

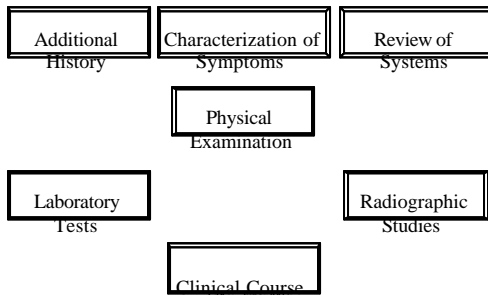
Clinical Pathologic Correlations – Case Three

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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



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



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Physical Examination

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

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Physical Examination

- Respiratory
 - No wheezes, rales or ronchi
- Cardiovascular
 - JVP – 16 cm of water
 - No significant "v" wave
 - No murmurs

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Physical Examination

- Abdomen
 - Protuberant
 - Ascites with + fluid wave
 - Liver
 - 5 finger-breadths below right costal margin
- Extremities
 - 4+ pitting edema to the sacrum



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Laboratory Examination

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



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Radiographic Studies

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



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Symptom Progression

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

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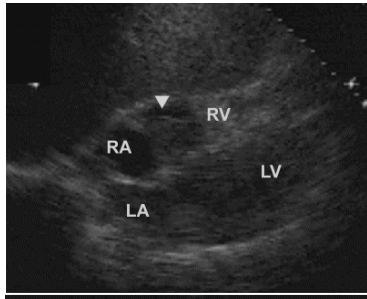
Emergent Echocardiography

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

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Echocardiography: Subcostal View



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Hospital Course

- Intractable hypotension develops
- PEA
- Aggressively coded
- Expires

**A Medical Autopsy
is Requested**

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Differential Diagnosis

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

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Autopsy Findings



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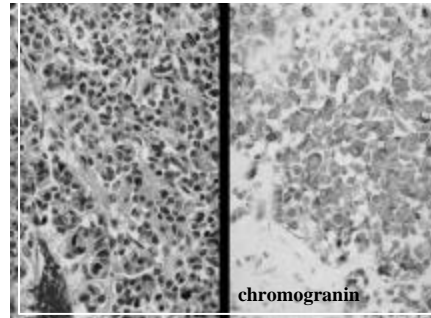
Additional Autopsy Findings

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

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Histology of Tricuspid Mass



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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

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Carcinoid

- Carcinoid tumor
- Carcinoid syndrome
- Carcinoid heart

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Carcinoid Tumor

- Tumors with malignant potential
 - Location & invasion dependent
- Appendiceal & rectal
 - Appendix most common
 - Infrequent metastasis
- Ileal, gastric, and colonic
 - Through $> \frac{1}{2}$ muscularis ~ 90% metastasize
- Many other

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Carcinoid Syndrome

Common

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

Infrequent

- Hepatomegaly
 - Metastasis
- Systemic fibrosis
 - Cardiac
 - Retroperitoneal
 - Pelvic

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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

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Carcinoid Heart

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

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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