

Myocardial Ischemia in Wegener's Granulomatosis

Armen Yuri Gasparyan, MD

Armenian Association For Theraputists-Internists

Republic Centre For Familial Mediterranean Fever

Yerevan, Armenia


 WG is one of the most common vasculitides

 Prevalence: 3 in 100,000 population, M-F 3:2; peak incidence at 50-60


① Granulomatous lesions of the airways and the kidneys are predominant features

① Saddle-nose deformity, epistaxis, sinusitis, hemoptysis, cough and dyspnea, arthralgia, optic neuritis, peripheral neuropathy, and necrotizing glomerulonephritis

Fauci AS, Wolff SM. Wegener's granulomatosis: studies in eighteen patients and a review of the literature. 1973. Medicine (Baltimore) 1994; 73: 315-24

 In 1 study with 158 pts, cardiac manifestations (predominantly pericarditis) were in 10 pts (6%), and, of these, 3 pts had coronary artery involvement

Hoffman GS et al. Wegener granulomatosis: an analysis of 158 patients. Ann Intern Med 1992; 116: 488-98

 Other reports. Heart block; pericarditis, coronaritis, thrombosis, myocarditis, noninfectious endocarditis, valvular disease with vegetations, mitral and aortal insufficiency, systolic heart failure

① The results of autopsy. Cardiac abnormalities can be present in one third of the patients who die from non-cardiac causes

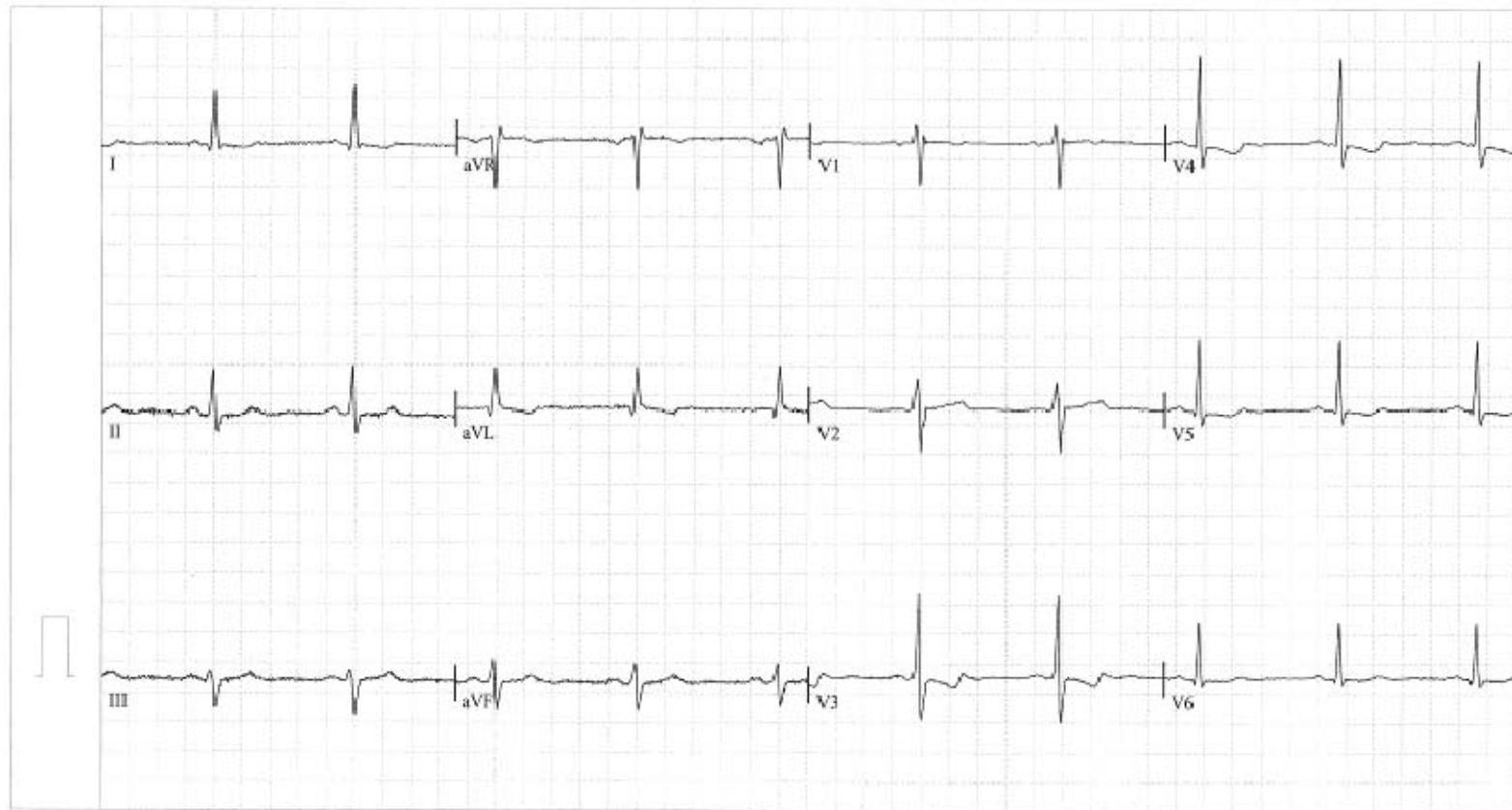
① Histology. Absence of immune deposits, presence of neutrophilic vasculitis, necrosis, microabscesses and granuloma. These changes are driven by antineutrophil cytoplasmic antibodies (ANCA) against cytoplasmic compound, proteinase-3 (cANCA)

Jennette JC, et al. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. Arthritis Rheum 1994; 37: 187-92

CASE PRESENTATION

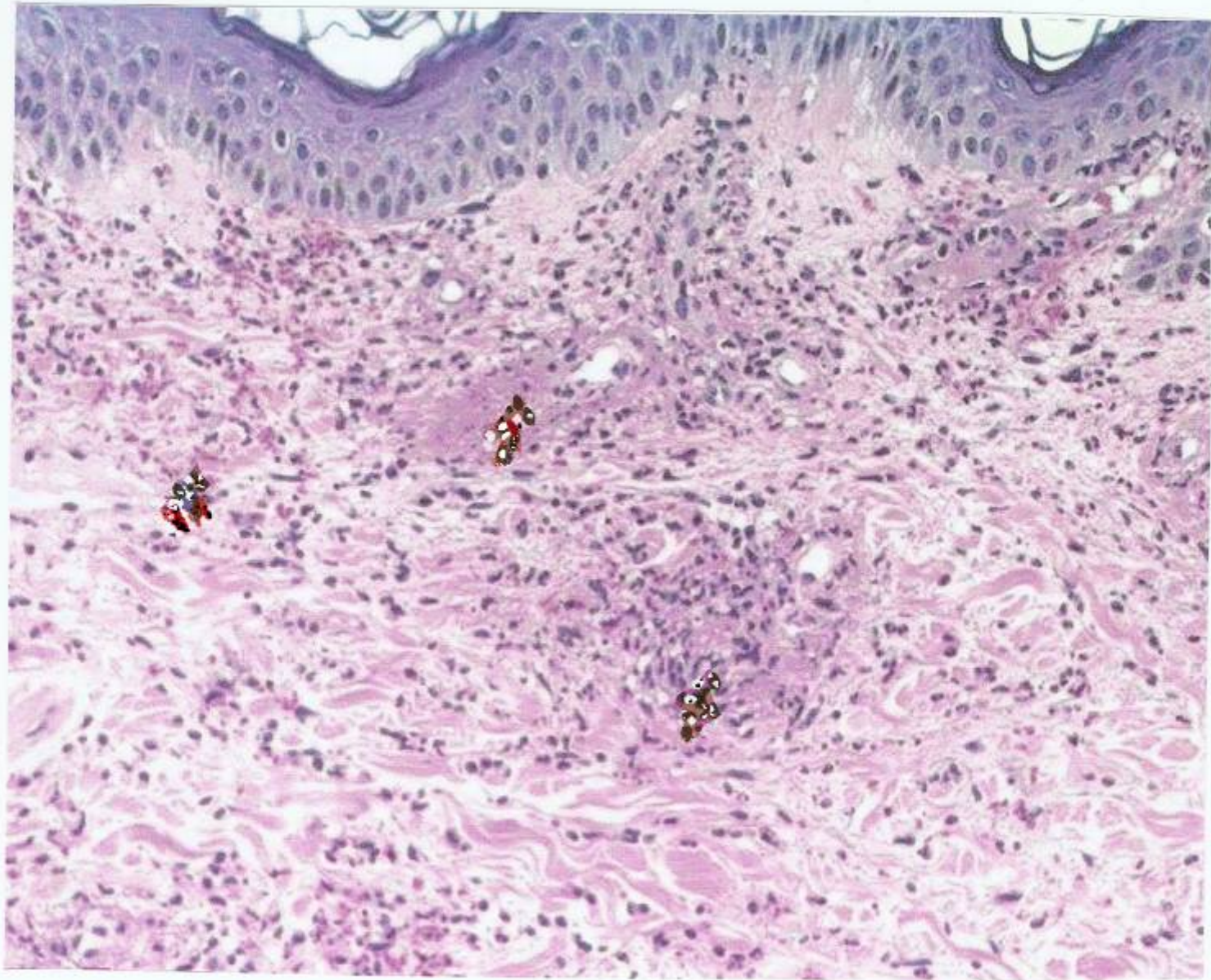
- 64 yo F
- Angina over the past 2 weeks
- Tenderness of wrists and proximal interphalangeal joints
- No pulmonary symptoms (on admission)
- A 2/6 grade systolic murmur over the all auscultation points
- No hepatosplenomegaly, lymphadenopathy

PQ Intervall 168ms
QRS Dauer 92ms
QT/QTc 408/410ms
P-QRS-T Winkel 68/188°
P Dauer 130ms
RR/PP Intervall 988/980ms

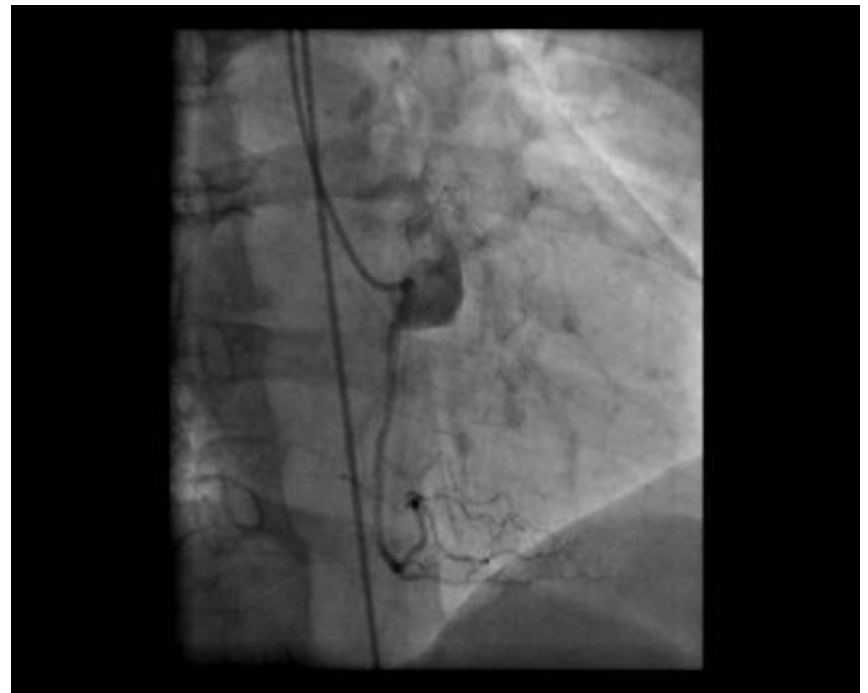
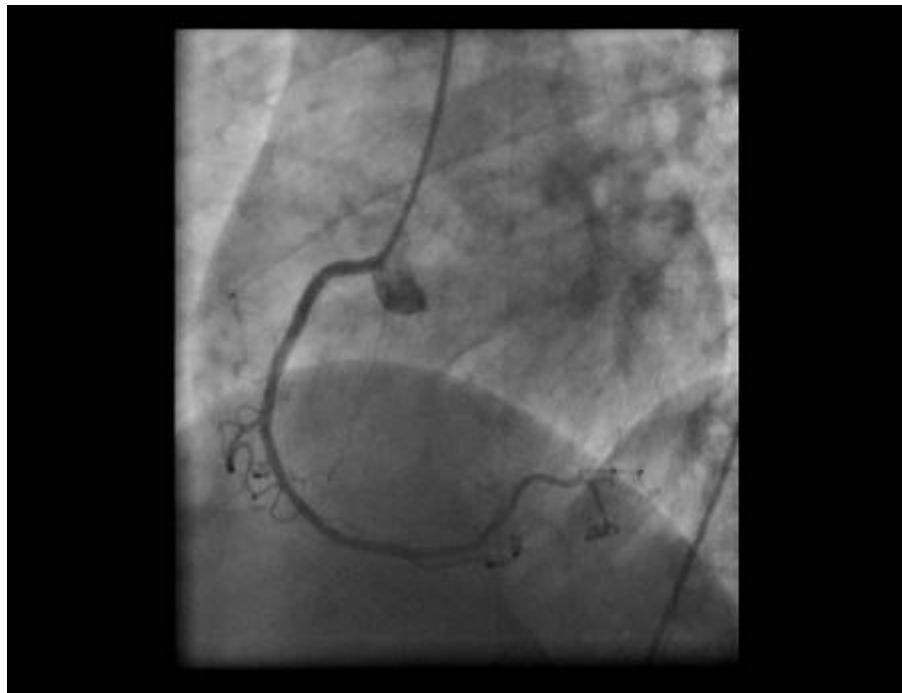
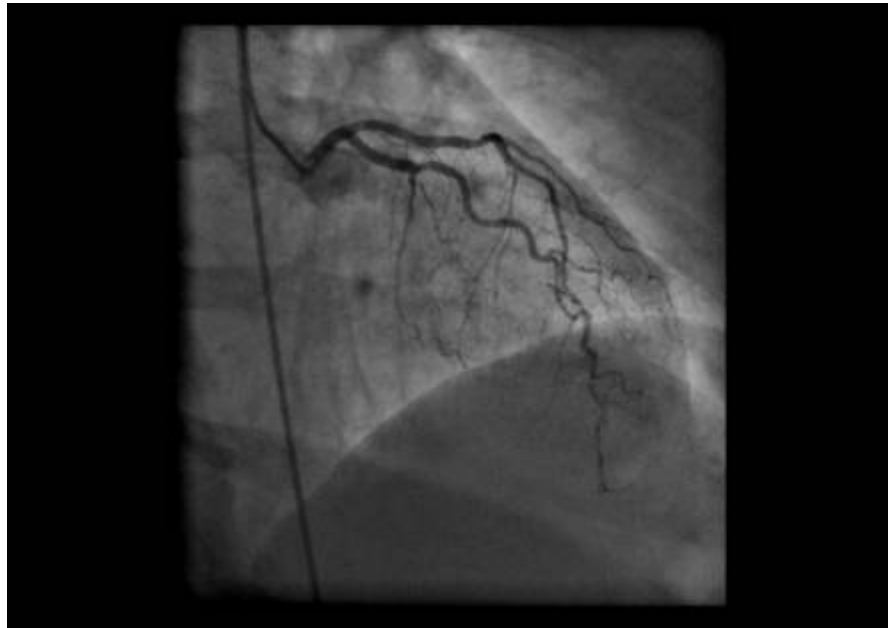


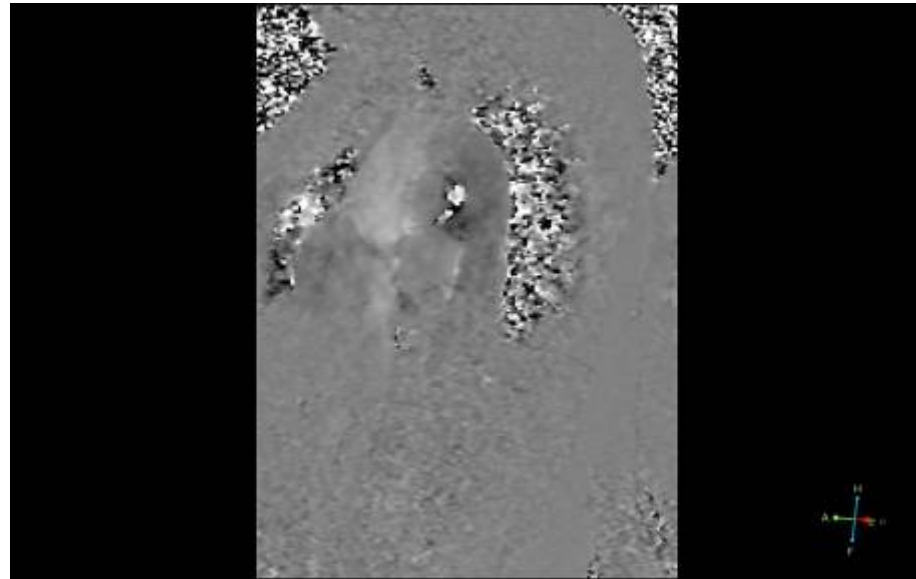
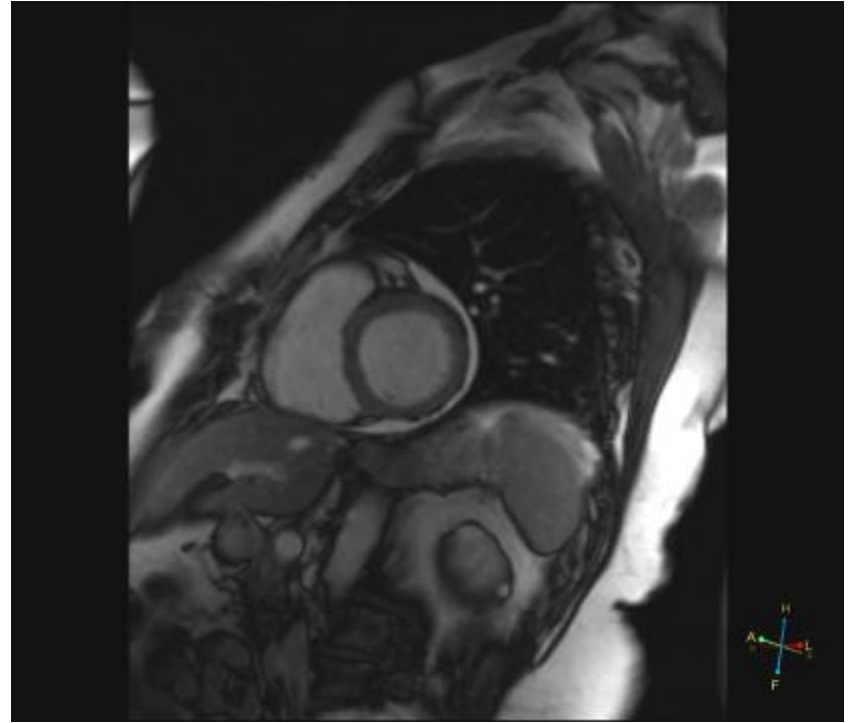
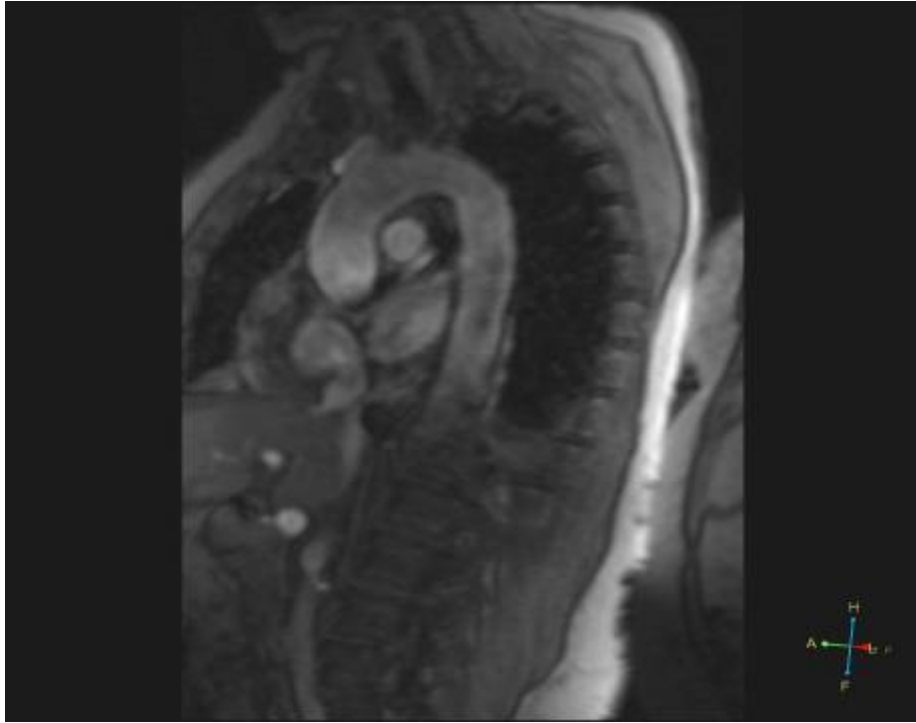
- EchoCG. Pericardial thickening
- Abdominal US and CT - N
- **ESR 90** mm/h, CRP 16.5 mg/L (N<9), Hb 10.8g/dL, WBC 7,450, Pt 434,000. Liver and kidney function tests – N
- HIV negative; blood and urine culture tests negative
- ASLO 1:400 (N<1:300), RF+, ANA -
- An enzyme-linked immunosorbent assay - positive for **cANCA** (43; N<20)

- 3rd d. Fever, SoB, paresthesia. Symmetrical peripheral neuropathy. Chest X-ray - infiltrate in the right lower lobe
- 6th d. Dyspnea worsened. Chest X-ray - **pulmonary infiltrates in both lungs**. Bronchoscopy with fluid examination - multiple erythrocytes. Biopsy - neutrophilic microabscesses, thrombosis, hemorrhage and **granuloma** with palisading histiocytes, lymphocytes, eosinophils
- 7th d. Erythematous rash; skin biopsy - **leukocytoclastic vasculitis**
- CT of paranasal sinuses - **thickening of the mucous membrane**



- + Cefuroxime, gentamycine and erythromycine
- + After the diagnosis – methylprednisolone (500mg daily, IV), cyclophosphamide (300mg daily, orally)
- + The patient was discharged on cyclophosphamide (150mg), which switched to azathioprine. Prednisone maintenance dose - 10mg





Learning points

- 👉 Cardiac manifestations are rare in WG
- 👉 Coronary involvement in WG can be specified by informative imaging techniques (MRI)
- 👉 Coronary microvascular disease is characteristic for WG