Thinking on systems

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- Former smoker (1 year). No treatment.
- First episode of severe anemia (Hb 6.4 g / dl) in January 2010. Transfusion required.
- Blood tests: iron deficiency:
 - MCV 77.1, MCH 23.9.
 - Reticulocytes 269,000 (6.93%).
 - Ferritin 95.3, Iron 7.
 - Transferrin 325, Transferrin saturation 7.2%.

51 year-old man

- Endoscopy: small hiatal hernia, internal hemorrhoids, no evidence of recent bleeding.
- Lung CT:
 - Patchy infiltrates in "ground glass"
 - numerous paratracheal, mediastinal, prevascular, perihilar and lower lobe levels, lymphadenopathy.
 - without forming clusters or associate necrosis.
 - Some infiltration associated calcium.

Diagnostic tests

- Endoscopic capsule: duodenal lymphangiectasia.
- **Pulmonary CT** (at 6 months): parenchymal pneumonitis.
- Spirometry: restriction pattern (FVC 80%; FEV1 81.3%, Tiffeneau Index 0.82).
- CO diffusion test: normal.

Diagnostic tests

- New progressive dyspnea of two weeks' duration, cough, occasional hemoptysis and anemia. No chest pain nor fever. January 2012.
- New emergency transfusion (Hb 7.1 g/dl).
- Scintigraphy labeled with Tc⁹⁹ erythrocytes: no radiotracer extravasation in the digestive tract.
- **High-resolution CT:** extensive pulmonary ground-glass attenuation, in lower and middle lobe. Subpleural consolidation areas in basal segments of the right lower lobe. No significant lymphadenopathy.

New episode

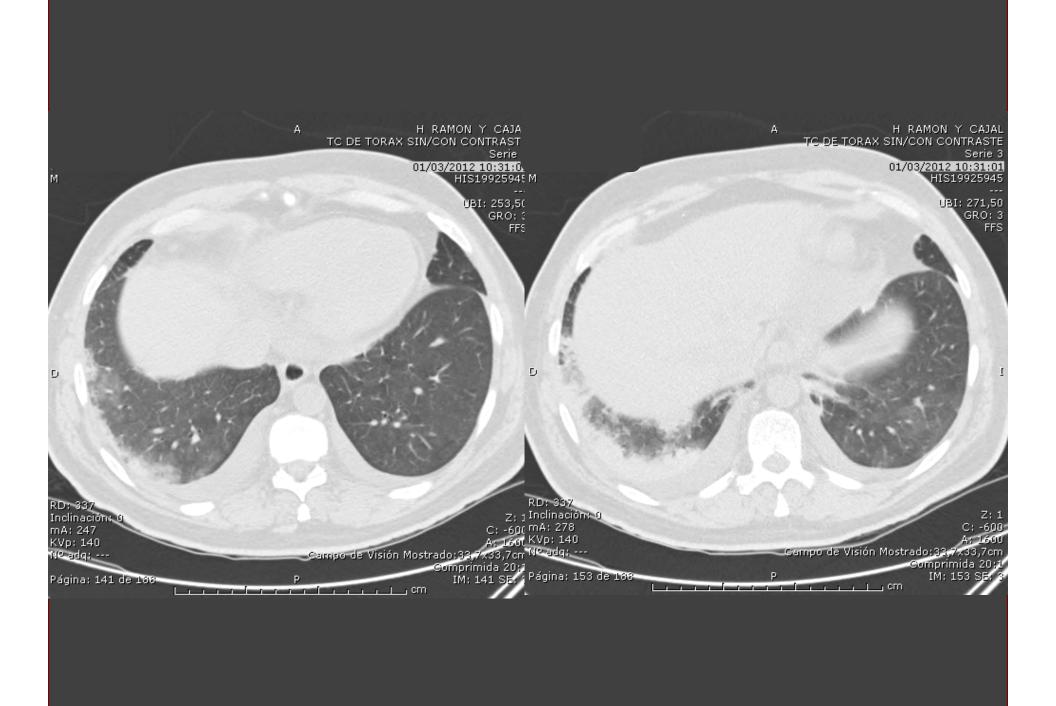


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- Mild chronic inflammation of the bronchial mucosa
- Bronchoalveolar lavage cell count:
 - 68% macrophages,
 - 30% neutrophils,
 - 2% lymphocytes and
 - abundant siderophages.
- Transbronchial biopsy: hemosiderophages in the lung parenchyma.

Bronchoscopy

- Renal biochemical profile, liver, lipid and thyroid is normal.
- Immunological study, including ANA, ANCA, complement and anti-GBM antibodies is negative.
- Antiphospholipid antibodies are negative.
- Quantification of immunoglobulin is normal.
- Serology for parvovirus B19, HIV, HBV and HCV are also negative.
- ESR 67 and C-reactive protein 38.

Other test results

Differential Diagnosis

- Idiopathic Pulmonary Hemosiderosis.
- Secondary causes :
 - congestive heart failure
 - pulmonary infarction
 - lung hemorrhage:
 - Goodpasture syndrome.
 - Granulomatous polyangiitis.

- Severe biventricular systolic dysfunction (ejection fraction 33%)
- Pulmonary hypertension without significant valve abnormalities (PSP 49)
- No pericardial effusion, left atrium 42 ml/m2, TAPSE 14.8 mm.
- Coronary angiography is requested to complete the study of ischemic heart disease.

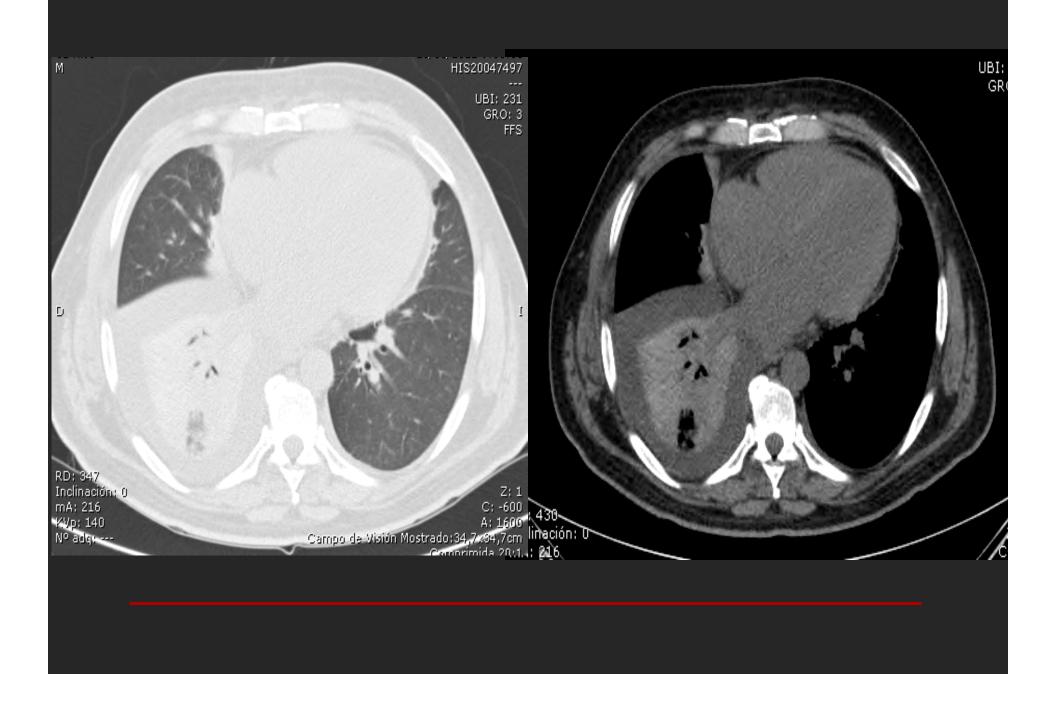
Transthoracic echocardiography

- Third day of admission.
- After starting with mild symptoms of right heart failure (perimaleolar edema in both legs)...
- Cardiac arrest while sleeping
- Non responsiveness to advanced resuscitation.

Outcome

- Marked facial cyanosis
- Pretibial pitting edema in the lower limbs
- Massive right hydrothorax: serous fluid, brownish yellow tinge (2 liters)
- Serous effusion pericardial sac (30 ml)
- Dilated right auricle
- Mediastinal lymphadenopathy of 1 cm
- Gallbladder lithiasis
- Right adrenal cortical neoplasm, 1 cm in diameter.
- Atheromatous plaques with complicated aspect at abdominal aorta level.

Postmortem examination



- Pulmonary thromboembolism and Pulmonary hypertension.
- Hemorrhagic infarction in both lungs, probably related to embolicischemic phenomena
- Abundant hemosiderophages
- Massive hydrothorax
- Heart: images of old and parenchymal infarcts and recent onset predominantly subendocardial. Left ventricular thrombus in right appendage
- Coronary arteries with atheromatous foci that in no case the gauge decreases below 50%
- Generalized arteriosclerosis
- Incidental papillary microcarcinoma in the left thyroid lobe
- Cortical adenoma of right adrenal gland of 1.2 cm.

Pathology (mycro)

- Silent heart attacks led to the situation of heart failure with of secondary *cor pulmonale*.
- Presence of abundant hemosiderophages in alveoli.
- Recurrent thromboembolism \rightarrow pulmonary hypertension.
- Immediate cause of death: microvascular thromboembolic disease of the lungs + right massive hydrothorax.

Cause of death