusion - M2 branch of middle cerebral artery

Diagnosis: Acute stroke
Cause: Known Afib with patient non-compliance of anti-coagulation meds

Treatment

- Not a candidate for tPA - hx anticoagulation
- Successful intra-arterial pharmacologic thrombolysis
  - *Intra-arterial thrombolysis* uses an endovascular microcatheter delivery system.
  - Catheter port is positioned immediately within and adjacent to the thrombus, and fibrinolytic agents are infused directly into the clot.
- Echo ordered

Echocardiogram (TTE)

- Chronic severely ↓ LVF
- Mildly ↓ RV function
- Mild - moderate TR
- LA mass (3.7-cm)
- Suggestive of myxoma

- PSAX
- LA: Smooth echogenic mass

- A4C - mass poorly visualized
- ICD wire seen in the RV
Echocardiogram (TTE)

- 3-chamber view
- Poorly visualized left atrial mass

Next Step

- TLS – No definitive diagnosis of Myxoma
- Thrombus vs. tumor??
- Cardiac MRI contraindicated due to implanted defibrillator
- TEE was ordered

TEE

2D TEE Findings

- Midesophageal short axis view
- Two smooth mobile masses consistent with myxomas
  1. Attached to the IAS *
  2. Attached overlying the orifice of the left atrial appendage (LAA) *
- Defibrillator wire in the right atrium

2D TEE

2D TEE Findings

- TEE Bicaval view
  - Myxoma with typical insertion
  - IAS - left atrial side
  - Defibrillator wire in RA
3D TEE Findings

• 3D TEE
• Myxoma attached to the IAS
• Visualization of the stalk.

2D TEE Finding

• Midesophageal view
• Smooth mass
• Perdunculated - characteristic stalk of a myxoma
• Attached near the left atrial appendage (LAA)
• Attachment point clearly identified
• Video: Rocking at attachment point above the left atrial appendage

3D Imaging Performed

3D TEE Findings

• Myxoma overlying the opening of the LAA
• Smooth borders consistent with myxoma
Treatment Plan

Patient was evaluated by cardiothoracic surgery due to:
- Distinctive characteristic findings of myxoma on TEE
- Location and embolic potential of the dual myxomas

Surgery

- Successful operation
- Histopathology:
  - soft, tan-pink cut surface
  - cystic-appearing mass
- Post-op uneventful
- Successful discharge to a skilled nursing facility

Myxoma – Surgical Images

- Intraoperative gross images
- Gross visualization of the two myxomas after surgical resection

Post-Op Echo

- Post-op echo demonstrated successful resection of the myxomas
- EF of 25% - 30% (previously reported)
- 6 month f/u echo
  - No recurrence of left atrial masses
  - EF 25% - 30%

Case Study Review

- Patient presented with signs of a stroke
- Echo & TEE - key diagnostic test in the management of stroke
- MRI was contraindicated due to the patient’s implanted defibrillator
- Echo was critical for diagnosis and plan of care

Case Study Review

- Initially, assumed the patient’s acute stroke was due to non-compliance of anticoagulation meds
  - Afib
  - Elevated risk scores
- TEE revealed a cardiac mass - not one but two left atrial myxomas!
Discussion: Myxoma

- Primary cardiac tumors are rare
- Myxomas account for almost half
- Myxomas are typically:
  - Pedunculated
  - Located in the LA
  - Vary in size
  - Usually only one tumor

Discussion: Myxoma

- Clinical manifestations include cardioembolic events
- Other symptoms include:
  - Weight loss, fever, laboratory abnormalities such as anemia and elevated C-reactive protein
  - Seen more in women

Discussion: Myxoma

- Recommendation treatment - Surgical resection due to the risk of embolization
  - TEE (especially 3D) allows precise localization of the attachment point of these tumors, allowing improved operative planning
  - Recurrence of tumor is rare
  - The presence of two primary cardiac myxomas is a rare finding, most often seen in patients with Carney Complex

Discussion: Carney Complex

- Rare endocrine disorder
- Characterized by multiple benign tumors, such as myxomas
- Spotty skin pigmentation
- Usually young men
- This patient (69M) did not have pigmented skin lesions, nor did he have evidence of any other endocrine tumors to suggest this diagnosis

Conclusion

- Myxomas are the most common primary tumors of the heart, most commonly located in the left atrium
- The discovery of multiple myxomas is a rare finding
- This case study demonstrates two left atrial myxomas diagnosed on transesophageal echocardiography (TEE)

End Case One
RV Thrombus in Transit
Pulmonary Embolism

Outline
• Signs & Symptoms
• History
• CT / CTA
• Echo (TTE)
• Echo (TEE)
• Treatment
• Outcome

Signs and Symptoms
• 40F
• 3 wks progressive dyspnea on exertion
• Normal cardiopulmonary exam in ER
• No signs of volume overload
• Hemodynamically stable but hypoxic

History
• HTN
• Obese
• Type 2 diabetes
• Unprovoked DVT (no known cause) and pulmonary emboli being treated with blood thinners
• Untreated pelvic mass of unknown etiology

Chest CTA
• Non-occlusive thrombi in various branches of the right and left pulmonary arteries
• Chronic emboli with interval improvement
• She was started on heparin and 4L supplemental oxygen
• Successful reversal of hypoxemia

Echocardiogram (TTE)
• Transthoracic echocardiography (TTE):
  • Mildly reduced RV function
  • RV fractional area change of 25%
  • TAPSE of 1.7 cm
  • New large RV mass suspicious for thrombus
• TEE was ordered
TEE: RV Inflow-Outflow View

**Large RV multi-lobed thrombus**

TEE: Long Axis View

- Large RV thrombus
- Multi-lobed
- Extending from TV to PV

3D TEE

- Large RV thrombus
TEE: TR

• TR Moderate-Severe
• Peak pressure gradient 94 mmHg
• Mean pressure gradient 46 mmHg

TEE: PFO

• 4 Chamber view
• Agitated saline
• PFO

TEE: Other Findings

• There was no evidence of PV stenosis/regurgitation
• Estimated right atrial pressure of 8 mmHg
• RVSP 102 mmHg
• Mean PA pressure 54 mmHg

Other Testing

• Lower Extremity Ultrasound
  • Chronic occlusive thrombi in various areas of the right lower leg
• Brain MRI
  • Multiple tiny, acute infarcts of the:
    • Bilateral cerebellar hemispheres
    • Bilateral parietal lobes
    • Left frontal lobe
• Neurological Exam
  • Normal neurological exam

Brain MRI

• Multiple tiny, acute emboli
• Bilateral cerebellar hemispheres
• Parietal lobes
• Left frontal lobe

Other Findings

• 12-cm ovarian mass found 1 year before admission
• Patient did not follow-up
• CT now shows 15 × 10 × 11 cm cystic and solid right ovarian mass
• ↑ Cancer antigen-125
• Placed on a new blood thinner plus meds to ↓ PHTN
Treatment

- IV filter was placed to prevent any further emboli
- Repeat echo 5 days later:
  - Slight decrease in RV thrombus size
  - Improvement in RVSP to 55 mmHg
  - Mean PA pressure to 44 mmHg

Initial Treatment Plan

- Initially considered: Emergent thrombolysis, surgery, and ECMO (heart-lung bypass support)
- None of these options were pursued because:
  - Patient was currently hemodynamically stable
  - Patient was improving on anticoagulation alone
  - Patient was discharged with anticoagulant medication and plans for out-patient surgery for pelvic mass

Pelvis Surgery

- 45 days later – pelvis surgery without any immediate complications:
  - Tumor debulking
  - Total hysterectomy
  - Removal of ovaries and fallopian tubes
  - Pathology revealed cancer (adenocarcinoma)

Intraoperative TEE

- Intraoperative TEE, at time of surgery showed a stable RV thrombus
- RVSP of 47 mmHg
- Mean PA pressure of 33 mmHg

CTA

- New pulmonary emboli in the right upper lobe arteries (arrow), right middle lobe artery, and left lower lobe arteries

Post Op

- On the day after surgery, the patient was extubated successfully but subsequently developed progressive hypoxemia
- Chest x-ray showed clear lungs
- Chest CTA showed new pulmonary emboli:
  - Right upper lobe
  - Right middle lobe
  - Left lower lobe arteries
Repeat TTE

- Repeat TTE showed worsening right heart pressures
- RVSP of 91 mmHg and mean PA pressure of 56 mmHg
- Patient was restarted on anticoagulation (it had been stopped for surgery)

Treatment

- Decision to proceed with thrombectomy, based on:
  - Worsening pressures
  - Continued risk for pulmonary and systemic emboli
  - Potential hemodynamic compromise

Surgery (sternotomy)

- Remove RV thrombus
- Close PFO

Surgical Finding

- RV mass- entwined in TV chordae
- Did not appear to be a thrombus
- Concerning for metastatic disease

Post-op

- Pathology
  - Organizing thrombus without any malignant cells identified
- Postoperative TTE
  - No evidence of interatrial shunting
  - Showed improved RVSP 53 mmHg / mean PA pressure 41 mmHg
  - Attributed to debulking of the RV mass

Post-op

- Post-op the patient remained hemodynamically stable and demonstrated good oxygen saturation on 3L of oxygen
- She was discharged with anticoagulants, sildenafil (PHTN) and home oxygen
- After discharge:
  - Improved symptomatically
  - Remained clinically stable
  - Serial echo showed improving right heart pressures

Follow-Up Echo

- 9 month F/U echo after thrombectomy
  - Patient was asymptomatic
  - No signs of recurring thrombus
  - TTE showed RVSP of 26 mmHg
  - Normal RV size
  - Mildly reduced RV function
Discussion: Right Heart Thrombus

- Right heart thrombus
  - Uncommon condition (4%) of patients with pulmonary emboli
  - It is often found in transit - originating from a systemic vein source
  - May also form within the cardiac chambers - primary processes, such as Afib

- Right heart thrombi are associated with significantly increased mortality
  - 27% - 45% with treatment
  - Near 100% in untreated patients
  - About 98% of cases of right heart thrombi are associated with concurrent pulmonary emboli

Discussion: Treatment

- The treatment options for RV thrombus include:
  - Anticoagulation
  - Thrombolysis
  - Surgical thrombectomy
- Overall, the existing literature has shown conflicting evidence, with no clear consensus on management recommendations
- Several studies have suggested a mortality benefit using a more aggressive approach with either thrombolysis or surgery

Discussion: Therapeutic Dilemma

This case presented a particularly difficult and complex therapeutic dilemma:

- Severely elevated right heart pressures
- RV dysfunction
- Fragile clinical status
- High risk for hemodynamic instability
- PFO with resulting cerebral infarcts
- Pelvic malignancy

There was concern that either thrombolysis or surgery could cause the thrombus to dislodge, leading to hemodynamic compromise:

- Thrombolysis - could lead to hemorrhagic conversion of her strokes
- Immediate surgery - anticoagulation would have to be held in the setting of:
  - Significant pulmonary embolic burden
  - Large RV thrombus
  - Significant right heart strain

Initial Decision - anticoagulation alone:

- Potentially poor outcome of intervention
- Patient’s initial improvement:
  - ↓ right heart pressures
  - ↓ thrombus size
Discussion: Therapeutic Dilemma

Surgical decision:
- Patient developed recurrent acute pulmonary emboli
  - After anticoagulation was interrupted for pelvic surgery
- Patient had worsening right heart pressures
- Continued high risk for pulmonary and systemic emboli

Surgery:
- RV thrombus was surgically removed
- PFO was closed

Case Summary

- Despite this successful outcome, the optimal treatment for right heart thrombi remains uncertain and warrants additional studies
- Until there is more definitive evidence, management decisions should be made on a case-by-case basis, with careful consideration of complicating factors

End Case Two

Double Chambered LV

Outline
- Signs & Symptoms
- History
- ECG
- Echo (TTE)
- Echo (TEE)
- Angiogram
- Cardiac MRI
- Treatment/Outcome

Signs and Symptoms
- 49M
- History of alcohol abuse
- Type 1 diabetes on insulin
- Sudden onset of CP & profound palpitations while exercising
- Called 911 – initial rhythm was monomorphic v-tach 280 beats/min
- Converted to sinus rhythm w/ bolus of amiodarone bolus
ECG in the field

• Monomorphic ventricular tachycardia at the time of presentation (EMS)

ECG

• Admission 12-lead ECG
• Normal sinus rhythm
• Normal limits, without ST-segment elevations or Q waves

History

NO known history of:
• MI
• Cardiac trauma
• Illicit drug use
• Infection
• No family history of sudden cardiac death

Physical examination was unremarkable and subsequent normal ECGs

TTE Echo ordered

TTE Echocardiogram

Transthoracic echo showed
• Unusual appearance to the LV
• Suspicious structure/attachment originating from LV apex
• Apical lateral wall - thinned & akinetic

TTE Echo

• Unusual double-chambered appearance of the LV
• Accessory Chamber (AC)?
• Lateral wall - thinned and akinetic

3D Echo

• 3D Echo
• Short Axis
• Double-chambered appearance of left ventricle
Echo Contrast

- Double-chambered LV
- Primary LV cavity with accessory chamber
- Subdivided by muscular ridge originating from apex
- Free unobstructed communication between the primary LV cavity and accessory chamber

TTE ECHO Color

- Color demonstrates the absence of flow acceleration between the primary LV cavity and the accessory chamber
- Normally function MV without stenosis or regurgitation

3D Echo

1. Dyskinetic lateral wall of the accessory chamber
2. Papillary muscles visualized
3. Cross sectional apex showing the subdivision of the LV
4. Color – no gradient between primary and accessory chamber
3D Echo

Echo 3-Chamber

- Subdivision of the left ventricle by an anomalous muscular ridge

Parasternal Long Axis

- Structurally normal mitral valve.
- Contractility of the IVS and inferolateral walls were normal.

PSAX

- 2 distinct papillary muscles
- Anterolateral papillary muscle is dominant & larger than the posteromedial papillary muscle
Papillary Muscle

- Distinct asymmetry of the anterolateral (AL) papillary muscle
- The AL papillary muscle was significantly larger than the posteromedial papillary muscle
- AL papillary muscle appeared to be attached to the questionable muscular ridge
- This anatomy likely falls within the wide spectrum of parachute-like mitral valve variants without the typical valvular dysfunction that is associated with a parachute mitral valve

Angiogram

- Coronary angiogram did not demonstrate obstructive CAD

Angiogram

- Left ventriculography reveals a large LV outpouching lateral to the LV apex, consistent with the presence of an accessory chamber

Cardiac MRI

- Presence of a large muscle band that subdivides the left ventricle into a primary and an accessory chamber. The wall of the accessory chamber is thin and akinetic
- EF 42% - supports congenital DCLV rather than an aneurysm or a pseudoaneurysm

Cardiac MRI

- No late gadolinium enhancement to suggest scar from prior ischemic injury
TTE Findings

- LV cavity subdivided by a vertical tissue ridge in the center of the LV
- This suspected cavity has a similar thickness and appearance to myocardial tissue in both 2D and 3D
- There was no evidence of structural narrowing at the site of communication, rather a large visible gap by 2D & color Doppler
- Structurally normal MV with mild MR
- Lateral wall - thinned and akinetic
- No other wall motion abnormalities identified
- Global LVF mildly reduced, EF 45%

Differential Diagnosis

Echo findings seemed consistent with DCLV, differential diagnosis of:

- Pseudoaneurysm at a prior silent myocardial rupture site
- True aneurysm
- Parachute Mitral Valve

Differential Diagnosis: Pseudoaneurysm

- Pseudoaneurysms
  - Do not contain all three layers of cardiac tissue
  - Have a narrow neck
  - May exhibit paradoxical movement during systole
  - DCLV contain all layers of cardiac tissue that typically contract synchronously with the rest of the ventricle
- **Important** - Pseudoaneurysm typically requires urgent surgical management!

Parachute mitral valve

- Parachute mitral valve can also have a similar appearance, but two distinct papillary muscles were identified that received chordae tendineae that appeared to be of equal lengths from both leaflets
- The mitral valve opened normally in a symmetric manner, without any stenosis and with minimal regurgitation

Case Study Review

- This is the case of a man who presented with V-Tach diagnosed with Double Chamber Left Ventricle
- Echo and Cath suggested DCLV, but cardiac MRI was diagnostic

MRI Finding

- The absence of late uptake enhancement indicated a lack of a scar tissue which ruled out a prior silent MI
- No signs of fibrosis, ruled out trauma or infarction
- MRI showed thinned wall consisted of myocardial tissue, which excluded the possibility of a pseudoaneurysm

- Surgical resection was deferred given the close proximity of the defect to the papillary muscle and submitral apparatus
- ↑ risk for complication that could lead to MV replacement in a young patient who otherwise had a fairly normal mitral valve
- Patient received implantable cardioverter-defibrillator (ICD) for secondary prevention of sudden cardiac death
- The patient had a recurrence of V-tach 1 year later and his ICD successfully converted him
Conclusion

Double-chambered left ventricle (DCLV) is a rare congenital defect (0.04% to 0.42%)
• Characterized by division of LV chamber into two chambers by
  abnormal muscular tissue
• Generally asymptomatic without complications
• Can present with life-threatening arrhythmias

DCLV should be suspected by echocardiography in the presence of:
• Unusual aneurysmal appearance
• Subdivision of the LV by an anomalous muscular ridge
• The echo diagnosis is aided by the use of contrast and 3D
• MRI is vital to help rule out differential diagnosis
• The anomalous muscle ridge can cause the development of VTach

References

1. Myxoma: https://www.cvcasejournal.com/article/S2468-6441(18)30190-7/fulltext

Right Ventricular Thrombus in Transit: Raising the Stakes in the Management of Pulmonary Embolism.
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Thank you!

Slides / Hand-out available at

www.cardioserv.net/MCSS