

Frantic About Marantic Valves

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Introduction

Nonbacterial Thrombotic Endocarditis (NBTE), also known as Marantic endocarditis, is a rare sterile endocarditis that most is often associated with malignancy and connective tissue diseases. Most reported cases have been associated with solid organ tumors though hematologic malignancies have been reported. It is associated with a high mortality rate and cases often require valve replacement. We present the case of a 46 year-old female with NBTE that was diagnosed with acute promyelocytic leukemia (APL), which represents the third reported case of this association.

Case Images

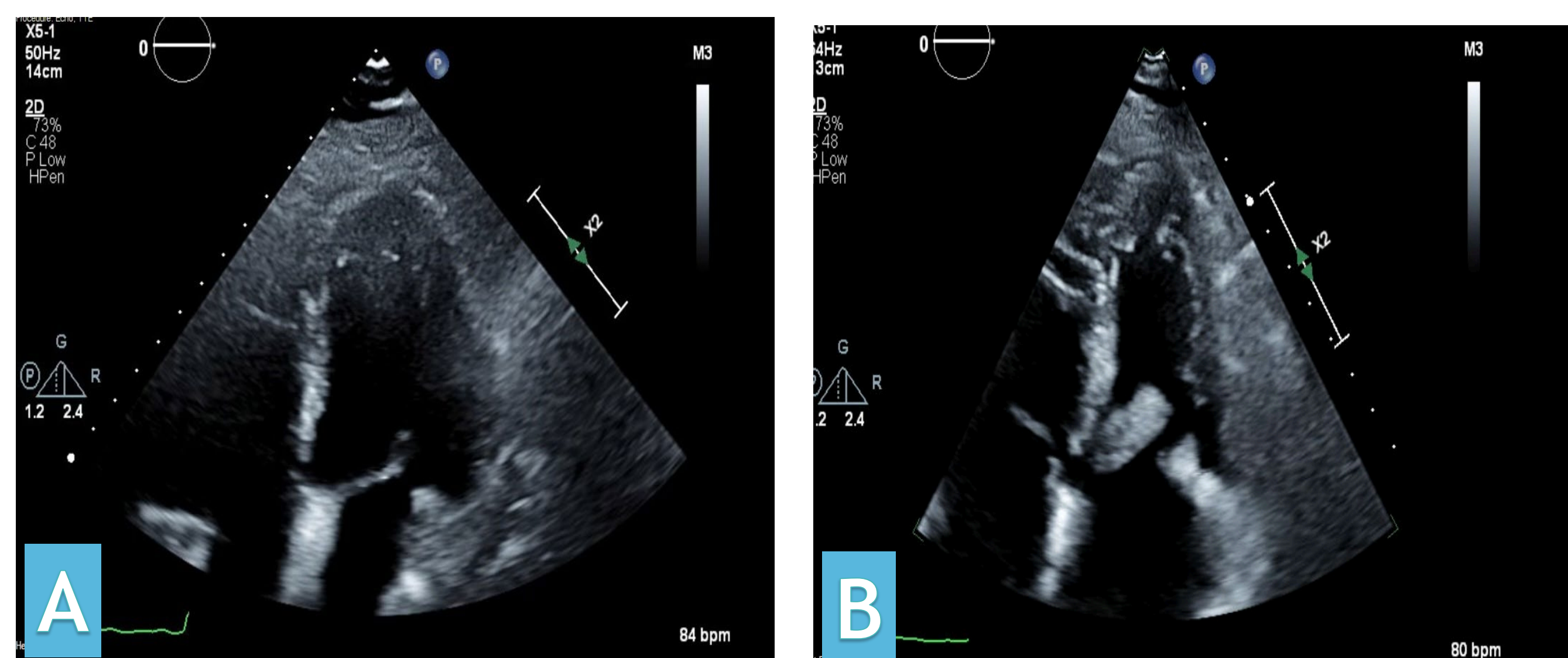


Figure 1: A. TTE from three months prior to presentation with normal mitral valve. B. TTE during hospitalization showing diffuse thickening of the mitral valve leaflets with decreased left ventricular cavity size and left atrial enlargement.

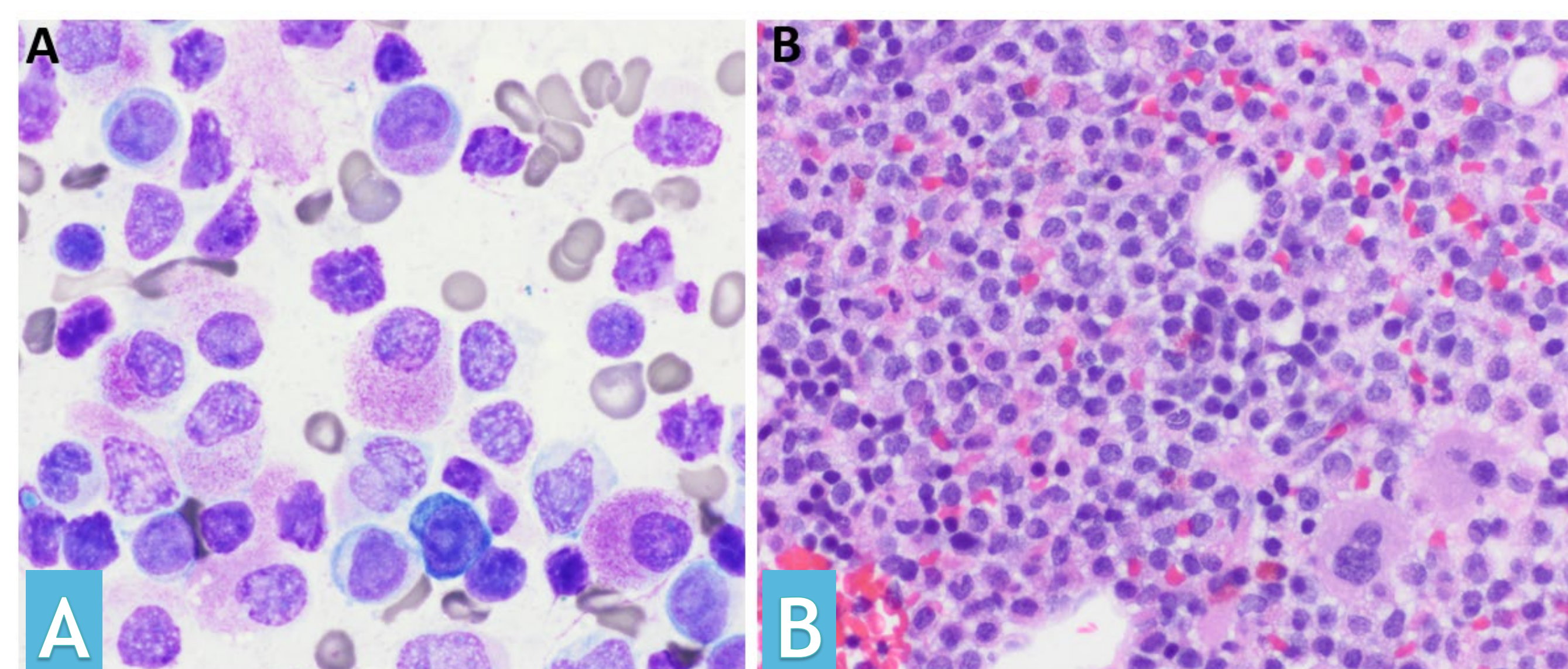


Figure 2: Bone marrow aspiration. A. Aspirate smear shows hypergranular neoplastic promyelocytes without Auer rods (Diff-Quik stain, 1000x). B. Aspirate clot section with granulocytic maturation arrest and partially preserved hematopoiesis (H&E stain, 500x).

Case Presentation

A 46-year-old female with recent history of cerebral vascular accident, deep vein thrombosis, and pulmonary embolism (on rivaroxaban) presented to the emergency department with worsening dyspnea. She was afebrile, hemodynamically stable, and on room air upon arrival. Physical exam demonstrated a soft diastolic murmur. Lab studies showed new pancytopenia with elevated INR of 2.3. Peripheral smear showed decreased cellularity but normal cell morphologies. Computed tomography angiography of chest showed acute splenic infarct and chronic pulmonary emboli. She was started on an intravenous heparin drip.

Transthoracic Echocardiogram (TTE) and subsequent Transesophageal Echocardiogram (TEE) showed echodense vegetations of the mitral valve leaflet tips consistent with nonbacterial thrombotic endocarditis (Figure 1). There was evidence of severe mitral valve stenosis with mean gradient of 17 mmHg. Severe pulmonary hypertension was noted with pulmonary artery systolic pressure of 62 mmHg. Blood cultures were negative. She was diagnosed with NBTE. She subsequently had right and left cardiac catheterization which showed preserved ejection fraction, reduced cardiac index of 1.87, and widely patent epicardial coronary circulation. She was started on diuretics for acute heart failure. Cardiac surgery was consulted for surgical replacement of her mitral valve.

Hypercoagulability work-up, including antiphospholipid antibodies, was negative. Serological work-up for connective tissue diseases including Lupus and Sjogren's syndrome was unrevealing. She underwent a bone marrow biopsy which demonstrated acute promyelocytic blasts without auer rods (Figure 2). Flow cytometry also showed promyelocytic blasts. Fluorescence in situ hybridization revealed t(15;17) PML-RARA fusion gene. She was diagnosed of acute promyelocytic leukemia. Evaluation for mitral valve replacement was suspended and she was then urgently transferred to a medical oncology unit at an outside institution for chemotherapy further staging and initiation of chemotherapy.

Discussion

- NBTE is a rare sterile endocarditis that involves fibrin-thrombi deposition on cardiac valve leaflets. Vegetations are more likely to embolize than those associated with infective endocarditis due to increased friability of the lesions [1].
- The lesions tend to form in areas of high flow on valvular leaflets and elevated levels of circulating cytokines have been reported. The most important pathogenic factor is likely hypercoagulability [2].
- Most patients are aged 40 to 80, with men and women being affected equally [4].
- The most prevalent associations were malignancy (52.1%; lung, ovarian, pancreatic) and connective tissue disease (37.5%; SLE, APS) [5]. Though there are no large epidemiological studies, current data shows only 3% of cases are associated with hematologic malignancy [3].
- Outcomes for NBTE are poor, with malignancy-associated cases having a 1-year mortality rate as high as 48% [5].
- Delays in etiological identification and treatment can impact patient survival.

References

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