Case report

Claudiu Onogea, MD

Colentina Hospital, Bucharest, Romania

55 year-old female, 50 kg, non-smoker

Initially dry, than productive cough (4 months)

Exertional dyspnea (2 months)

Chest pain (recently)

Fatigue

Clinical exam

- Telangiectasia
- Digital ulcers and pitting at the fingertips
- Thick, hardened skin
- Discreet central cyanosis
- Midsystolic murmur at the apex (grade III/VI)
- Fine inspiratory crackles at the lung bases
- BP=120/60 mmHg, HR=90 bpm
- SaO2=95%

Known Clinical History

- Systemic sclerosis – diffuse form
  - Skin (Rodnan score=23)
  - Vascular – Raynaud phenomenon
  - Gastrointestinal – esophageal hypomotility
- Treatment before admission:
  - Methylprednisolone 0,5 mg/kg/day
  - Pentoxifylline 400 mg x 3/day
  - Nitroglycerin 1 tablet as needed
  - Different Antibiotics

Which is the right answer?

- Interstitial lung disease (pulmonary fibrosis) ?
- Infection of the RT
- Pulmonary vascular disease ?
  - Pulmonary arterial hypertension
  - Thrombosis of the pulmonary vessels
- Cardiac disease ?
  - Secondary to PAH (heart failure)
  - Myocardial fibrosis (restrictive cardiomyopathy)
  - Conduction system disease and arrhythmias
  - Pericardial disease

Investigations

- Laboratory:
  - Leukocytosis with neutrophilia
  - Polycythemia (RBC=5.7mil/ml, HGB=16.6g/dl)
  - Hypercholesterolemia
  - No inflammatory syndrome
  - d-Dimer – negative
  - pro-BNP – 20 x ULN
  - GOT, GPT – 1.1 x ULN
  - CPK=1.1 x ULN, CPK-MB – normal, troponins (-)
- ECG:
  - sinus rhythm, 96bpm, QRS +10° axis
  - LVH
  - ST-T wave abnormalities suggestive of anterior subendocardial myocardial ischemia
  - Minor intraventricular conduction abnormality.

Investigations

- Chest X-ray: slightly enlevated ICT, otherwise normal.
- Sputum: normal.
- Spirometry test: suggestive for mild obstructive disease.
- DLCO: 70% of predicted (mild decrease)
- Fundoscopy: retinal arteriolar narrowing

Investigations

- HRCT-scan:
  - mild pulmonary fibrosis
  - no thrombosis of the pulmonary vessels
  - normal size of pulmonary arteries

Investigations

- 24h Holter monitor:
  - sinus rhythm with normal variability;
  - a few isolated VPBs;
  - no other conduction abnormalities or arrhythmias;
  - ST-T wave abnormalities suggestive of myocardial ischemia;
  - QTc > normal at 40% of QRS complexes.
  - BP within normal range.

Investigations

- Doppler TT echocardiography:
  - Ao=24mm, LA=32mm, LV=30/15mm; IVS=18mm, LV posterior wall=15mm (asymmetric), RV 22mm, RA 25mm
  - Normal systolic function
  - Resting outflow gradient LV=60mmHg (N=30mmHg), LV outflow tract obstruction during systole
  - Hyperkinetic RV with outflow tract obstruction during systole
  - Diastolic dysfunction (impaired relaxation)
  - No tricuspid regurgitation. Mitral regurgitation
  - Normal pericardium.

Diagnostic

- Obstructive hypertrophic cardiomyopathy
- Systemic sclerosis – diffuse form – with skin, vascular, gastrointestinal, pulmonary involment

Treatment

- Verapamil 240mg/day
- Gradual lowering of the Methylprednisolone dose
- Lower dose of Pentoxifylline (400 mg x 2/day)
- NTG ointment

Evolution

- Real improvement of dyspnea
- Relief of chest pain
- 3 months after: improvement of outflow tract obstruction and IVS hypertrophy

Key points

- No relatives with HCM, not an athlete
- No aortic stenosis; Fabry disease – unlikely
- Good response to high doses of Verapamil and lowering the dose of vasodilator drugs
- A few cited cases in literature, but no causality proven
- HOC is associated with HLA DR3, common in SSc
- Cold season – worsening of Raynaud phn.
- Without serious renal involvement
- Hypertension??? – normal BP due to outflow tract obstruction??
- High doses of corticosteroids
- No salt diet ???