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Recovery of non-bacterial thrombotic endocarditis and severe aortic regurgitation in a young patient with acute promyelocytic leukemia

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Introduction: Non-bacterial thrombotic endocarditis (NBTE) is characterised by non-infectious vegetations on the cardiac valves and is typically seen in patients with malignancies or systemic lupus erythematosus. Risk of thromboembolic events is high and treatment of choice consists of anticoagulation, if feasible, as well as treatment of the underlying disease. Severe valvular dysfunction is rarely seen and in these cases, surgical valve replacement is usually performed.

Case presentation: A 28-year old female patient presented with left-sided hemiparesis persisting for several hours. Angio-CT scan of the brain revealed a branch occlusion of the middle cerebral artery with a correlating infarct core and without an area of penumbra. Blood tests showed decreased thrombocyte levels ($45 \times 10^9/l$). Given the thrombocytopenia and the time between the onset of symptoms and presentation, no thrombolysis was administered.

A subsequently performed transoesophageal echocardiography for diagnostic work-up of cardioembolic sources of stroke revealed a right ventricular thrombus and vegetations on the aortic valve causing severe aortic regurgitation with a holodiastolic flow reversal in the descending aorta (Fig. 1 (a) and (b) showing a large central coaptation defect). There were no signs of infection or inflammation and blood cultures remained negative, thus infectious endocarditis was unlikely. A bone marrow biopsy led to the diagnosis of acute promyelocytic leukemia (PML). Despite the inherent high risk of bleeding due to PML, anticoagulation with unfractionated heparin was initiated and treatment with prednisolone, all-trans retinoic acid and arsenic trioxide was started. After three weeks of therapy aortic valve vegetation size decreased considerably leading to dramatic reduction of aortic regurgitation (Fig. 1 (c) and (d)). No further embolic event or bleeding occurred.

Conclusion: This case is remarkable in many aspects. First, PML typically causes bleeding complications whereas NBTE is characterised by a high risk of thromboembolic events. Thus, the decision to start or not to start anticoagulation is challenging.

Second, NBTE, unlike infectious endocarditis, rarely causes severe valvular dysfunction and reported treatment in most cases consisted of surgical valve replacement or repair. However, in this case, conservative treatment alone led to almost complete resolution of valvular vegetations and of aortic regurgitation.

We conclude that conservative treatment of severe valvular disease in the setting of NBTE should be strongly considered, especially given the small number of cases reported worldwide.

Abstract P630 Figure. Fig.1

