A 57-year-old female was referred because of an aortic valve vegetation. Medical history revealed aortic valve replacement 7 years ago, because of an aortic valve vegetation. Two years ago, patient underwent aortic valve re-replacement with a bioprosthetic valve because of a mechanical valve vegetation. Evaluation showed thrombotic and fibrotic vegetation and no signs of infection.

At the current presentation, the patient was asymptomatic. Medication consisted of acenocoumarol and acetyl salicylic acid (ASA). Physical examination revealed a systolic murmur (grade IV/VI) on the second right intercostal space. Laboratory results were normal except for a sedimentation rate of 21 mm/h. Blood cultures demonstrated no bacterial growth.

Echocardiography revealed an aortic valve bioprosthesis vegetation (Figure, left panel). Despite addition of unfractionated heparin, there was an increase in the initial vegetation accompanied by a mitral valve vegetation.

Haematological screening showed positive lupus anticoagulans (LAC) and elevated levels of cardiolipin IgG antibodies, suggesting antiphospholipid syndrome (APS). Unfractionated heparin was switched to low-molecular weight heparin (LMWH). Also, high-dose corticosteroids were administered. After 3 weeks, a decrease in initial vegetation and vanishing of the mitral valve vegetation were observed (Figure, right panel).

In our patient there was APS combined with non-bacterial thrombotic endocarditis or Libman-Sacks endocarditis. There was increased prevalence of valvular regurgitation, which could be caused by fibrinogenic and thrombogenic mass formation. The ineffectiveness of unfractionated heparin despite adequate activated partial thromboplastin time was solely caused by LAC. Therefore, switching unfractionated heparin to LMWH, continuation of acenocoumarol and ASA, and addition of corticosteroids finally resulted in adequate treatment of the recurrent cardiac vegetations.