EOSINOPHILIC MYOCARDITIS

WITH LEFT VENTRICULAR APICAL ANEURYSM

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Challenging Case Reports from the ESC Cardiologists of Tomorrow
• Declaration of interest: Nothing to declare
A 27-year old male patient from the island of São Tomé was evacuated to Portugal and admitted in our hospital.

Previous history:

- Cerebral malaria 7 years before admission, smoking and drinking habits
- No regular medication
Four months before admission:

- Oppressive, constant, chest pain, located in the precordium, without irradiation
- Increased with exertion and deep breathing
- Associated with profuse sweating

- Orthopnoea and nocturnal paroxysmal dispnoea
- No dyspnoea on exertion, lower limbs oedema, palpitations, nausea, vomiting, fever, rash or other symptoms
Admitted to a hospital in São Tomé e Príncipe:

- Medicated with propranolol, nitroglycerin, isosorbide dinitrate, aspirin, diclofenac, cimetidine and diazepam, with no pain relief

- Evaluated by physicians from a non-governamental organization

- Evacuated to Portugal for further diagnostic work-up
• Emergency department:
  
  – Physical examination: Patient tachycardic and on distress
  
  – Laboratory evaluation:
    ✓ Microcytic anaemia (Hb = 11.0 g/dL)
    ✓ Elevated CRP (9.6 mg/dL) without elevated WBC or neutrophil count
    ✓ No eosinophilia (330 cells/μL; 4.0% WBC)
    ✓ Elevated liver enzymes (AST = 70 U/L, ALT = 95 U/L) and LDH (724 U/L)
    ✓ Negative myocardial necrosis biomarkers
• Emergency department: 12-lead ECG
Case Report

- Emergency department: Urgent TTE
Eosinophilic myocarditis with LV apical aneurysm

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Case Report

• Emergency department: Urgent TTE
• Emergency department: Urgent TTE
During admission:

- Chest pain persisted and epigastric pain and hiccups developed

- Serial laboratory evaluation:
  - Eosinophil count = 10 – 2640 cells/μL
  - Cardiac troponin I = 0.04 – 0.15 ng/mL
• During admission: 2D speckle tracking echocardiography
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Case Report

- 5th day of admission: Cardiac catheterization
Eosinophilic myocarditis with LV apical aneurysm

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Case Report

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Case Report

• 5th day of admission: Chest and abdomen CT
Eosinophilic myocarditis with LV apical aneurysm

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Case Report

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Case Report

- 5\textsuperscript{th} day of admission: Chest and abdomen CT
Case Report

- 5th day of admission: Endoventricular repair
5th day of admission: Endoventricular repair
• 5th day of admission: Endoventricular repair
Pathological examination:
• 17th day of admission:
  
  – Aorto-distal left renal artery bypass

  – Partial removal of the left renal artery aneurysm

  – Left renal artery ostium closure
Pathological examination:

- Arterial wall necrosis
- Chronic inflammation with neutrophils, xanthomatous cells, hemosiderophages
- Extensive fibrosis
- No microorganisms or vasculitis
Eosinophilic myocarditis

- LV apical aneurysm
- Pseudoaneurysm
Serological tests:

- Raised titers of *Chlamydia pneumoniae* IgA; Negative IgG

- Equivocal Coxsackie virus antibodies

• Microbiology exams:

  ─ Negative blood and urine cultures

  ─ Negative stool parasitologic exam

  ─ Negative LV wall, LV thrombus, pericardial fluid, renal artery aneurysm wall and thrombus cultures
• Bone marrow examination:
  
  — Normocellular bone marrow

  — Increased eosinophil count (15% total cells)

  — Erythrocitic and granulocytic precursors and megakaryocytes with no morphological abnormalities

  — No atypical cells
Case Report

- **Autoimmunity screening:**
  - Negative anti-nuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies (ANCA), anti-double stranded DNA antibodies, SS-A and SS-B antibodies, anti-Smith antibodies, anti-U1 RNP antibodies, anti-histone antibodies and anti-Jo1 antibodies

- **Other tests:**
  - Negative FIP1L1-PDGFRα fusion gene screening
  - No thrombofilia disorder
Post-operative course:

- Prolonged inotropic support, mechanical ventilation and CVVHDF
- *Klebsiella pneumoniae* VAP with severe sepsis

Discharged home on the 41st day of hospitalization
Follow-up: TTE
• Follow-up: TTE
Follow-up: Cardiac MRI
Follow-up: Cardiac MRI
Follow-up: Brain, chest, abdomen and pelvis CT angiography
• Follow-up:

  14 months

  No cardiac symptoms or events

  Laboratory evaluation: Eosinophil count = 1080 – 1920 cells/μL

  Medicated with prednisone 10 mg/day, ramipril, carvedilol and warfarin → aspirin
• Hipereosinophilia:
  - Parasitic infections
  - Allergic reaction
  - Less common aetiologies (leukaemia, lymphoma, solid tumours, myeloproliferative disorders, connective tissue disorders, vasculitis, other infectious diseases and cutaneous disorders)
  - Idiopathic

  - Cardiac involvement in 80% of patients
Eosinophilic cardiopathy or Eosinophilic endomyocardial disease:

- Acute necrotizing stage: eosinophilic myocarditis
- Thrombotic stage
- Fibrotic stage → endomyocardial fibrosis

Eosinophilic myocarditis:

- 0.5% unselected autopsy series
- 57 cases in adults published since 1957
- Mean age 41.6 years (range: 19-83 years), 47% female
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Discussion

- **Etiology:**
  - Idiopathic 63%
  - Churg-Strauss syndrome 20%
  - Drug hypersensitivity 9%
  - Infections 7%
  - Malignancy 2%

- **Presentation:**
  - Heart failure 48%
  - AMI 22%
  - Chest pain 13%
  - Cardiogenic shock 11%
  - Syncope 4%
  - Sudden death 2%
  - Cardiac tamponade 2%
  - Fever 13%
  - 6 cases of VT / VF
  - 1 case of bradycardia
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Discussion

• Laboratory evaluation:
  — 20% with normal eosinophil counts
  — 21% eosinophil counts between 500 and 1500 cells/μL
  — Range: 0 – 31080 cells/μL

• Echocardiography:
  — Segmental or diffuse LV wall motion abnormalities
  — Increased wall thickness
  — LV or RV thrombus (5 patients)
  — Pericardial effusion (23 patients)
  — Isolated or predominant RV dysfunction
  — First description of a LV aneurysm
Therapy:
- Identification of aetiology
- Corticosteroids
- Immunosuppressive drugs, hydroxyurea, IVIG, IFN-α, mepolizumab
- Imatinib if FIP1L1-PDGFRα fusion gene is present
- Heart failure therapy, anticoagulation
- Mechanical haemodynamic support, heart transplantation

Prognosis:
- 19.6% mortality rate
- 3.6% required heart transplantation
In summary:

- Eosinophilic myocarditis

- LV apical aneurysm

- Idiopathic mild peripheral-blood eosinophilia
Eosinophilic Myocarditis

With Left Ventricular Apical Aneurysm

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