Clinical Pathologic Correlations – Case Three

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Case Three – History
- 24 year old female
- Previously healthy
- 4 week history
  - Fatigue
  - Dizziness
  - Shortness of breath
  - Ankle swelling

Case Three – Road Map

Additional History
- Gravida 1, para 1
- Uneventful pregnancy 4 years earlier
- Social
  - Smokes "occasionally" since age 15
  - Denies drug or alcohol abuse
- Medications
  - Oral contraceptives since delivery
Additional History (continued)

- PMH
  - Usual childhood illnesses
  - No history of rheumatic fever
  - Routine childhood checkups

Characterizing Symptoms

- Fatigue
  - "No energy" worsening over 4 weeks
  - Now just wants to "sit and do nothing"
  - Restless, difficulty sleeping
- Dizziness
  - Light headed without syncope
  - Non-vertiginous

Characterizing Symptoms (cont'd)

- Shortness of breath
  - Worsening over 4 weeks
  - Initially with moderate exertion
  - Now, unable to pick up her four year old
  - Insignificant cough, not productive
  - No wheezing

Characterizing Symptoms (cont'd)

- "Ankle swelling"
  - Poor appetite
  - Can eat only small amounts of food at a time
  - Despite this, she has noted a 20 pound weight gain during this time

Despite this, she has noted a 20 pound weight gain during this time.
Review of Systems

Negative
- Fevers, chills, rashes, wheezing, chest pain, palpitations
- Temperature intolerance, flushing

Positive
- Occasional diarrhea, now needs a second pillow to sleep at night

Physical Examination

Afebrile, pale, diaphoretic
60”, 195 pounds
Pulse 120, regular
Respirations 24, labored
Blood pressure
- Laying 100/60
- Sitting 75/50

Respiratory
- No wheezes, rales or ronchi

Cardiovascular
- JVP - 16 cm of water
- No significant "v" wave
- No murmurs

Abdomen
- Protuberant
- Ascites with + fluid wave
- Liver
  - 5 finger-breadths below right costal margin

Extremities
- 4+ pitting edema to the sacrum
Laboratory Examination

- WBC, Hb, and Hct - normal
- Electrolytes, BUN, Cr
  - K - 5.4 meq/ml
  - BUN - 98
  - Creatinine - 7.8 mg/dl

Radiographic Studies

- Chest X-Ray
  - No congestion or pulmonary edema
  - Prominent azygous vein
  - No cardiomegaly

Symptom Progression

- Patient develops severe hypotension
- Resuscitative measures
- A procedure is emergently requested...

Emergent Echocardiography

- Surface study
  - LV size & function
    - Normal
  - RV size & function
    - Normal
  - Normal aortic, mitral, and pulmonic valves
  - Tricuspid
    - Inflow gradient 12 mmHg (doppler)
    - ?????
Hospital Course

- Intractable hypotension develops
- PEA
- Aggressively coded
- Expires

A Medical Autopsy is Requested

Differential Diagnosis

- Rheumatic tricuspid stenosis
- Right atrial myxoma
- Carcinoid heart disease
- Metastatic cardiac tumor

Autopsy Findings

Tricuspid

RV
Additional Autopsy Findings
- Hepatomegaly
- Ascites and extremity edema
- Right mainstem bronchus
  - 2cm firm white nodule
  - Proximal parabronchial

Histology of Tricuspid Mass

Diagnosis - Cause of Death
- Metastatic carcinoid tumor to tricuspid valve
- Tricuspid stenosis, severe
  - Progressive right heart failure
  - Progressive edema
  - Progressive preload reduction
  - Hypotension
  - Renal failure
  - Death

Carcinoid
- Carcinoid tumor
- Carcinoid syndrome
- Carcinoid heart
Carcinoid Tumor

- Tumors with malignant potential
  - Location & invasion dependent
- Appendiceal & rectal
  - Appendix most common
  - Infrequent metastasis
- Ileal, gastric, and colonic
  - Through >½ muscularis – 90% metastasize
- Many other

Carcinoid Syndrome

Common

- Vasomotor disturbances
  - Cutaneous flushes and cyanosis
- Intestinal hypermotility
  - Diarrhea, cramps, nausea, vomiting

Infrequent

- Hepatomegaly
- Metastasis
- Systemic fibrosis
  - Cardiac
  - Retropertoneal
  - Pelvic

Carcinoid Syndrome

- Incidence
  - 1% of all patients with carcinoid
  - 20% with metastatic disease
- Etiology
  - Uncertain
  - Serotonin (5-HT, hydroxytryptamine)
    - Inactive metabolite (5-HIAA) in blood/urine
    - GI tumors must have liver mets for syndrome to occur

Carcinoid Heart

- Indirect effect of carcinoid syndrome
  - Pulmonic and tricuspid valves
  - Thickening
  - Stenosis
- Endocardial fibrosis
  - Right ventricle
  - Bronchial carcinoids affect left side
Metastatic Cardiac Tumors

- Most frequent
  - Lung, breast, melanoma, leukemia & lymphoma
- Usually clinically silent
  - Most common - pericardial effusion
  - Restrictive pericardial effect
- Spread direct, hematogenous, or lymphatic