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
- Characterization of Symptoms
- Review of Systems
- Physical Examination
- Laboratory Tests
- Radiographic Studies
- Clinical Course

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
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

Physical Examination

- 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-breathths 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)
    - ?????
Echocardiography: Subcostal View

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
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 >1/2 muscularis – 90% metastasize
- Many other

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 Syndrome

- Common
  - Vasomotor disturbances
    - Cutaneous flushes and cyanosis
  - Intestinal hypermotility
    - Diarrhea, cramps, nausea, vomiting
- Infrequent
  - Hepatomegaly
  - Metastasis
  - Systemic fibrosis
  - Cardiac
  - Retroperitoneal
  - Pelvic

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