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Acute myocarditis in patient with IgG4-related disease

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Abstract. We present case of 27-year-old woman in the chronic care of her gastroenterologist for IgG4-related disease, with pancreatitis, cholangitis, colitis, and perimyocarditis in the past. Now in long-term remission on combination therapy with budesonide and biological therapy with rituximab, she was admitted to our Intensive care unit (ICU) for chest pain and dyspnoea with increased frequency of stools. She did not present with any remarkable findings during physical examination and she was hemodynamically stable, without requiring oxygen. Laboratory findings showed leukocytosis with neutrophilia, elevated CRP (C-reactive protein) without PCT (procalcitonin), slightly elevated Troponin (hs-cTnT) without typical dynamic for acute coronary syndrome, and high NTproBNP (N-terminal prohormone of brain natriuretic peptide). We performed transthoracic echocardiography showing preserved left ventricular systolic function, localized hypokinesia, and reduction of the longitudinal strain of the basal segment of the interventricular septum. With her history of allergy to gadolinium, CT coronarography was done that showed small pericardial effusion, without pericardial thickening, and calcifications and stenosis of coronary arteries. Abdominal ultrasound showed non-homogenous structure of the pancreas, cholecystolithiasis, solid-cystic lesion of the right adrenal gland, and thickened wall of the colon. After these results we decided to add PET/CT scan, which showed diffuse activity of bone marrow, right adrenal gland expansion with low activity, and colitis without any findings of malignancy or inflammation. We concluded the condition as acute myocarditis associated with the IgG4-related disease. She was then treated with oral prednisone with good effect and was discharged after 13 days. Fig. 2, Ref. 4, on-line full text (Free, PDF), www.cardiologyletters.sk

Keywords: IgG4-related disease – acute cardiac injury – myocarditis – cardiac markers

IgG4-related disease is an autoimmune disease affecting most organs (1). The disease starts with an early inflammation later followed by fibrosis. It typically affects the pancreas, kidneys, retroperitoneal tissue, large vessels, bile ducts, and salivary and lacrimal glands, causing inflammation and progressive fibrosis. Among many, the disease's involvement of medium-sized vessels and pericardium is less common. These may include valvular

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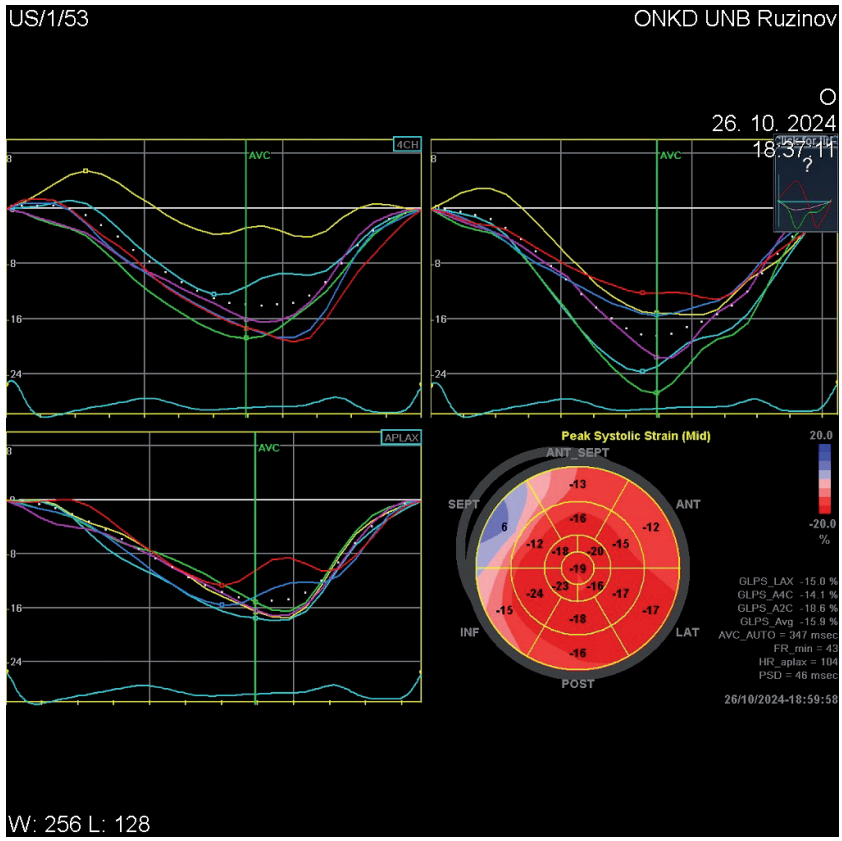

Figure 1 Reduced global longitudinal strain of basal segment of the IVS

heart disease, pericardial disease, heart masses, and stenosis of coronary arteries not meeting typical organ involvement criteria of the disease. Our case presents possible cardiovascular involvement of the disease in a young woman.

Case presentation

A 27-year-old woman with history of IgG4-related disease visited the emergency department (ED) of our hospital for pressure chest pain. Her IgG4-related disease has been of predominantly colonic involvement, with only discrete signs of type 1 autoimmune pancreatitis, and history of perimyocarditis in the past. She has previously received steroid therapy and several doses of rituximab. Before admission, the patient had been in long-term remission of the IgG4 disease. The current pain started suddenly at night and worsened in the prone position and during deep breathing. The pain was alleviated while sitting. She has tried ibuprofen without sufficient effect. The patient also reported having shivers and similar symptoms during the previous episode of pericarditis. She didn't report any abdominal pain or discomfort.

Her objective findings were without any significant abnormalities, her blood pressure was normal with borderline tachycardia, and she had normal oxygen saturation. ECG showed normal sinus rhythm with an incomplete right bundle branch block. Initially, the patient had leukocytosis with absolute and relative neutrophilia, there was slight increase in serum creatinine (115.2 umol/l), hs-cTnT was elevated without typical dynamics consistent with the acute coronary syndrome (19.94 to 14.58 ng/l), NTproBNP and CRP were also significantly increased (1787 ng/l and 98 mg/l respectively). D-dimer was only slightly elevated (0.55 mg/l) and hyperfibrinogenemia (4.48 g/l) was also present. Urine tested positive for leukocyturia without clinical correlation. chest x-ray was performed with normal findings.

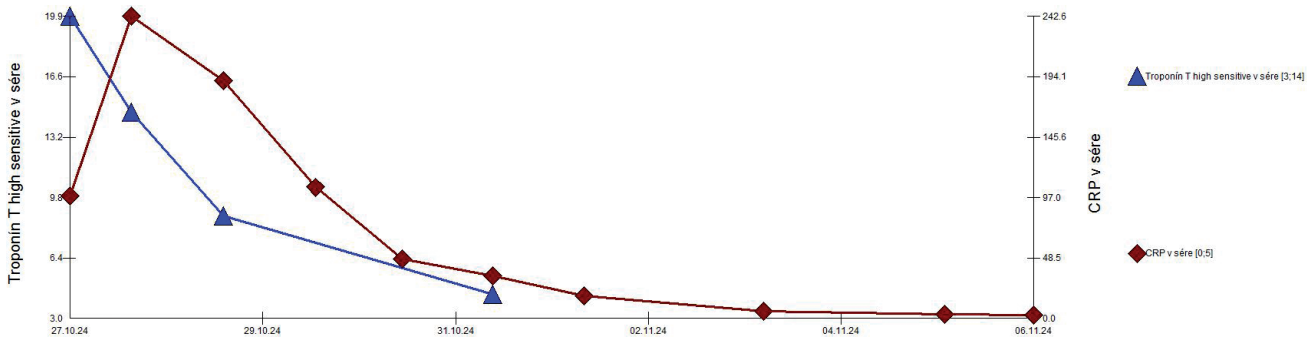

Figure 2 Trends in CRP and hs-cTnT levels for this patient

The patient was admitted to our ICU for close monitoring given the cardiac involvement: transthoracic echocardiography was performed showing normal ejection fraction of the left ventricle, and normal diastolic function without significant valvulopathy. However, we observed reduced longitudinal strain of the basal segment of the interventricular septum (IVS) with hypokinesis (**Figure 1**). Based on the clinical, laboratory, and imaging findings, the diagnosis of focal myocarditis of IVS was established. Further workup was performed with additional laboratory markers showing increased serum immunoglobulin (IgA, beta-2-microglobulin (B2MG), increased thyroid-stimulating hormone (TSH) without abnormality of free T4 (fT4) and thyroid antibodies, and borderline increase in alpha-1-globulin. Swabs for microbiological examination revealed non-pathological flora. Rectal swabs disclosed the massive presence of Saccharomyces cerevisiae and Candida albicans complex.

With her history of gadolinium allergy, cardiac CT with coronarography was performed showing normal findings on coronary arteries without any significant wall-motion abnormalities, normal ejection fraction of the left ventricle, with only small localised pericardial effusion ventrobasal to right atrium. Abdominal ultrasound showed non-homogenous structure of the pancreas, multiple cholecystolithiasis of small diameter, without cholestasis, thickening of the wall of the ascending, descending, and sigmoid colon, and lesion in the right adrenal gland. PET/CT with FDG verified the expansion of the right adrenal gland but without any signs of malignancy. There was increased activity in the descending and sigmoid colon, Waldeyer's ring, and the bone marrow.

Management and Outcome

Based on the presumed diagnosis of IgG4-related disease complication as the culprit of myocarditis, we consulted the gastroenterologist and started treatment with 30 mg of oral prednisone. The patient remained in stable clinical condition with gradual alleviation of the chest pain. Laboratory follow-up showed improvement of cardiac and inflammatory markers (NTproBNP 681 ng/l, hs-cTnT 4.34 ng/l, and CRP 2.6 mg/l) (**Figure 2**). She was discharged after 13 days.

Discussion

The diagnosis of IgG4-associated perimyocarditis was established based on the patient's symptoms, positive cardiac markers (NTproBNP, hs-cTnT), and CRP with the echocardiographic finding of decreased longitudinal strain of IVS with negative findings on CT coronarography. Most of the secondary findings can also be attributed to the base disease characteristics. Unfortunately, the patient's allergy to gadolinium restricted us from performing cardiac MR. Prednisone orally was chosen as treatment with good effect and the patient was discharged without any symptoms.

Cardiac manifestation of IgG4-related disease is not unheard of the case series by Seshika Ratwatte et al. in 2022 showcases multiple cases including myocardial infarction with soft-tissue masses surrounding coronary arteries, pericarditis progressing to constrictive, and pericarditis, after ibuprofen treatment (2). The limiting factor is always a challenge in diagnosing Ig4-related

disease, which involves clinical, laboratory, radiology, and pathology findings (3). The most typical cardiovascular target organ is the aorta, and involvement of coronary arteries, myo- and or peri-cardium does not meet the criteria of disease affecting the typical organ. Cardiac involvement, therefore, is probably under-represented.

Novel and more commonly used cardiac imaging methods in conjuction with other imaging methods, typically used in IgG4-related disease, can help with diagnosis. This is meant in patients presenting with symptoms of cardiovascular involvement, and also laboratory markers of cardiac injury (most commonly used hs-cTnT, NTproBNP) and ECG. In rare cases cardiac biopsy can also be performed (4).

Patients with history of IgG4-related systemic disease presenting with any symptoms commonly associated with myocardial damage should be thoroughly investigated. Although the involvement of the myocardium is uncommon, delayed therapy can potentially lead to devastating consequences.

References

1. Wallace ZS, Naden RP, Chari S, Choi H, Della-Torre E, Dicaire JF, et al. American College of Rheumatology/European League Against Rheumatism IgG4-Related Disease Classification Criteria Working Group. The 2019 American College of Rheumatology/European League Against Rheumatism Classification Criteria for IgG4-Related Disease. Arthritis Rheumatol. 2020;72:7-19. doi: 10.1002/art.41120. Epub 2019 Dec 2. PMID: 31793250.
2. Ratwatte S, Day M, Ridley LJ, Fung C, Naoum C, Yiannikas J. Cardiac manifestations of IgG4-related disease: case series. Eur Heart Case Rep. 2022;6:ytac153. doi: 10.1093/ehjcr/ytac153. PMID: 35481260; PMCID: PMC9036079.
3. Khosroshahi A, Wallace ZS, Crowe JL, Akamizu T, Azumi A, Carruthers MN, et al. Second International Symposium on IgG4-Related Disease. International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. Arthritis Rheumatol. 2015;67:1688-1699. doi: 10.1002/art.39132. PMID: 25809420.
4. Matsumura I, Mitsui T, Tahara K, Shimizu H, Yanagisawa K, Ishizaki T, et al. IgG4-related Disease with Cardiac Mass. Intern Med. 2020;59:1203-1209. doi: 10.2169/internalmedicine.4054-19. Epub 2020 Jan 17. PMID: 31956206; PMCID: PMC7270767.