Multiple fatal images in right heart

Sağ kalpte multiple ölümcül görüntüler

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Right answer: 4. Behçet's disease

This patient had been admitted to hospital several times with eleven years history of Behçet's disease. Behçet's disease (BD) is a multi-systemic disease and may present with vascular, neurological, cutaneous, pulmonary, gastrointestinal, rheumatological, genito-urinary and cardiovascular manifestations (1). Cardiovascular involvement in BD is seen as 7-46% and different prevalence's have been observed in many trials (2). The main areas of cardiac involvement of BD are dilatation of proximal aorta, interatrial septal aneurysm, patent foramen ovale, mitral valve prolapse, and mitral regurgitation also there are less commonly affected areas such as pancarditis, myocardial infarction, conduction system disturbances, and intracardiac thrombosis. In this case, intracardiac thrombus formation is a rare, fatal and serious complication of BD (2). The pathologic mechanism underlying the thrombotic tendency among patients with BD is not well known. There are different hypothesis about thrombosis such as endothelial cell ischemia or disruption, which leads to enhancement of platelet aggregation, presence of antiphospholipid antibodies, presence of prothrombotic factors, such as deficiencies of protein S, protein C, and antithrombin, fibrinolysis anomaly due to endothelial cell damage after the deposition of antigen-antibody and elevated von Willebrand factor antigen levels (3-6). The main sufficient imaging tools that have been generally used for diagnosis of cardiac thrombosis are transesophageal and transthoracic echocardiography. The mass is usually heterogeneous and echogenic, involving the ventricles rather than the atria and the right ventricle more often than the left (3). Transesophageal and transthoracic echocardiography imaging might suggest a differential diagnosis a large vegetation, an intracardiac tumor such as a myxoma, or endomyocardial fibrosis but the clinical context helps to clarify the diagnosis (7).

In the present case, the differential diagnoses for the right ventricle thrombus were infective endocarditis and right ventricle tumors. Infective endocarditis was rule out based on the clinical, laboratory findings and the echocardiographic features of the cardiac mass. This current mass was not observed on his previous transthoracic echocardiography so, right ventricle tumors were also rule out. Based on his clinical course, laboratory results and the echocardiographic characterization of the multiple masses the cause of his death has been considered as embolism resulted from right ventricle thrombi. He was being treated with cyclophosphamide (1000 mg/month) and azathioprine 50 mg twice a day for BD.

In conclusion, we present an unusual case of multiple cardiac thrombosis in a patient with an established diagnosis of BD. It is suggested that Behçet's syndrome be kept in mind in the differential diagnosis of intracardiac thrombus. In addition, it is clear that the early echocardiography imaging is crucial to identify any cardiac involvement in patients with BD.

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Video 1. Modified right heart two-dimensional echocardiography views of multiple mobile thrombosis in right atrium and ventricle with normal right ventricle function

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