Carcinoid

Sunday, February 14, 2016 | 7:00 – 7:15 PM | 15 min

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Disclosures

Speakers Bureau
Philips, Medtronic
In 1907, Siegfried Oberndorfer coined the term *Karzinoide* (based on καρκινώδης, later anglicized as carcinoid).

Karzinoide Tumoren des Dünndarmes. [Carcinoid tumors of the small intestine] 
*Frankfurter Zeitschrift für Pathologie* 
1907;1:426-429.

*Carcinoid = ‘carcinoma like’*

*Histologically, a malignant tumor...* 
*... but clinically often behaves like a benign nonmetastatic tumor*
Carcinoid Tumor vs. Carcinoid Heart Disease

What’s the difference?
Carcinoid Tumor

- **Malignant tumor** of neuroendocrine cells in the gut or bronchial tree
- **Slow growing** and occasionally metastatic
- Secretes **vasoactive substances & hormones** including serotonin (5-HT) resulting in **carcinoid syndrome**:
  - Facial flushing
  - Diarrhea
  - Carcinoid heart disease

What portion of the heart gets involved depends on the primary tumor’s location:

- **Intestinal** carcinoid >> Preferential **RIGHT** heart involvement
- **Bronchial** carcinoid >> Preferential **LEFT** heart involvement
Carcinoid Heart Disease

Carcinoid heart disease is NOT a cardiac malignancy.

- Carcinoid heart disease is a paraneoplastic syndrome caused by tumor-derived vasoactive substances.

Paraneoplastic manifestation of a carcinoid tumor
- Mural plaques
- Valvulitis >> Stenosis + Regurgitation

Because intestinal carcinoid are more common than bronchial ones, carcinoid disease involves primarily the right heart.

Carcinoid heart disease is often associated with liver metastases of an intestinal carcinoid.
Carcinoid Heart Disease

Case #1
Clinical Presentation

56-year-old previously healthy man
- Presented with right upper quadrant pain
- Referred for abdominal CT

Abdominal CT

Multiple liver metastases of an unknown primary tumor
TRANSTHORACIC ECHOCARDIogram | APICAL 4-CHAMBER VIEW

Dilated RV

Dilated RA

Noncoapting Tricuspid Valve
Noncoapting Tricuspid Valve

Dilated RV

Dilated RA

AV

LA
Transthoracic Echocardiogram | Parasternal Short-Axis View

Severe Tricuspid Regurgitation

Dilated RV

Dilated RA

AV

LA

HR = 61 bpm

Cardiac Easy
General
Pwr = 0 dB
Mlcd = 1.4
TIS = 1.7

T1/-2/ 0/V:A
1/2
CD: 2.5 MHz
CD Gain = 50

Store in progress
TRANSTHORACIC ECHOCARDIOGRAM | CW DOPPLER OF TRICUSPID VALVE

TR JET
Low velocity
Triangular shape
Severe Pulmonic Regurgitation
**Transthoracic Echocardiogram | Pulmonic Valve**

**Pulmonic Valve**

Frozen in semi-open, semi-closed position

**Pulmonic Regurgitation Jet**

Rapidly decelerating
Triangular shape

Diastole

Systole

To-and-Fro Flow
Carcinoid Heart Disease

Case #2
CLINICAL PRESENTATION

71-year-old female with 8-year history of metastatic carcinoid

- Severe abdominal pain & increased diarrhea x 2 weeks

- On somatostatin analog for many years

- For the last 3 years, she has been taking tincture of opium for her diarrhea with relatively good control; however, she stopped 4 days ago because she developed severe right upper quadrant pain when she took it.

- Since then, she has been taking at least 12 Lomotil antidiarrheal pills with poor diarrhea control.

**OCTREOTIDE**

Somatostatin analog used to counteract effects of carcinoid-derived hormones
TRANSTHORACIC ECHOCARDIOGRAM | RV INFLOW VIEW

Dilated RV

Dilated RA

TRICUSPID VALVE
Frozen in semi-open, semi-closed position
TRANSTHORACIC ECHOCARDIOGRAM | RV INFLOW VIEW

Dilated RV

Dilated RA

SEVERE TRICUSPID REGURGITATION

Laminar flow
TRICUSPID REGURGITATION JET

Low velocity
Triangular shape
Pulmonic Regurgitation Jet
Rapidly decelerating
Carcinoid Heart Disease

Case #3
57-year-old female with shortness of breath over the past 6 weeks.

- Previously unlimited exercise tolerance
- Progressive bilateral leg edema, increasing abdominal girth and loose bowel movement.
- Recently noticed intermittent flushing and shivering without fever

**Octreotide Abdominal CT Scan**

*Multiple foci of uptake within the liver and in a calcified mesenteric mass compatible with octreotide avid tumors such as carcinoid.*
Increased urinary excretion of serotonin metabolites, consistent with **carcinoid syndrome**.
TTE showed **severe tricuspid** and **pulmonic regurgitation**... and something else in subcostal view.
**Carcinoid Tumor**
Metastatic well differentiated neuroendocrine tumor

**Carcinoid Tumor**
Positive staining for chromagranin
consistent with carcinoid
CARCINOID HEART DISEASE

Case #4
Clinical Presentation

64-year-old men with recent weight loss and bilateral lower extremity edema

• A year ago, he noticed some flushing and rosy appearance to his cheeks
• Six months ago he started having diarrhea
• Abdominal MRI
  • Mesenteric soft tissue mass
  • Multiple hepatic metastases
• Liver biopsy
  • Carcinoid tumor

MRI | Mesenteric Carcinoid

MRI | Innumerable Liver Metastases
3D TRANSTHORACIC ECHOCARDIOGRAM | BIPLANE VIEW

Dilated RV

Dilated RA

Frozen Tricuspid Valve

Dilated RV

Dilated RA
Dilated RV

Dilated RA

Severe Tricuspid Regurgitation
TRANSTHORACIC ECHOCARDIOGRAM | SUBCOSTAL VIEW

- Dilated RA
- Hepatic Vein
TRANSTHORACIC ECHOCARDIOGRAM | HEPATIC VEIN

Systolic Flow Reversal (sign of severe TR)
3D Transthoracic Echocardiogram | LV Perspective

- Frozen Tricuspid Valve
- Normal Mitral Valve
Surgery for Carcinoid Valve Disease

Underwent surgical replacement of both tricuspid & pulmonic valve

33-mm Edwards Magna Pericardial TVR

21-mm Sorin Pericardial PVR
TRANSTHORACIC ECHOCARDIOGRAM | TRICUSPID BIOPROSTHESIS

RV

TV
Bioprosthesis

RA
Subcostal View

TV Bioprosthesis
TRANSTHORACIC ECHOCARDIOGRAM | PULMONIC BIOPROSTHESIS

Pulmonic Bioprosthesis
TRANSTHORACIC ECHOCARDIOGRAM | PULMONIC BIOPROSTHESIS
CASE #4 EPILOGUE

At present, 4 years after TVR & PVR, patient is doing well on octreotide.
CARCINOID HEART DISEASE

Case #5
Carcinoid Heart Disease

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Thank you

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