A large left ventricular pseudoaneurysm in a case of Behçet's disease and literature review

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Abstract

Behçet’s disease is a collagen-vascular disease which is most commonly seen in Asia and Mediterranean area. It can cause involvement of different organs and systems including cardiovascular system. Pseudoaneurysm is the most common form of arterial involvement in Behçet’s disease, however, cardiac pseudoaneurysm is rare. We are presenting a 13 year-old boy with a 4-year history of Behçet’s disease who was admitted because of cough, chills and fever, and chest pain. A very large pseudoaneurysm in his left ventricle was found in further workups. He was treated surgically and was followed up without any complications for 24 months.

Keywords: Behçet’s disease, cardiac, left ventricular, pseudoaneurysm
Background

In 1937 a Turkish dermatologist, Hulusi Behçet, first described a chronic autoimmune disease bearing his name with characteristic orogenital aphtous ulceration and uveitis (1). The etiology of this disease, which is most common in Asia and Mediterranean area, is still unknown. The primary pathology is a vasculitis affecting skin, joints, and pulmonary, gastrointestinal, urinary and nervous systems (2). Its vascular complications are most frequently manifested as thromboembolism in veins and pseudoaneurysm in arteries (3). Although pseudoaneurysms are the most common form of arterial involvement in Behçet’s disease, we could only find one case reported by Rolland et al. (4) with Behçet’s disease and cardiac pseudoaneurysm. Occasional cases of cardiac pseudoaneurysms have been reported in association with rheumatoid arthritis (5) and Kawasaki’s disease(6); yet, large cardiac pseudoaneurysms are usually complications of myocardial infarction, endocarditis, chest trauma, and surgical manipulations (7).

In this study we present a patient with Behçet’s disease with a large left ventricular pseudoaneurysm.
Case presentation

A 13-year-old boy with Behçet’s disease was referred to our hospital with chills, fever, cough and chest pain of one month duration in June 2001. The diagnosis of Behçet’s disease was established 4 years prior to this admission and had presented with oral aphtae, orchitis, right eye uveitis leading to blindness, recurrent pseudofolliculitis, knee arthritis, and lower extremity deep vein thrombosis, all attributable to his autoimmune disorder. He had been treated by Prednisolone (15 mg/day) and Methotrexate (7.5 mg/week) for last 7 month.

Physical examination revealed III/VI to-and-fro murmur along the left sternal border, and an S3 gallop.

The PA chest X-ray (Fig 1.), Computed tomography scan with IV contrast media at the level of T5-T8 (Fig. 2), Magnetic resonance imaging (MRI) (Fig. 3), coronary angiography and echocardiography all revealed a 10.1*14.8 cm left ventricular pseudoaneurysm. At surgery the ostium (arrowhead in Fig. 4) was measured 2.5*3.0 cm.

The above mentioned results warranted a surgery with Median Sternotomy approach. After pericardotomy, a pulsatile mass appeared at the tip of LV with a fistula to the heart. The pseudoaneurysm and large amounts of thrombus within it were resected, the defect in the left ventricular wall was repaired by Teflon Plegeted Prolen 4/0.

The patient made an uneventful recovery. The pathologic examination results revealed a fibrous pseudoaneurysm including areas of old hemorrhage and thrombosis, and chronic inflammation.

The patient was followed up without any complications for 24 months after the operation.
Conclusions

We have presented a 13 year-old boy with a large (10.1*14.8 cm) left ventricular pseudoaneurysm in association with Behçet’s disease which is rare.

Behçet's disease is a systemic disorder with mucocutaneous, ophthalmic, neurological, cardiovascular, pulmonary, gastrointestinal, urogenital and musculoskeletal involvement. Its vascular manifestations are thrombophlebitis and, less frequently, arterial lesions such as pseudoaneurysms and occlusions/stenoses (8). About 8% of the patients with Behçet's disease have severe vascular complications such as arterial pseudoaneurysms and occlusions (1). Pseudoaneurysms are the most common form of arterial involvement in Behçet’s disease (3).

Cardiac involvement is rare in Behçet's disease (9). Only about 6% of patients with Behçet's disease have this complication (4).

Cardiac involvement in this disorder is a diffuse process which involves both cardiac structure and vascular elements. Higher incidences of interatrial septum aneurysm (6% to 31%), mitral valve prolapse (3% to 25%), mitral regurgitation (6% to 40%) and aneurysmal dilatations of valsalva sinus and ascending aorta were observed in the Behçet's disease patients than in the normal subjects (10).

Although Left ventricular aneurysms with Behçet's disease have occasionally been reported, we found only one case of cardiac pseudoaneurysm in these patients reported in literature. Rolland et al. (4) reported a 29 year old patient with Behçet's syndrome and a false left ventricular aneurysm and coronary artery aneurysm which were repaired under cardiopulmonary bypass with no postoperative complications.
Cardiac pseudoaneurysm is defined as a rupture of the myocardium that is contained by pericardial adhesions or the epicardial wall. This phenomenon can be explained by myocardial fragility induced by ischaemia due to vasculitis process of Behçet’s disease (4). In our case, pseudoaneurysm was resulted from rupture of the left ventricle.

In contrast to a true ventricular aneurysm, in which the wall is composed of myocardial scar tissue, the wall of a pseudoaneurysm is composed of thick fibrous tissue and pericardium (11). In our case, pseudoaneurysm was consisted of profuse fibrous tissues and thromboses in various stages.

These pseudoaneurysms have the potential to leak, rupture, or can be the source of peripheral emboli (10). Different reports recommended that such a contained rupture has a greater propensity for rupture than a true aneurysm, whose wall contains myocardium. Rupture of a left ventricular pseudoaneurysm is usually fatal, so appropriate recognition and treatment (early surgery) -even for asymptomatic patients- was recommended (7).

The diagnosis of pseudoaneurysm is rarely suggested by clinical signs and symptoms and can be difficult to diagnose (7). In our case the pseudoaneurysm presented with nonspecific symptoms and signs. Thus, such a diagnosis was highly unlikely before getting the results of imaging.

Various imaging methods have been used to diagnose pseudoaneurysm, including two-dimensional echocardiography, computed tomography, magnetic resonance imaging, and left ventricular angiography. Each has its advantages and disadvantages but echocardiography has become the most common examination used for first recognition because it can evaluate other associations, such as valvular regurgitation, thrombus formation, and ventricular function, are often important additions to clinical management (7).
Like our case, chest radiography sometimes shows a localized bulge on the cardiac silhouette. On computed tomography, pseudoaneurysms are characterized by an abrupt disappearance of the myocardial wall at the border of the pseudoaneurysm. Magnetic resonance imaging shows the low signal of the pericardium, which constitutes the only wall of the pseudoaneurysm (7).

Regarding its fatality and nonspecific manifestations, one should consider cardiac pseudoaneurysms as a potential risk in any patient with Behçet’s disease. Thanks to early diagnosis and surgery, he was treated successfully and had no complications in a follow-up period of 24 months duration.

**List of abbreviations used**

SVC = Superior Vena Cava  
IVC = Inferior Vena Cava  
LV = Left Ventricle

**Competing interests**

Authors declare that there is no financial or non-financial competing interests

**Authors' contributions**

SMM: Data collection, participated in the design of the study, critical review of the manuscript.

PE: conceived the study and wrote the manuscript, participated in the design of the study.

All authors read and approved the final manuscript.
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References


Figure legends:

**Figure 1:** The PA chest X-ray: left-sided pleural effusion and a large mass in anterolateral part of left lung which had overshadowed the left border of the heart and caused the shifting of the heart to the right side (Arrowhead).

**Figures 2:** Chest computed tomographic scan with contrast at the level of T7 showing the large pseudoaneurysm. The lesion was a well-defined partially calcified mass with tubular density adjacent to the heart. Arrowheads indicate the calcifications. It was filled with contrast medium concurrently with the heart. This lesion, which was mostly occupied by thrombosis, had a mass effect on heart.

**Figure 3:** The MRI indicated a mass with inhomogeneous signals implying the presence of blood and clots in addition to calcification. (SVC = Superior Vena Cava; IVC = Inferior Vena Cava; LV = Left Ventricle; P = Pseudoaneurysm)

**Figures 4:** Chest computed tomographic scan with contrast at T7 level. Arrowhead marks the ostium.