Unusual Heart Involvement of Wegener’s Granulomatosis and Literature Review

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To the editor,
A female patient, 60 years of age, was presented to our hospital with chest pain and monomorphic ventricular tachycardia (VT, Figure1.A). On physical examination she had normal blood pressure. She was transferred to the Coronary Care Unit and amiodarone perfusion restored basal rhythm in atrial fibrillation. She had history of AF, with normal transthoracic echocardiogram (TTE) six months before.

Serial cardiac enzymes were in normal limits and continuous telemetry monitoring did not record other VT episodes. Chest radiography was unremarkable. A TTE was performed and an one mitral mass was found at atrioventricular junction with displacement of the posterior mitral leaflet (Red asterisk in figure1.B-C) and also had significant mitral regurgitation (MR). The mass had no contrast uptake and the patient had good biventricular function. However, global speckle tracking echocardiography (STE) was decreased with longitudinal strain value of 10% and circumferential strain of 16%. A transesophageal echocardiogram (TEE) demonstrated a mass at atrioventricular junction sized 20x18 mm, with similarly echodensity to the myocardium and well-defined edges (Figure1.D, red asterisk) and severe MR secondary to displacement of the posterior mitral leaflet (Figure1.E).

Coronary angiography showed normal epicardial coronary arteries. VT or other arrhythmias were not induced on electrophysiological study. In the blood test was elevated creatinine, erythrocyte sedimentation rate and myeloperoxidase-antineutrophil cytoplasmic antibodies (MPO-ANCA) antibody were 1/80. Other abnormal parameters were not presented. Urinary analysis and microscopy were negative for protein, red blood cells or red cell casts.

Cardiac Multidetector Computed Tomography and Cardiac Magnetic Resonance (CMR) confirmed the mass at atrioventricular junction level (Figure 1.F-G, red asterisk) and these test ruled out other thoracic abnormalities. The remaining anterolateral papillary muscle was thickening and hypertrophied with hyper-enhancement consistent with fibrosis. Moreover, T2-weighted imaging demonstrated hyperintense mass with respect to the surrounding myocardium in relation of inflammatory mass. (Figure1.H, red arrow).

The patient had involvement of other organs as well. She had saddle nose by destruction of the septum, bilateral hearing loss, sinusitis and scleritis and renal involvement. A biopsy of the septal mucosa was performed showing fibrinoid necrotizing vasculitis with granulomatous inflammation.

This patient was diagnosed of Wegener’s Granulomatosis (WG) and she was treated with methylprednisolone during 3 days, continued with prednisolone and cyclophosphamide. An 8 days later echocardiogram did not find the mass (Figure2.A-C,
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Review of the literature

Wegener's granulomatosis is one of the pauci-immune small vessel vasculitis and was characterized as a clinical syndrome in 1936. The etiology remains unknown [1]. The prevalence in the general population has been reported to be 20-50 per million1. This illness affects respiratory and renal tracts and is characterized by granulomatous lesions [1]. The percentage of cases that involve the heart is unknown, with range 6-44% [2], but cardiac manifestations are rare and heart involvement is associated with poor outcome [2]. The frequency of cardiac involvement varies depending on the level of disease activity and the diagnostic test employed. The ECG and TTE are the most often used screening tools for heart evaluation [2]. Recently, CMR has emerged as a novel non-invasive imaging modality providing comprehensive and accurate evaluation of myoccardial function and structure. Therefore, T1 and T2 intensity on CMR can be helpful in doing the differential diagnosis in cardiac tumors and for monitoring therapeutic efficacy [3].

A wide spectrum of cardiac manifestations have been described in the course of WG such as myocarditis, coronary vasculitis, valvular heart disease, pericarditis and/or rhythm disorders [4]. In the setting of WG cardiac masses are not common and can be presented as a tumor-like mass. These findings are reported in few cases on papillary muscles, left or right ventricular cavity or valves and only two of them have been associated with VT [5-6]. Treatment with cyclophosphamide and corticosteroids is described that improve the cardiac symptomatology [7]. So far, high dose of corticosteroids and cyclophosphamide may treat cardiac affection in this pathology. In our patient CMR imaging revealed inflammatory involvement of the heart and after

Figure 1

Pre-immunosuppressive therapy: Image A) ECG on admission showed monomorphic ventricular tachycardia with left bundle-branch block morphology and left-axis deviation. TTE images: Image B) Parasternal short axis at mitral level evidenced an atrioventricular junction mass with secondary displacement of posterior mitral valve leaflet. Image C) Zoom of apical 4-chamber showing the mass at atrioventricular junction. TEE images: Image D) 2-chamber view confirmed an atrioventricular junction mass with sized to approximately 20x18 mm. E) Doppler color imaging at longitudinal TOE view displayed a severe mitral regurgitation. E) Coronal imaging of CMR showing atrioventricular junction mass. F) Axial 4-chamber imaging confirmed the mass. G) T2-weighted imaging CMR showed well defined hyperintense mass, with respect to the surrounding myocardium (indicated by red arrow). The mass is indicated by red arrow and red asterisk. LV: Left ventricle. LA: Left atrium.

Midline sternotomy was performed and mitral mass was not found. The surgeon performed mitral valve replacement using mechanical prosthesis. Histological of biopsies showed central ischemic necrosis of the myocardium and perivascular calcifications of papillary muscles with myocard degeneration and lymphocytic infiltrate of mitral valve (Figure 2.D-F). There was no evidence of giant cells, granulomas or vasculitis.

Since the admission, any other episodes of VT have not been presented. Post-operative TTE showed normal left ventricular size with good overall systolic function, normal mitral prosthetic valve function and normal limits of pulmonary artery pressure. The patient was discharged in relatively good condition on mild doses of corticoids, acenocoumarol, beta-blocker and angiotensin-converting enzyme inhibitor. She was no discharge with implantable cardioverter-defibrillator. Three years after surgery she is in stable condition with no cardiac symptomatology.

Figure 2

Post-immunosuppressive therapy: TTE images: Image A) Parasternal long axis without the mass. Image B) Apical 4-Chamber confirmed that the mass was disappeared. Image C) Zoom of apical 3-Chamber could not find the mass as well. Histologic assessment. Image D) Histological section of mitral valve with hematoxylin eosin tinction x40 of mitral valve and papillary muscles is showing myoccard degeneration and chronic inflammation. Image E) Histological section x100 of papillary muscle showing lymphocytic infiltrate. Image F) Histological section x200 of papillary muscle x 200 confirmed lymphocytic infiltrate. The previous position of the atrioventricular mass is pointed with red arrow. LV: Left ventricle. LA: Left atrium. RV: Right ventricle. RA: Right atrium.
treatment the inflammation decrease with improving of global values of STE and also the mass disappears. ICD implantation was not considered regarding in the moment of VT the patient was in acute phase of inflammation. In this point of view, Jon Torgny et al. described a patient with WG and complete heart block and after treatment with high dose of steroid and cyclophosphamide he recovered normal rhythm and definitive pacemaker had not been implanted [8]. In resistant cases, rituximab or other monoclonal antibodies can be helpful [9]. Nonetheless, the effect of immunosuppressive therapy in patients with WG has not been systematically evaluated. Prospective studies are necessary to assess the real impact of this treatment in this pathology.

On the bases of these findings, we recommended cardiac screening in patients with WG with serial echocardiography and electrocardiograms to identify and monitor heart involvement. STE and Cardiac CMR should be considered to assess the subclinical systolic impairment, the real extension of the cardiac affection and in the correct assessment of the tumor-like mass. Also, aggressive treatment with immunosuppressive therapy may be started when WG is diagnostic.

Declarations of Interest
The authors declare no conflicts of interest.

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The authors state that they abide by the “Requirements for Ethical Publishing in Biomedical Journals” [10]

Abbreviations
CMR: Cardiac Magnetic Resonance.
WG: Wegener’s Granulomatosis.
STE: Speckle tracking echocardiography
TTE: Transthoracic echocardiogram
TOE: Transesophageal echocardiogram
VT: Ventricular tachycardia.

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