CLINICAL CASE

Colleagues from Portugal: Gonçalo Sarmento Ricardo Fernandes Sandra Morais (presenting)





History of



Chronic obstructive pulmonary disease • GOLD category C, stable in the past 2 decades



High blood pressure





Chronic atrial fibrillation (CAF)



Chronic right heart failure (HF) •Classified as New York Heart Association functional class II



60 pack-year history of cigarette smoking in the past



Pulmonary Hypertension due to lung disease (group 3)

•Transthoracic echocardiograph: "moderate to severe dilation of both atria... Mild to moderate tricuspid insufficiency. Pulmonary artery systolic pressure (PASP) of 60 mmHg..."



MARCH OF 2012

Physical examination



MARCH OF 2012

Laboratory tests

- Leukocytes count of 4100/µl, 59.7% neutrophils
- Hemoglobin 12.2 g/dL
- C-reactive protein 5.6 mg/l (Normal < 5 mg/l)
- Brain Natriuretic Peptide (BNP) level elevated (577.4 pg/mL; Normal < 300 pg/mL).



Electrocardiography - AF (63/min), no significant ST-T abnormalities

 Admitted to our hospital with severe right heart failure.



2012



Mediastinal enlargement



POSSIBLE CAUSES OF MEDIASTINAL ENLARGEMENT?

What tests should be performed?



The patient was admitted to the medical ward with the diagnosis of heart failure and was started on high dose diuretics with significantly clinical improvement.



Thoracic CT scan February/2011

Thoracic angio CT scan March/2012



Pronounced dilation of pulmonary trunk (72 mm), and a principal pulmonary arteries enlargement (right - 40 mm, left - 33 mm)

GIANT (> 50 MM) PULMONARY ARTERY ANEURYSM





- Congenital cardiac malformations with intracardiac shunt
- Vascular abnormalities such as arteritis (e.g., Takayasu disease) and primary pulmonary hypertension
- Vasculitis (e.g., Behçet's disease)
- Connective tissue disorders (Marfan, Ehlers-Danlos and Hughes-Stovin syndrome and systemic lupus erythematosus)
- Tuberous sclerosis
- Atherosclerosis
- Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu Disease)
 - Vascular trauma **No history of major thoracic trauma in the past**
- Infections (such as tuberculosis, syphilis, bacteria or fungi)

No history of significant clinical infection in the past: Serology for HIV 1 and 2 and VDRL-TPHA test were negative

ANA autoantibodies were negative

GIANT (> 50 MM) PULMONARY ARTERY ANEURYSM



GIANT HIGH-PRESSURE PULMONARY ARTERY ANEURYSM



Many authors defend an aggressive surgical approach in a presence of severe complications

- Airway compression
- Pulmonary artery dissection
- Intravascular thrombosis

Presented case

- 20 years evolution without any significant complication
- Favoring a conservative approach even for high-pressure uncomplicated PAA





THANKS FOR LISTENING

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