Exacerbation of eosinophilic granulomatosis with polyangiitis (EGPA) with heart involvement mimicking acute coronary syndrome

Zaostrzenie ziarniniakowatości eozynofilowej z zapaleniem naczyń przebiegającą z zajęciem serca imitującym ostry zespół wieńcowy

A case of a 49-year-old male with exacerbation of eosinophilic granulomatosis with polyangiitis (EGPA) with heart involvement mimicking acute coronary syndrome is presented. Institution of intensive immunosuppressive treatment resulted in the improvement of clinical condition and systolic left ventricular function. Coronary angiography excluded atherosclerosis as a primary cause of heart damage.

Background

Eosinophilic granulomatosis with polyangiitis (EGPA, Churg-Strauss syndrome) is a rare (annual incidence 0.5–4.2 cases/106 inhabitants), necrotizing inflammation of small and medium sized vessels with accompanying bronchial asthma, chronic rhinosinusitis, blood eosinophilia, eosinophilic infiltrations of various tissues and organs [1,8]. Although etiology and pathogenesis of the disease have not been completely elucidated, the role of abnormal immune response is suggested and favored by the presence of pANCA (anti-neutrophil cytoplasmic antibodies directed mainly against neutrophil myeloperoxidase) in the patients serum (~38% patients) [5,8,11]. Cardiac complications, depending on data source, are reported in 16–92% of cases and are usually associated with poor prognosis by contributing importantly to mortality of the patients [9]. These complications may include dilated cardiomyopathy, congestive heart failure, myocarditis, endocarditis, pericarditis, cardiac tamponade, arrhythmias, acute myocardial infarction and eventually sudden cardiac death (SCD) [3,4,7,10]. Even though the diagnosis EGPA is certain, causes of heart damage may remain unclear.

Case report

A 49-year-old male chronic smoker, diagnosed with EGPA 4 years ago, suffering also from steroid-induced diabetes, hypercholesterolemia and arterial hypertension was admitted to the Department of Internal Medicine of the University Hospital due to the exacerbation of the disease. He complained of fever of 39°C for 2 weeks, abdominal pain, diarrhea, musculoskeletal pain associated with decreasing body weight (8 kg within 2 weeks) and generalized weakness. Routine ECG revealed ST segment depression, symptoms of abnormal ventricular repolarisation in leads II, III, aVF, V3-V6 and single extrasystolic ventricular beats. On physical examination he was febrile with temperature of 38,5°C, HR 80/min, BP 130/80 mmHg, with bilateral crackles over the lung bases and tenderness in epigastric area. Laboratory tests revealed elevated markers of inflammation with marked leukocytosis 20.600/μl (normal range 4.5-10.000/μl), CRP 76.5 mg/l (normal range to 5 mg/l), absolute eosinophil count 8424 cells/μl (normal range up to 400cells/μl), elevated troponin I 3.84 ug/l (normal range up to 0.11 ug/l) and CK-MB max 32 U/l (normal range to 12 U/l). The patient did not report angina symptoms. Transthoracic echocardiography was performed, showing akiinesia of the basal segments of inferior and posterior wall and left ventricular ejection fraction value of 48%. The suspicion of myocardial infarction (NSTEMI) was made as a result of the clinical picture and the additional tests. At first due to the life-threatening exacerbation of EGPA pharmacological treatment with high doses of glucocorticosteroids (methylprednisolone i.v. 500 mg for 3 days, then 1 mg/kg body weight/day) and cyclophosphamide 75 mg/day were administered. With numerous coronary heart disease risk factors present, acute MI due to atherosclerosis was considered in differential diagnosis. Due to unstable state of the patient and elevated troponins in consecutive tests coronary angiography was performed but no critical narrowings in the coronary arteries were found. The diagnosis of myocarditis in the course of

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the exacerbation of EGPA was suspected. Clinical improvement of the patient and rapid regression of the left ventricular dysfunction after 5 days of immunosuppressive therapy confirmed the preliminary diagnosis.

Discussion

There are two main mechanisms of myocardial damage in the course of EGPA: eosinophilic infiltration within myocardium and ischemia caused by inflammation in the small and medium-sized vessels, including coronaries [8,9]. Clinical presentation of our patient suggested both mechanisms: myocarditis together with acute myocardial infarction without ST segment elevation (NSTEMI). In the acute phase of myocarditis damage of cardiomyocytes and release of myocardial necrosis enzymes occurs, therefore ECG in myocarditis is almost always abnormal and may show characteristics of acute myocardial damage, supraventricular and ventricular arrhythmias, changes in atrio-ventricular and intraventricular conduction. Considering the burden of multiple atherosclerosis risk factors (age, male gender, hypercholesterolemia, diabetes mellitus, smoking, family history of premature coronary artery disease), all putting the patient in a high cardiovascular risk category, the diagnosis of myocardial infarction should have been considered. ECG and echocardiography results were indicative of ischemia in the right coronary artery supply region. In order to verify the diagnosis coronary angiography was performed, which excluded any atherosclerotic changes. Myocardial biopsy was never performed to confirm myocarditis, but the patient has shown significant clinical improvement with anti-inflammatory and immunosuppressive medications.

ANCA antibodies are present in 38-50% of EGPA [5]. Their absence, as in the presented case, associates with higher incidence of inflammatory lung infiltrates and heart involvement [2]. Cardiac involvement is reported to be the major predictor of a fatal outcome in the course of EGPA [2,8]. For this reason more attention should be placed to implement both intensive immunosuppressive therapy to diminish the risk of disease progression and even earlier active search (echocardiography, heart magnetic resonance) for heart involvement in the early and/or asymptomatic phase of the disease [6,9].

Conclusions: Acute myocarditis and myocardial infarction are among life-threatening complications of EGPA. On the other hand, even if there are strong indications that heart damage is a result of EGPA exacerbation, other possible etiologies should always be considered. Coronary angiography, therefore, should be performed at even the slight suspicion of an atherosclerotic background and suspected infarction.

Bibliography


