



A Rare Case of Ovarian Tumor Presenting as Cerebral Embolism from Nonbacterial Thrombotic Endocarditis with Giant Vegetation

Li Z[#], Dai T[#], Yang Y¹, Lujia Ni^{2*} and Daoyuan Si^{1*}

¹Department of Cardiology, China-Japan Union Hospital of Jilin University, Jilin Provincial Engineering Laboratory for Endothelial Function, China

²Department of Ultrasound, China-Japan Union Hospital of Jilin University, China

[#]These authors contributed equally to this work

Abstract

Background: Nonbacterial Thrombotic Endocarditis (NBTE) is a rare condition characterized with non-infection vegetation on cardiac valves which usually leads to systematic embolism. Large vegetations are rare, however, we report a case of NBTE with giant vegetations, resulting in multiple organ embolism in brain, lung and lower limb arteries. Further examination revealed an ovarian tumor.

Case Report: A 54-year-old female was referred to our hospital with multiple cerebral infarctions. Of note, she had bilateral lower extremity deep vein thrombosis and bilateral pulmonary arterial branches embolism 3 months ago. Therefore, she is currently on anticoagulant therapy. Subsequent transthoracic echocardiograms demonstrated giant vegetations on anterior and posterior mitral leaflet. Serial blood cultures and procalcitonin remained negative raising suspicion for nonbacterial thrombotic endocarditis. Further examination of an unexpected ovarian tumor confirmed this diagnosis.

Conclusion: This current report demonstrates that the possibility of NBTE should be considered especially in a patient suffering from vegetation on cardiac valves presenting with the embolic phenomenon, and further causes should be actively sought such as malignancy, autoimmune disease and so on. Strive for early diagnosis and treatment to improve the survival rate of these patients.

Keywords: Nonbacterial thrombotic endocarditis; Cancer-associated thrombosis; Stroke; Case report

Introduction

Nonbacterial Thrombotic Endocarditis (NBTE) or marantic endocarditis is a rare form of non-infectious endocarditis associated with advanced stage malignancy, autoimmune disease, connective tissue disease, hypercoagulable states and so on [1,2]. It is characterized by the presence of sterile vegetation, predominantly on the mitral or aortic valves due to platelet and fibrin deposits [3]. In this report, we described a case of NBTE with giant vegetation on mitral valve presenting as cerebral embolism and multiple lower extremity arterial embolism. Further examination of the patient revealed a new diagnosis of ovarian cancer.

Case Presentation

A 54-year-old female presented to our hospital with left-sided hemiparesis, which was manifested as inability to walk independently and lift the arm. The patient's medical history was notable for bilateral lower extremity deep vein thrombosis and bilateral pulmonary arterial branches embolism 3 months prior to this presentation and an inferior vena cava filter had been placed 2 months earlier.

Initial laboratory tests revealed prolonged prothrombin time (15.2s, 9.4-12.5) and thrombin time (20.4s, 10.3-16.6), raised D-dimers (21.4 µg/mL, 0-0.5) and reduced fibrinogen (0.9 g/L, 2-4). Brain Magnetic Resonance Imaging (MRI) (Figures 1A-1D) and diffusion-weighted imaging (Figure 1E, 1F) indicated multiple infarcts. In combination, these clinical and imaging findings were suspicious for a thromboembolic cerebrovascular accident and she was started on anticoagulant and antiplatelet therapy.

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*Correspondence:

Lujia Ni, Department of Ultrasound,
China-Japan Union Hospital of
Jilin University, Xiantai Street No.
126, Changchun, Jilin, China, Tel:
(+86)13943187009; Fax: (+86)431-
84995104;

E-mail: nilujia2012@jlu.edu.cn

Daoyuan Si, Department of Cardiology,
China-Japan Union Hospital of Jilin
University, Jilin Provincial Engineering
Laboratory for Endothelial Function,
China;

E-mail: Nil

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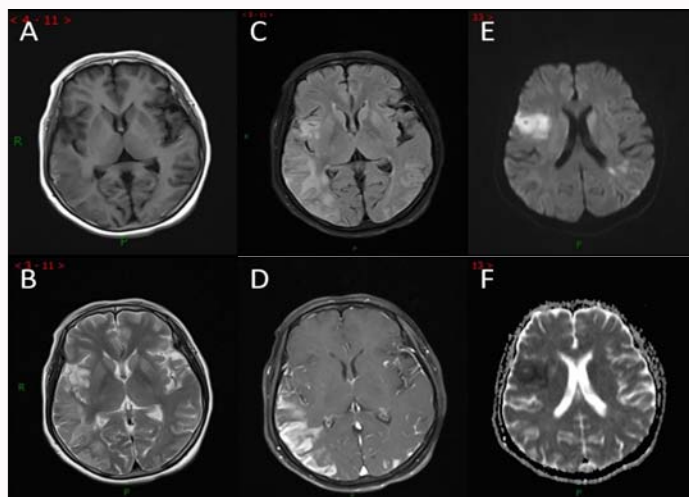


Figure 1: Brain magnetic resonance imaging and diffusion-weighted imaging on admission. (A-D) Brain magnetic resonance imaging on admission. (E, F) Diffusion-weighted imaging on admission.

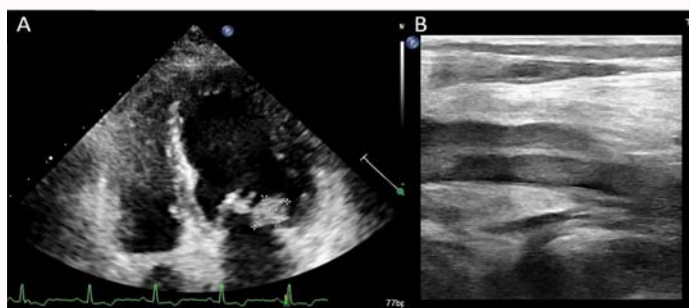


Figure 2: Transthoracic echocardiography and Lower limb arterial ultrasound on presentation. (A) Transthoracic echocardiography on presentation. (B) Lower limb arterial ultrasound on presentation.

During her hospitalization, the patient continued to undergo workup for her cerebrovascular accident. Transthoracic Echocardiography (TTE) revealed severe mitral valve insufficiency as well as two masses over 10 mm in diameter (10.4 mm × 7.4 mm, 16.5 mm × 11.8 mm) attached to the anterior and posterior mitral leaflet, pulmonary hypertension and left atrial enlargement (Figure 2A). Then left lower extremity arterial embolism was also found (Figure 2B). As a consequence, anticoagulation management began with low-molecular-weight heparin twice daily, and therapy with oral anticoagulation of rivaroxaban at a dosage of 20 mg once daily was started.

Infective Endocarditis (IE) was first suspected, however, the patient remained afebrile without a leukocytosis, serial blood cultures and procalcitonin were also remained negative. No evidence of IE was evident. Multiple blood cultures showed no growth raising suspicion for NBTE. A broad workup was initiated for causes of culture-negative and NBTE. Autoimmune labs were negative, including antinuclear antibody and antiphospholipid antibody screen. The patient had a sudden onset of dysuria and hematuria, and bladder ultrasound revealed unexpected ovarian occupancy. An enhanced computed tomography of the abdomen revealed a 7.5 cm × 5.4 cm soft tissue mass in the right accessory area. CA-125 was elevated at 218 (normal <35 U/mL). In light of negative blood cultures, mitral valve vegetations on echocardiogram, massive thromboembolism and ovarian cancer, a diagnosis of NBTE was made.

The patient refused biopsy and discharged with continued anticoagulation and neoadjuvant treatment.

Discussion

NBTE refers to a spectrum of lesions ranging from microscopic aggregates of platelet and fibrin to large vegetations on previously undamaged heart valves in the absence of a bloodstream bacterial infection [4]. Valves most commonly affected include aortic, mitral, or a combination of both. The vegetations in NBTE are usually small (<1 cm), broad based and irregular in shape [1]. They can also easily dislodge and readily cause multiple systemic embolic infarcts, especially the cerebral vascular bed [5]. There is a strong association between NBTE and neoplastic disease. Key points of malignancy-associated NBTE likely include hypercoagulable state and elevated levels of circulating cytokines, such as tumor necrosis factor, interleukin-1 and 6, resulting in local tissue damage that promotes vegetation formation [3].

Antemortem diagnosis was once rare, therefore NBTE was often undervalued. With improved imaging modalities, more NBTE can be diagnosed, of which echocardiography is critical. However, TTE may not reveal any valvular vegetation of size less than 3 mm because of its image resolution. Therefore, Transesophageal Echocardiography (TEE) is often preferred over TTE for NBTE, which usually presents as smaller vegetations. Roldan et al. compared the diagnostic accuracy of TTE compared to TEE in detecting Libman-Sacks endocarditis

[6]. He considered TEE is superior to TTE and should be considered either as complement to a nondiagnostic TTE.

In the current case, our patient presented with cerebral embolism and MRI showed indicated multiple infarcts. He was also on long-term anticoagulant therapy because of a medical history of bilateral lower extremity deep vein thrombosis and bilateral pulmonary arterial branches embolism. On ultrasound, he was found to have two large vegetations over 10 mm with high mobility attached to the mitral valve. Considering the history and imaging findings, the diagnosis of multiple cerebral embolisms was considered rather than orthotopic cerebral infarction. According to echocardiogram, the diagnose of endocarditis is established, including infective and non-infective. Because of multiple negative cultures and serology, the patient was more likely to be diagnosed with NBTE. As we continue to search for the possible cause, there was an accidental discovery of a huge ovarian mass due to hematuria. This supports the diagnosis of NBTE. Patients with a cardioembolic event and echocardiographic evidence of vegetation in the absence of hemodynamically significant valvular heart disease and no evidence of an infective process should be inclined to suspect NBTE.

Thromboembolic events (particularly stroke) are a common presentation for NBTE because of the more friable vegetations. Systemic embolism occurs in nearly half of patients with NBTE affecting mainly cerebral, lung, coronary, renal, and mesenteric circulations, among them neurological symptoms being the most common presentation [7]. It is in concordance with our case report. In other cases, however, the vegetations are often small and undetectable by TTE. Our patient had giant vegetations that were easily detected by ultrasound.

In conclusion, we report a case of cerebral embolism due to NBTE in a patient with a newfound malignant disease. Echocardiogram is crucial in the diagnosis of NBTE. The possibility of NBTE should be considered even if the vegetation is large, and further causes should be actively sought. Early diagnosis is crucial for establishing rapid treatment to prevent further valvular damage and systemic embolization.

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