



Aneurysms of the Sinus of Valsalva in a Patient with Behçet's Disease

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Abstract

A 38-year-old man with a history of recurrent genital ulceration initially diagnosed as genital herpes was admitted after presenting with paroxysmal nocturnal dyspnea. Echocardiography revealed aneurysm formation of the sinus of Valsalva. On diagnosis of an aneurysm of the sinus of Valsalva associated with Behçet's disease (BD), surgeon repaired the affected sinus only, and continuous steroid therapy maintained the integrity.

Keywords: Behçet's disease, Aneurysms, Sinus of Valsalva

Introduction

BD is a multisystemic disease that may affect any organ in different combinations, characterized by recurrent oral aphthous ulcers, genital ulcers, uveitis, and skin lesions. Involvement of the heart is called Cardio-BD, which includes pericarditis, myocarditis, endocarditis, endomyocardial fibrosis, atrial fibrillation, ventricular arrhythmias, coronary arteritis, acute myocardial infarction, and dilated cardiomyopathy(1). We report a rare case of this syndrome, associated with aneurysm of the non-coronary sinuses of Valsalva, resulting in symptomatic aortic valve regurgitation.

Case Report

A 38-year-old man was referred for investigation of a 5-day history of paroxysmal nocturnal dyspnea in May 2008. He had a three-year history of recurrent genital ulceration initially diagnosed as genital herpes, but never confirmed on culture or nucleic acid amplification testing; sequential treatment with aciclovir over a four-month period did

not alleviate his genital symptoms. And he had no history of cardiac or respiratory disease, nor any cardiac risk factors. His family history included his brother, sister and his nephew had recurrent aphthous. Clinical examination revealed aphthous stomatitis, genital ulceration, erythema nodosum bilaterally and a grade 3/6 systolic murmur, audible at the 2nd intercostal space on the left sternal border. Hematologic examination showed that high sensitive C-reactive protein (hs-CRP) was $105 \text{ mg}\cdot\text{L}^{-1}$ and erythrocyte sedimentation rate (ESR) is $70 \text{ mm}\cdot\text{hr}^{-1}$. Anti nuclear antibody (ANA), anti double strand DNA antibody (anti ds DNA) and muscle biopsy specimens for connective tissue disease were negative. His electrocardiogram showed Sinus tachycardia. Chest X-Ray revealed cardiomegaly with a cardiothoracic ratio of 55% and a slightly widened mediastinum. Echocardiography revealed the non-coronary-sinus of Valsalva aneurysm (Fig. 1). The left ventricular (LV) end-diastolic was 58 mm, and the LV ejection fraction was 53%. Based on these

findings, the diagnosis of aneurysm of the sinus of Valsalva associated with BD was made.

Because the patient's condition was in the active phase, prednisolone was administered over one month until the inflammatory signs (hsCRP and ESR) became negative. Considered that our patient presented a competent valve and only one sinus involved, surgeon repaired the affected sinus

only. Oral administration of prednisolone was started on the first postoperative day. No aortic regurgitation was detected on postoperative transthoracic echocardiography. He was discharged without any complications on postoperative day 15, and was doing well at five-year follow-up after the operation.

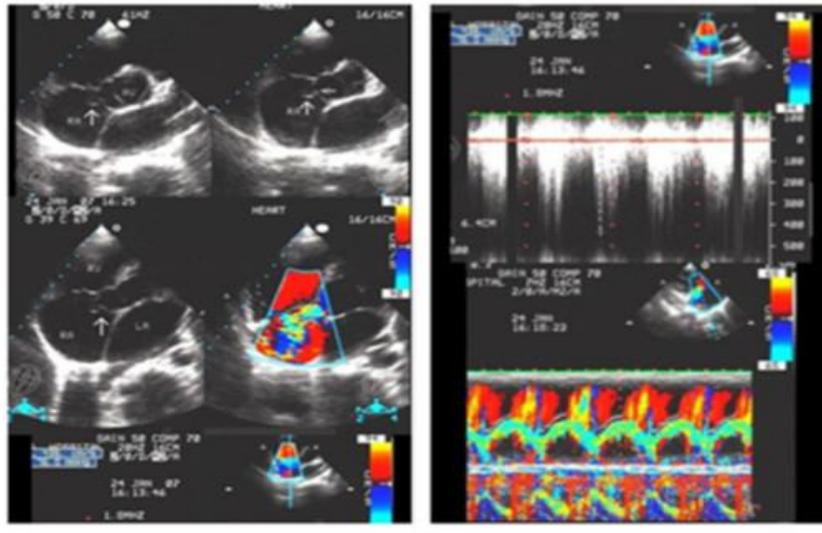


Fig.1: showing non-coronary-sinus of Valsalva aneurysm

Discussion

Aneurysms of the sinus of Valsalva's etiologies include atherosclerosis, infection such as syphilis, congenital disease such as Marfan's syndrome, trauma, and autoimmune disease such as BD. BD is a chronic, relapsing multisystem vasculitis with predominant involvement of the oral and genital mucosa (1, 2). BD has a worldwide distribution but is prevalent in Japan, the Middle East, and some Mediterranean countries, and it often affects younger adults and is more common in men than women. James et al. demonstrated that Valsalva sinus aneurysm was the leading causes of death in patients with BD (3).

In our patient, echocardiography revealed aneurysm formation of the sinus of Valsalva, and should BD is the etiology? The patient had a history of recurrent genital ulceration, on admission,

clinical examination revealed aphthous stomatitis, genital ulceration, and erythema nodosum bilaterally, which could provide clues to differentiate diagnosis. If neglected these symptoms, it maybe at a loss of initial diagnosis of genital herpes. In addition to recurrent genital ulceration, aphthous ulcers and skin lesion, a clinical diagnosis of BD should be made, because other examination showed that high CRP and ESR, and ANA, anti dsDNA and muscle biopsy specimens for connective tissue disease were negative.

Aneurysms of the sinus of Valsalva may incur complications if untreated, such as right ventricular outflow tract obstruction, coronary artery occlusion, aortic regurgitation and congestive heart failure, so surgical treatment should be considered (4, 5). But to aim directly at etiology is the most of all, and Continuous steroid and immunosuppressive agent therapy are most important to maintain the integrity to prohibit recur-

rence of BD. Our patient was doing well at five-year follow-up after the operation. It also highlights the importance of recognizing systemic disease and maintaining a holistic approach when treating patients.

Acknowledgement

The authors declare that there is no conflict of interests.

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