

Case Report

A case with giant right coronary artery aneurysm in a patient with Behçet's disease

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KEYWORDS

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Abstract

Behçet's disease is a chronic, relapsing, multi-systemic inflammatory disorder characterized by recurrent orogenital aphthous ulcers, uveitis, and skin lesions. Vascular involvement affects the veins more commonly than the arteries of these patients. Arterial involvement may include aneurysm formation, occlusive disease, and stenosis, with the most common sites being the abdominal aorta and femoral and pulmonary arteries. Thus, coronary arterial involvement is very rare but can cause serious complications when it occurs. In this report, we present a 28-year-old male patient with Behçet's disease who developed a giant right coronary artery aneurysm with minimal cardiac manifestation of a recently developed slight palpitation. The patient underwent surgical repair. We present the results of cardiovascular imaging and histopathological examinations. Our case demonstrates the necessity of regular cardiovascular examination for patients with Behçet's disease, even for those without cardiac symptoms.

Case report

A 28-year-old male had a positive fecal occult blood test. Colonoscopy demonstrated that there was a deformity in the ileocecal valve and semicircular ulceration was observed from the ileac side of the ileocecal valve to the top of the valve. The floor of the ulcer was covered with granulomatous tissue. A biopsy sample from the ileocecal valve showed chronic inflammatory change. He had no history of oral ulcer, genital ulcer, or visual disturbance, and was diagnosed with intestinal Behcet's disease. He was transferred to the Department of Gastroenterology of our institution and treated for Behcet's disease with infliximab and low-dose steroid. During the course, he felt slight palpitation and was referred to our department. His height was 151.3 cm and body weight was 42.9kg. He had no history of Kawasaki disease, scarlet fever, infantile polyarteritis nodosa, or traumatic injury. Laboratory tests revealed a white blood cell count of $10,400/\mu\text{L}$ without eosinophilia; hemoglobin of 13.9 g/dL; platelet count of $179,000/\mu\text{L}$; C-reactive protein of 0.5 mg/dL. Cardiac enzymes and serum levels of electrolytes including Na^+ , K^+ , Ca^{2+} , and phosphate were all normal. On physical examination, he had a temperature of 36.5°C with a respiratory rate of 20/min. He had no skin rash or muscle or joint ache. The blood pressure was 120/70 mmHg without differences between both arms, and the pulse was 70/min. He had a Levine III/VI systolic murmur at the apex and moist rale in both lungs. Chest radiography showed right atrial enlargement (Fig. 1A), and an electrocardiogram (ECG) showed sinus rhythm, right axis deviation, and tall P- and negative T-waves, which suggested the existence of right atrial and ventricular overload (Fig. 1B). Transthoracic echocardiography revealed a giant pericardial mass compressing the right atrium and ventricle (Fig. 1C). The patient underwent cardiac computed tomography (CT) imaging

for the diagnosis of the cardiac mass. There was a 90×79×84 mm mass on the apico-inferior side of the heart, and the mass was connected with the right coronary artery (Fig. 1D). The mass was inhomogeneously enhanced with contrast material, which suggested that it was a giant coronary artery aneurysm with thrombus formation. There were no abnormalities of the aorta, abdominal arteries, or venous systems. Cardiac magnetic resonance imaging confirmed the diagnosis (Fig. 2A). It also indicated that the mass was connected to the right coronary artery. A T1-weighted image indicated that there was blood flow inside the aneurysm, and co-axial layer enhancement indicated mural thrombus formation inside the giant coronary aneurysm (Fig. 2B). Coronary angiography revealed that he had a normal left coronary artery. The proximal part of the right coronary artery was also normal but there was a giant coronary aneurysm in the distal part of the right coronary artery (Fig. 2C). Left ventriculography in a right anterior oblique view showed compression of the left ventricle by the enlarged aneurysm. Mitral regurgitation (II~III/IV) was also observed. The patient underwent an operation to remove the aneurysm. After resection of the aneurysm, there was still moderate to severe mitral regurgitation due to mitral valve prolapse, and mitral valve plasty was conducted. The posterior papillary muscle was atrophic and most of the chordal tendons were attached to the anterior papillary muscle. There were no complications in the post-operative period. ECG recording showed the disappearance of tall P- and negative- T waves in II, III and aV_F. Pathologic specimens of the aneurysm confirmed that there was no elastic lamina in the aneurysm wall, and the vascular wall had almost been replaced with fibrous tissue (Fig. 3A and B). Inflammatory cell accumulation was also observed, which may represent ongoing chronic activity of Behçet's disease (Fig. 3C).

Discussion

Behçet's disease exists worldwide but is more prevalent in areas along the ancient silk route spanning from Japan to the Middle East and Mediterranean countries [1]. It is an autoimmune disease characterized by oral aphthosis, genital ulcers, and ocular lesions. Vasculo-Behçet's is a concept that has been adopted for cases in which vascular manifestations dominate the clinical features of this disease. Unlike other forms of inflammatory vasculitis, Behçet's disease can involve both the arterial and venous systems, with more frequent manifestations in the venous system. Venous disease commonly consists of superficial thrombophlebitis, thrombosis of large veins, and cerebral venous sinuses. Arterial lesions account for 12% of all vascular complications and most frequently affect the abdominal aorta, followed by the thoracic aorta and pulmonary, iliac, and femoral arteries [2,3].

Coronary arterial involvement in Behçet's disease is very rare, and the prevalence is reportedly 0.5% [4,5]. Aneurysms are seen less frequently than arterial occlusions [6]. The pathogenesis of the arterial lesion in Behçet's disease has been well documented. Active arteritis occurs initially, followed by destruction of the media and fibrosis. Saccular aneurysms are probably produced by severe destruction of the media through intense active inflammation [7].

Because coronary aneurysms are rare, an evidence-based approach is difficult to establish. Most of the patients are managed on the basis of its size and clinical symptoms. Resection is generally considered in symptomatic patients who are good candidates for surgery. However, surgical intervention for asymptomatic patients is controversial. In the largest reported cohort of coronary aneurysms (CASS), Swaye et al. did not find any case of rupture in 978 patients [8]. In a recent review, Burns et al.

commented that, since the advent of angiography and the premortem diagnosis of coronary aneurysm, no case of rupture has been documented [9]. This led the authors to conclude that, despite the theoretical risk of coronary aneurysm rupture, this outcome seems extremely rare and unpredictable. However, it can cause serious complications when it occurs, and cardiac tamponade is reportedly due to rupture of a right coronary artery aneurysm [10]. Thus, surgical resection was performed in the present case.

Although this patient's entire history of Behçet's disease is unknown, a giant aneurysm was formed in his coronary artery, which may represent silent, ongoing chronic inflammation for years without any manifestations. In this case, this patient's complaint was only slight palpitation, and the diagnosis of coronary aneurysm would not have been made if the cardiovascular examinations had not been performed.

Conclusion

Thus, our case demonstrates the necessity of the regular noninvasive check-up of coronary arteries even without cardiovascular-thoracic manifestations in patients with Behçet's disease.

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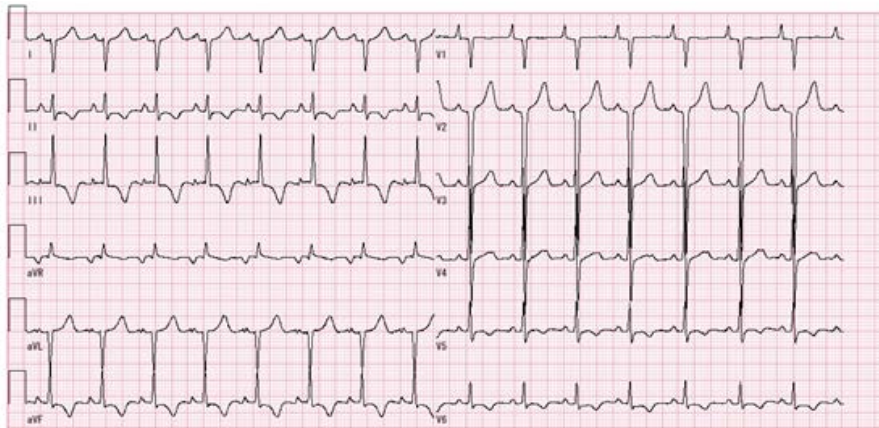
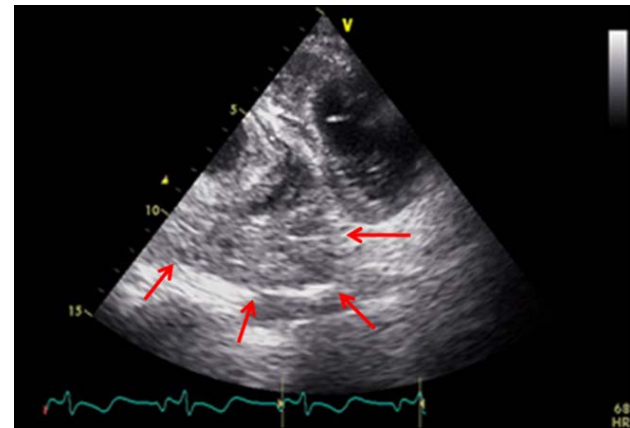
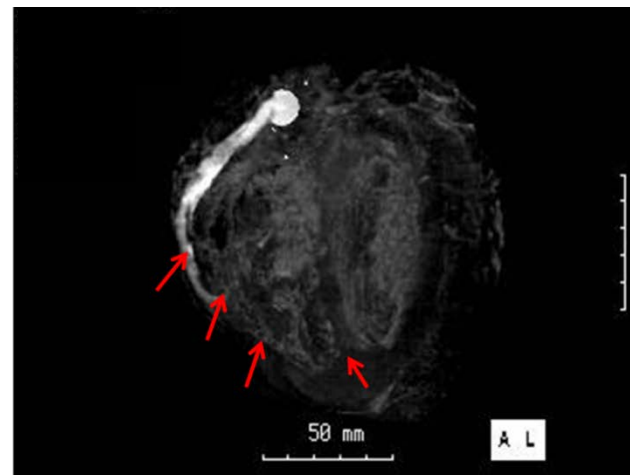
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Figure 1. (A) Chest X-ray demonstrates the enlarged second arc of the cardiac shadow. (B) Electrocardiogram shows right axis deviation, and tall P- and negative T-waves in II, III, and aV_F. (C) Echocardiography presents a parasternal short axis view showing a mass adjacent to the apex of the left ventricle. (D) Coronary CT angiography showing an aneurysm in the distal part of the right coronary artery.

Figure 2. (A) T1-weighted right anterior oblique image of the aneurysm. (B) T1-weighted axial image of the right coronary artery aneurysm, which indicates the existence of a mural thrombus. (C) Imaging by cardiac catheterization. Coronary arteriography indicates aneurysm of the right coronary artery.

Figure 3.

Histological findings of the aneurysmal wall. (A) Hematoxylin-eosin staining indicates severe destruction of the aneurysmal wall. (B) Elastica van Gieson's staining showing the vascular wall almost fully replaced with fibrous tissue. (C) Focal accumulation of inflammatory cells.

A**B****C****D**

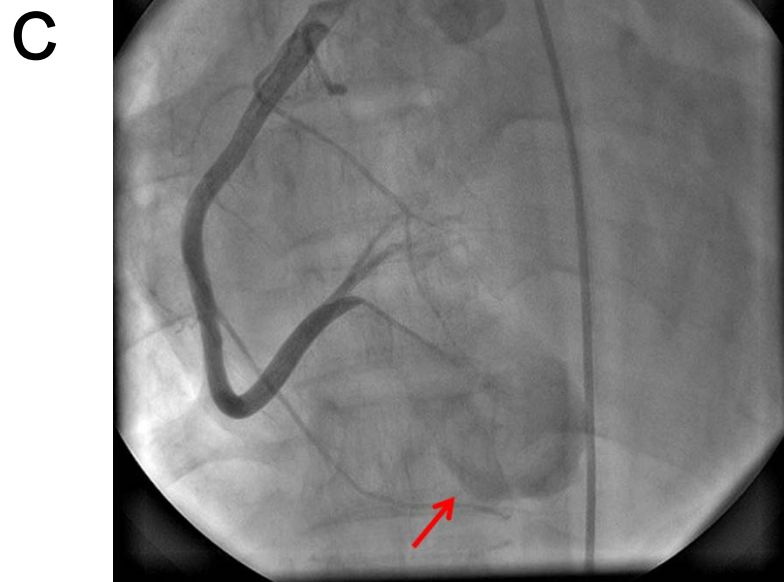
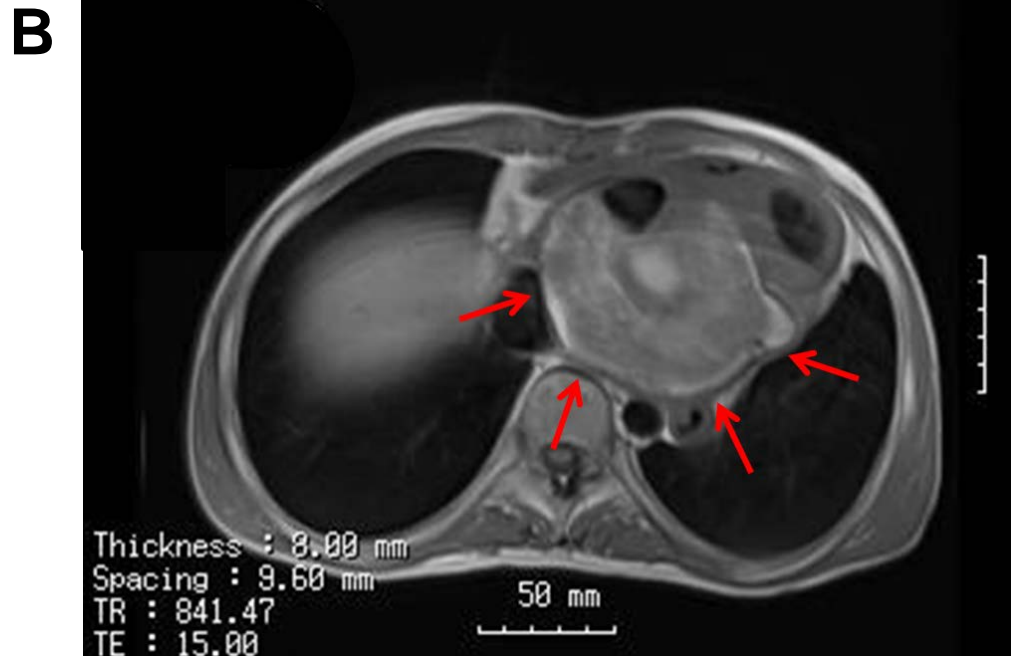
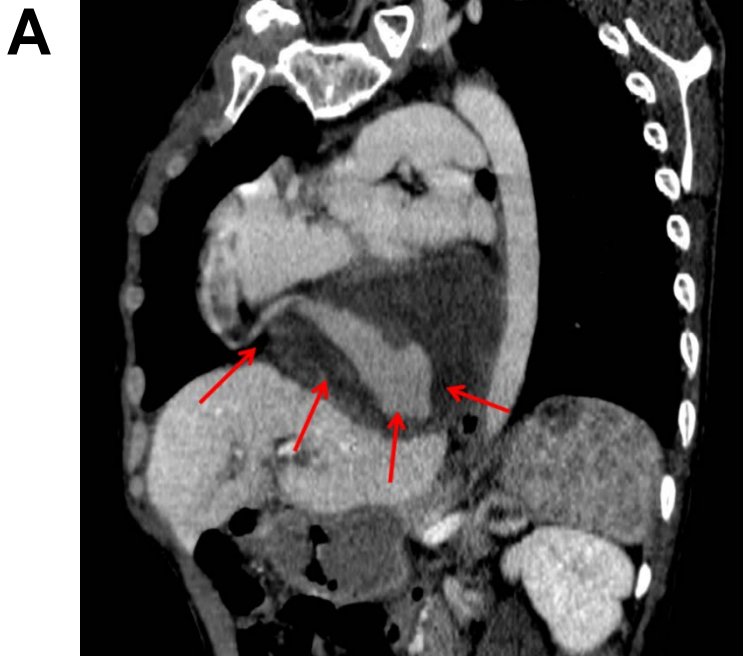
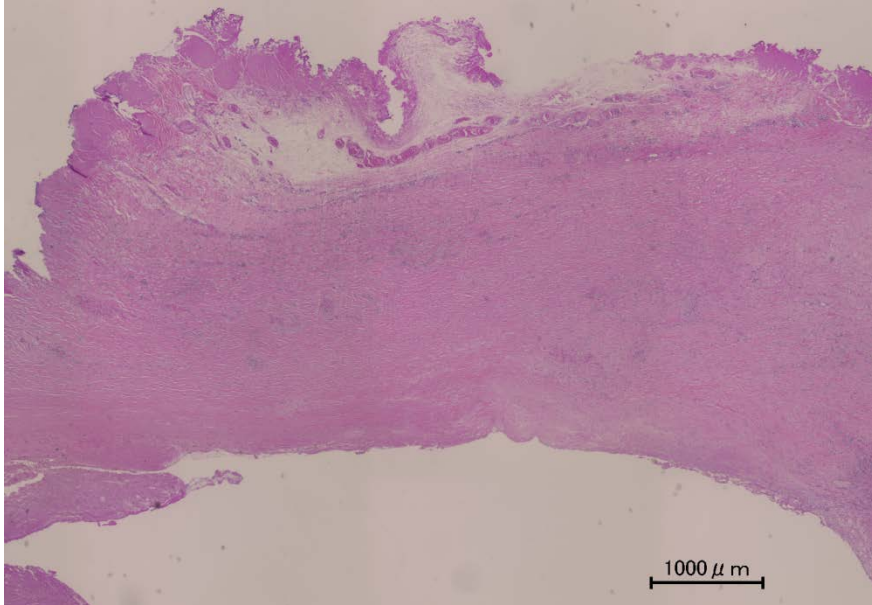
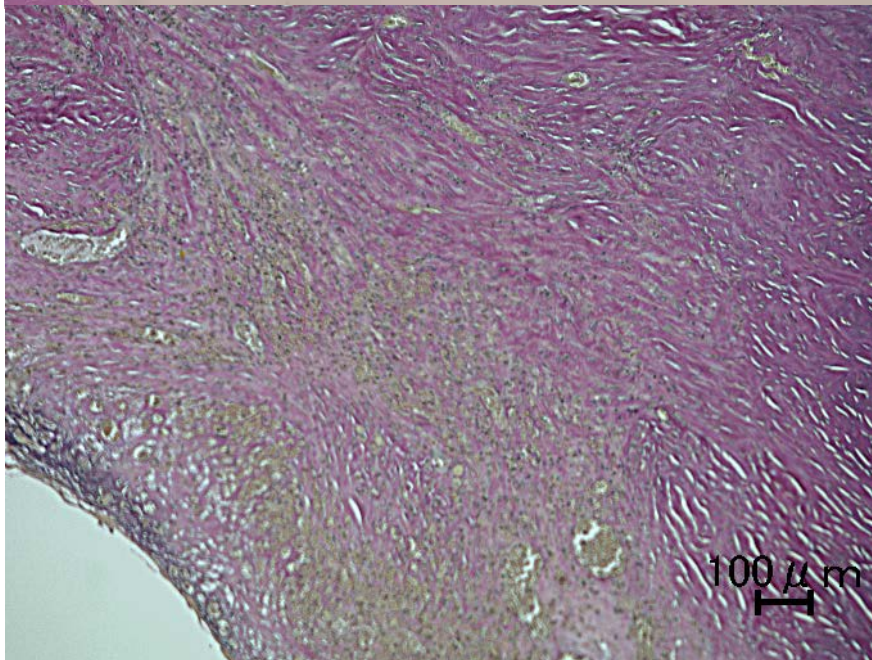
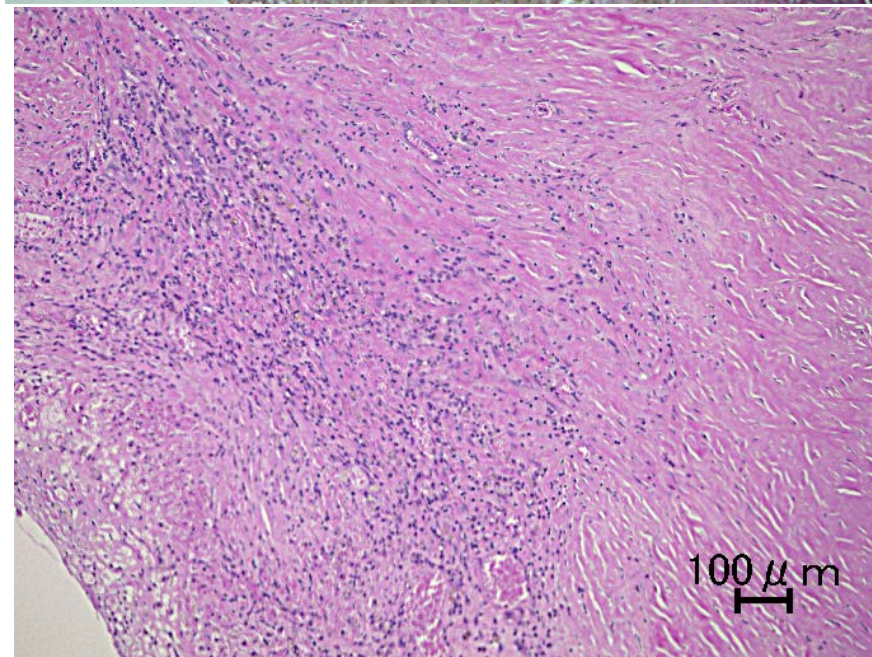


Fig. 2

A**B****C****Fig. 3**