

Granulomatosis with polyangiitis complicated by myocardial infarction: case report

Ziarniniakowatość z zapaleniem naczyń powikłana zawałem serca – opis przypadku

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Abstract

Granulomatosis with polyangiitis (GWP) is a rare systemic disease of small and medium vessels of the unknown aetiology. Virtually, every organ can be affected by the disease, but GWP mainly attacks the respiratory tract and the kidneys. Cardiac involvement occurs unfrequently, but may be insidious and dangerous. In this article we present a case report of a 55-year-old female patient with myocardial infarction during the course of GWP.

Key words: granulomatosis with polyangiitis, myocardial infarction

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Introduction

Granulomatosis with polyangiitis (GWP, former Wegener's granulomatosis) is a rare systemic autoimmunological disease of small and medium vessels, respiratory tract and kidneys. The hallmarks of the disease are: formation of necrotic granulomas and presence of cytoplasmic anti-neutrophilic cytoplasmic antibodies (c-ANCA). Cardiac involvement in the course of GWP occurs uncommonly and is very heterogenic. Frequent cardiac manifestation includes pericarditis, coronary arteritis, myocarditis, rarely valve abnormalities and endocarditis, while arrhythmias and myocardial infarction happens occasionally [1]. In this article, we would like to present our experience with GWP patient complicated by myocardial infarction.

Case report

A 55-years-old female was admitted to Department of Pulmonology and Allergology. She complained of fever (39

degrees Celsius), dyspnoea, dry cough and retrosternal pain lasting from two weeks. Before the admission she was treated with amoxicillin with clavulanic acid and moxifloxacin. Her previous medical history since last nine months includes recurrent sinusitis, nose and sinus polyps (the patient underwent several laryngological interventions), left myringotomy bilateral hearing impairment, tracheitis and laryngitis. A number of specimens for histopathological analysis were taken, but results did not show any abnormalities. The patient during the stay at guesthouse last year was repeatedly bitten by bugs and exfoliating erythematous lesions appeared on her skin. Antihistamine treatment was applied – without significant effect.

During the admission, the erythematous papules on upper and lower limbs, as well as on face and neck were noticed. Physical examination revealed no clinical changes. Chest X-ray demonstrated II and III right lung segment and upper zone of left lung opacities. Slight consolidations were found in parahilar zones (Figure 1). The electrocardiogram (ECG) showed ST depression up to 3 mm in V3–V5,



Figure 1. Chest X-ray showing II and III right lung segment and upper zone of left lung opacities; slight consolidations visible in parahilar zones

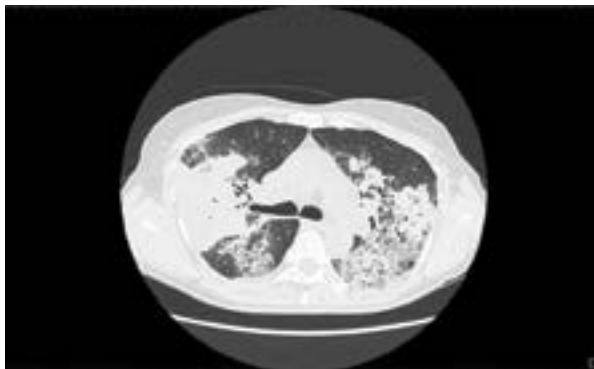


Figure 2. Computed tomography chest angiography showing interstitial consolidations in the upper zones of both lungs

II, III and aVF leads and ST-elevation in aVR lead. Abnormal laboratory tests results performed after ECG were as follows: C-reactive protein (CRP) 213 mg/L, D-dimers 3710 ng/ml, high-sensitivity troponin T 492.4 ng/L (after three hours 812.3 ng/L). Computed tomography chest angiography excluded pulmonary embolism. However, this study confirmed a presence of interstitial consolidations with predilection to upper lobes (Figure 2). The suspicion of non ST-elevation myocardial infarction (NSTEMI) has been made and the patient has been referred to a haemodynamic laboratory at Clinical and Didactic Centre in Lodz for further therapy.

After cardiovascular intervention, the patient returned to the Department of Pulmonology and Allergology. The suspicion of GWP (ANCA antibodies were negative) was made and the immunosuppressive agents were used – prednisone and cyclophosphamide. Due to radiological progression and elevated markers of inflammation, cyclophosphamide was discontinued. Unfortunately, the dyspnoea still worsened and bronchoscopy

with tracheal biopsy were performed. Histopathological findings included the presence of granulomas and cyclophosphamide were again implemented. Clinical improvement of respiratory symptoms and skin lesions was achieved. Furthermore, the progressive anaemia was noticed (haemoglobin [Hb] 6.8 g/dL) and four units of packed red blood cells were transfused. Diffusing capacity for carbon monoxide corrected to actual haemoglobin concentration (DLCO SB K) was 14.5 (168%), which suggested the alveolar bleeding. The presence of kidney failure (estimated glomerular filtration rate [eGFR] 30 mL/min/1.73 m² and haematuria) caused the patient admission to the department of nephrology. After that, kidney biopsy was performed and microscopic pattern matched glomerulonephritis with crescents and necrosis of capillary loops.

Discussion

Oliveira and colleagues demonstrated that 86% of patients with GWP have echocardiographic abnormalities, but only 36% have direct connection with the disease [2]. In similar study, ECG and echocardiography pathologies were observed in 46% of patients. After using the magnetic resonance technique for heart imaging, the percentage of patients increased to 61%. The Authors of this study noticed the increased risk of death in a group with asymptomatic heart involvement [3]. In echocardiography, detected defects concerned mainly abnormalities of aortic valve (stenosis, insufficiency), the presence of fluid in pericardial cavity, regional wall motion abnormalities and systolic dysfunction of left ventricle [2, 4]. The GWP patients have higher risk of ischaemic heart disease (1.9 [95% confidence interval {CI} 1.4–2.4]) and myocardial infarction (2.5 [95% CI 1.6–3.7]) than healthy subjects. Particularly, the risk is even higher for males, patients above the age of 50 years and in a group who received high cumulative dose of cyclophosphamide [5]. Moreover, cardiac involvement predisposes to disease recurrence after the remission (2.9 [95% CI 1.3–6.5] [6]. Additionally, in patients resistant to induction treatment (> 6 months), cardiac involvement occurs more often and is characterised by late gadolinium enhancement in heart MRI and signs of myocarditis [7]. Myocardial infarction during the course of GWP happens infrequently and concerns about 10% of patients with heart involvement [1]. So far, six cases of asymptomatic myocardial infarction in GWP patients has been reported. Interestingly, myocardial infarction was the first sign of cardiac involvement and had not been preceded by the chest pain. In all these patients myocardial infarction was lethal and may appear after clinical improvement in the time of initial therapy of GWP [8].

Conclusions

In summary, we have reported a rare complication during the course of GWP, which is myocardial infarction. The physician should be aware of cardiac involvement in GWP and we recommend basic cardiovascular examination (ECG

and echocardiography) as a routine procedure in every case of GWP patient.

Conflict of interest(s)

None declared.

Streszczenie

Ziarniniakowatość z zapaleniem naczyń (GWP) to rzadka ogólnoustrojowa choroba małych i średnich naczyń krwionośnych o nieznannej przyczynie. Teoretycznie każdy organ może być zajęty przez chorobę, jednak GWP dotyczy głównie układu oddechowego i nerek. Zajęcie serca występuje sporadycznie, ale może przebiegać podstępnie i być niebezpieczne. W tym artykule autorzy chcieli zaprezentować opis przypadku 55-letniej kobiety z zawałem serca w przebiegu GWP.

Słowa kluczowe: ziarniniakowatość z zapaleniem naczyń, zawał serca

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