A rare cardiac manifestation of Wegener's granulomatosis

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Answer: C

The correct answer is C. The perioperative view and after excisional view of cardiac mass was shown in Figure 4-5. This is a case of myocarditis related to WG. As the ECGs and high level of cardiac markers with echocardiographic findings clearly showed us typical myocarditis case related to Wegener. ECGs indicate not only conduction abnormalities but also cardiac injury caused by myocarditis. And in the end, systolic dysfunction facilitate the development of cardiac thrombus. Spontan echo contrast described in text is also the precursor formation of cardiac thrombus. Transthoracic echocardiography displayed severe left ventricular systolic dysfunction with ejection fraction 25-30% and very large cardiac thrombus (4x4 cm) in the left ventricule apex (Video 1-2). One of treatment choice in such a cardiac thrombus is thrombectomy by surgery but there is a myocarditis process and patient is still on therapy for active inflammation. Timing of surgery is very important because of mortality on going myocarditis process. And another choice described in most of the cardiac thrombus cases is thrombolytic therapy (1). But this is a left ventricule cardiac thrombus and there is a risk of embolization. We opted for a treatment based on intravenous heparin dose adjustment by aPTT, intravenous furosemide, spiranolactone, carvedilol, ramipril and prednisolone. Two weeks later, thrombus size has mildy decreased (3 x 3 cm) and thrombus was more mobile (Video 3-4). Because of the risk of embolism, he underwent LV thrombectomy. Postoperative pathologic examination of a specimen revealed an organizing thrombus (Fig. 4-5). Postoperative course was uncomplicated, and warfarin therapy was started. Postoperative transthoracic echocardiography revealed mild left ventricular systolic dysfunction with EF 40-45%. There was no thrombus in LV. The patient was referred to rheomatologist for further treatment.

Other choices are incorrect because a mass that rapidly occurs in the heart can be only thrombus. Granuloma and sarcoma are slow growing masses relatively to cardiac thrombus. Wegener's granulomatosis (WG) is a rare form of vasculitis of the small- and medium sized blood vessels affecting mainly the upper and lower respiratory tracts as well as the kidneys (2). The clinical signs and symptoms can be various forms including upper airway disease such as epistaxis, chronic sinusitis. In the lower respiratory tract, nodules, infiltrates, cavitary lesions and pulmonary hemorrhage presenting as hemoptysis are the signs of this vasculitis. WG can also involve the renal microvasculature in up to 75% of cases in the form of rapidly progressive glomerulonephritis and renal failure. Cardiac involvement in WG is not common manifestation, approximately 6-44% of cases having some degree of heart disease (3, 4). Pericarditis and coronary vasculitis are the most common findings (50% of cases), but myocarditis, endocarditis and conduction system defects are also described (5). Cardiac muscle involvement due to WG was noted to occur in 2%< one series of 158 patients (6). The overall mortality rate of WG with cardiac involvement has been

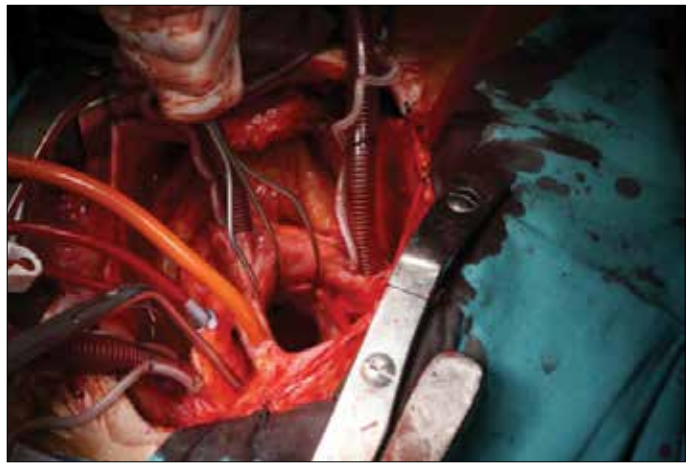

Figure 4. Perioperative view of cardiac mass

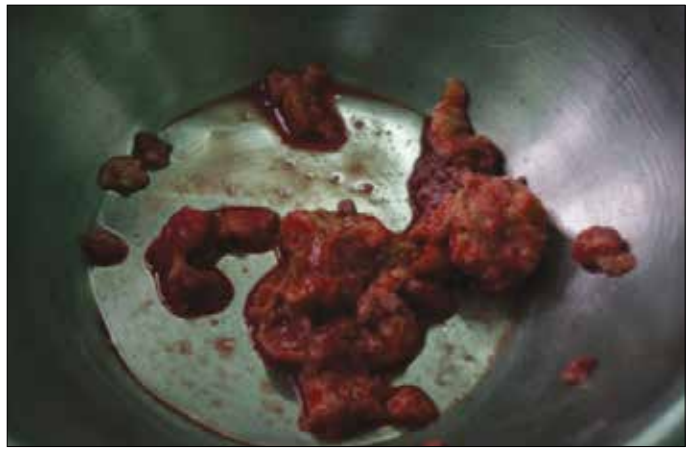

Figure 5. After excisional view of cardiac mass

reported to be between 15-45% (7). We present a case of WG in which the diagnosis of cardiac involvement was made with echocardiographic imaging modalities. Severe left ventricular systolic dysfunction with giant cardiac thrombus has not been reported previously in any WG patients. We believe that extreme intensity of inflammatory process of WG in our patient resulted in this rare manifestation. Routine cardiovascular surveillance including echocardiography and electrocardiograms are important for the screening and monitoring of the patients with WG.

Ali Elitok, Samim Emet, İmran Önür, Ekrem Bilal Karaayvaz, Ömer Ali Sayın*, Berrin Umman, Zehra Buğra, Fehmi Mercanoğlu

Departments of Cardiology and *Cardiovascular Surgery, İstanbul Faculty of Medicine, İstanbul University, İstanbul-*Turkey*

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