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Case Report

Chest Pain in A Patient with Polyarteritis Nodosa and Behcet's Disease: Myocardial Infarction or Aortic Dissection?

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ABSTRACT

Aortic diseases have high mortality and are usually late or misdiagnosed. Especially in patients with inflammatory vasculitis, diagnosis is often confused with other causes of chest pain and this causes a delay in diagnosis. Vascular complications are the most important predictors of mortality and morbidity in Behcet's disease and also polyarteritis nodosa. The diagnosis of aortitis is usually obtained by vascular imaging, but partly made only by biopsy on occasion of an operation. Here we present a case of pathologically proven fatal aortitis and aortic dissection in a patient with Polyarteritis nodosa and Behcet's disease.

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Introduction

Vasculitis develops reactive damage that causes inflammation in the vascular wall. They cause bleeding and ischemia as a result of damage to the vascular lumen and impaired vascular permeability.

They are usually classified according to the size, type, and location of the affected vessel. Although it involves specifically according to the vessel size, there are also forms with a wide variety of vascular involvement. Organ structures affected are also important in the diagnosis of vasculitis. Vasculitis is serious diseases that can sometimes be fatal. Therefore, early diagnosis and treatment are crucial. Aortic dissection is one of the most fatal cases of vasculitis. Aortic dissection associated with polyarteritis nodosa has been reported in the literature only in a few cases, but cases of aortic dissection associated with Behçet's disease have been reported.

It is difficult to diagnose because of diseases that mimic vasculitis. Systemic lupus erythematous, atherosclerotic diseases and drug reactions can be given as examples.

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Case Report

A 44-year–old man was admitted to our hospital with chest pain that started 2 hours before admission. His medical history had celiac disease, hypertension, Behcet's disease, Polyarteritis nodosa, hepatitis B. His medications were 40 mg prednisolone once a day and azathioprine 50 mg three times in a day. ECG was normal and the cardiac troponin value at the time of admission was 38.3 ng/mL (0-17.5 ng/mL), while it was 110.6 ng/mL after 6 hours of admission. Transthoracic echocardiography demonstrated with mild hypokinesis of the anterior wall and an ejection fraction was around 50%. In accordance with these results, the patient was diagnosed with acute non-ST-segment elevation myocardial infarction (NSTEMI) and decided to perform a coronary angiography.

As a result of coronary angiography slow flow was observed in the LAD and several attempts to identify the right coronary artery (RCA) ostium failed. Injection of contrast medium didn't show any coronary artery originating from right coronary cusp. An aortogram revealed an ascending aortic dissection flap and real lumen early opacification and then the false lumen opacified. (Figure 1A, 1B) The patient was transferred to

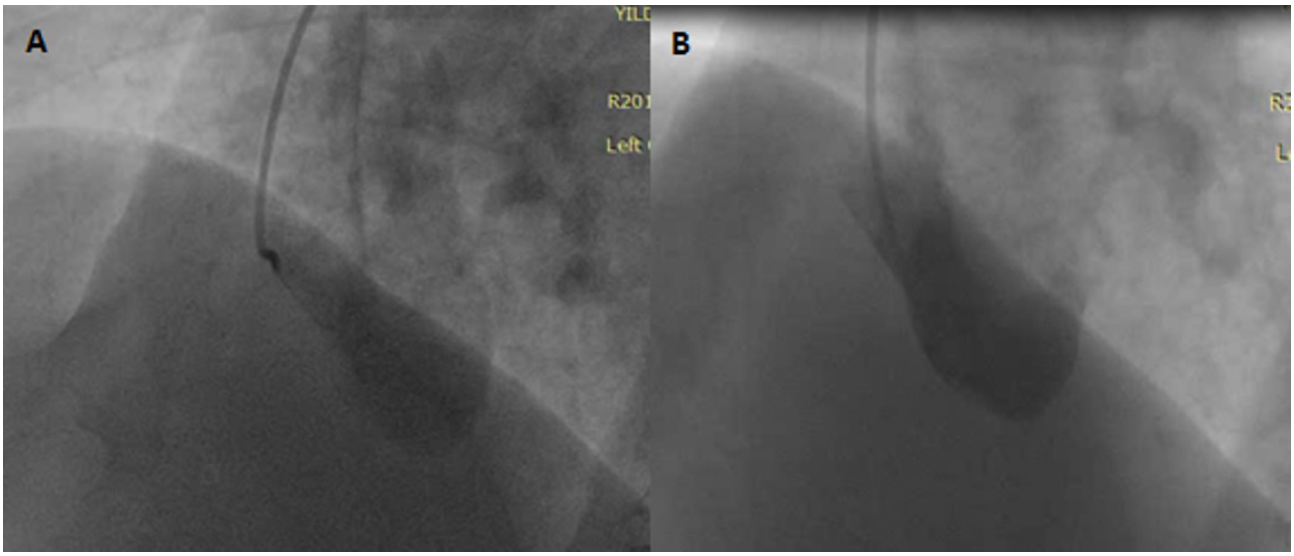

Figure 1A. Aortic dissection flap is seen in aortography with right Judkins catheter; **Figure 1B.** Early opacification of the real lumen is observed.

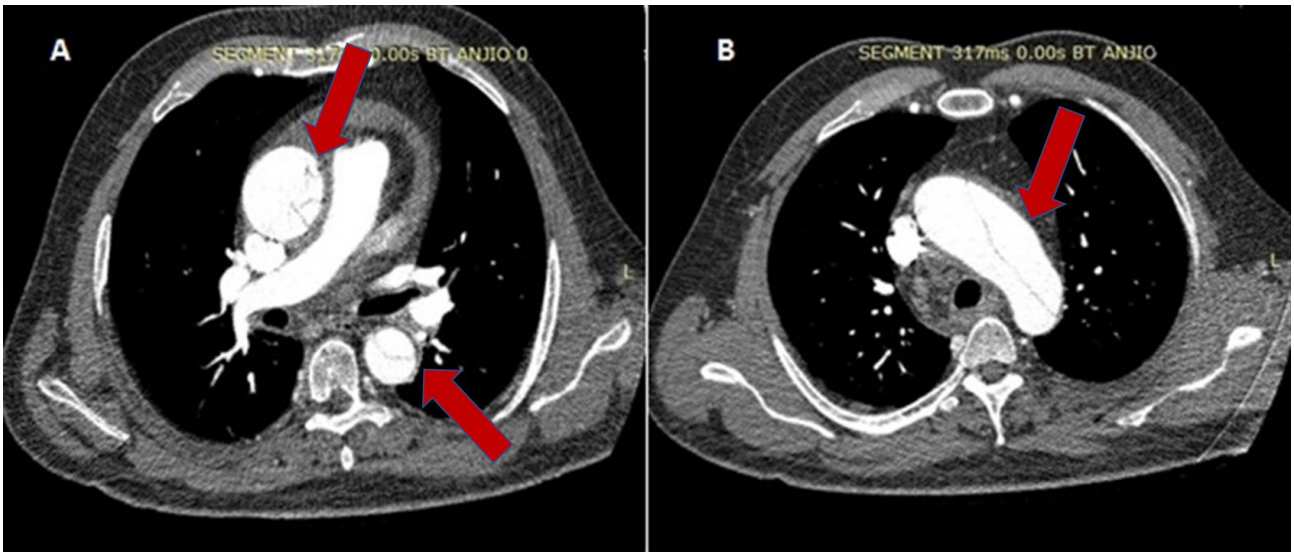

Figure 2A and **Figure 2B.** Aortic dissection images with multi-slice computerized tomographic angiography.

an intensive care unit and urgent cardiovascular surgeon consultation was requested. The infusions of esmolol and nitroprusside were up-titrated and the so that the patient's blood pressures could be closely monitored. Hydromorphone and fentanyl were initiated for a pain control.

An emergent contrast-enhanced computed tomography angiography scan was obtained and revealed aortic dissection of Stanford A and DeBakey type 1. (Figure 2A, 2B) The origin of the intimal tear was detected just distal to the coronary sinus. The dissection reached the abdominal aorta and pervaded both external iliac arteries. The celiac trunk, superior mesenteric artery and right renal artery originated from the true lumen. The left renal artery and inferior mesenteric artery originated from the false lumen.

The patient was transferred for cardiac surgery. The surgical repair consisted of graft replacement of the ascending aorta with the reconstruction of the left brachiocephalic and left common carotid artery. Despite the successful implementation of the procedure bleeding control was not achieved and the patient died. Histopathological examination of the resected specimen was revealed cystic medial degeneration and dissection at the medial layer, active and chronic inflammation and vasculitic process in the adventitia. (Figure 3A, 3B)

Discussion

Behcet's Disease is a chronic multisystemic immune-mediated disorder characterized by recurrent oral and/or genital ulcers, arthritis, skin manifestations, and ocular, vascular, neurological, or intestinal involvements. Vascular involvement may be seen in 25–50% of Behcet's disease patients.[1] Four types of vascular lesions were described: arterial occlusion, aneurysms, venous occlusion, and thrombophlebitis or thrombosis. Venous manifestations are more common than arterial involvement (88% vs. 12%).[1] Arterial lesions are frequently localized in the aorta, cerebral, carotid, subclavian, brachial, ulnar, renal, and popliteal arteries.[2] Sporadic cases of endocarditis, myocarditis, pericarditis, acute myocardial infarction, aortic aneurysm, ventricular thrombosis, congestive cardiomyopathy, and valvular dysfunction have been reported.[3,4] In patients with Behcet's disease, the vasculitis may damage the aortic wall and may predispose to dissection and development of aneurysms.

Polyarteritis Nodosa is characterized by a necrotizing inflammation of the entire vascular wall which develops in a segmental pattern. Fibrinoid necrosis is frequently observed in active lesions, and neutrophils are more frequently present in vessels with fibrinoid necrosis.[5] Approximately, 20% of polyarteritis nodosa may be associated with chronic viral hepatitis caused by the Hepatitis B virus infection.[6] Cardiac involvement is stated to occur in up to 35% of patients with polyarteritis nodosa and carries prognostic significance.[6]

Polyarteritis nodosa is a vasculitis of medium-sized and small vessels, small arteries may be involved, but small vessels, including arterioles, capillaries, and venules, are not, and therefore involvement of the ascending aorta is not expected.[6] The most common cardiac involvement of is myocardial infarction due to coronary artery involvement.[7,8] Lino T described a 59-year-old woman with polyarteritis nodosa with hepatitis

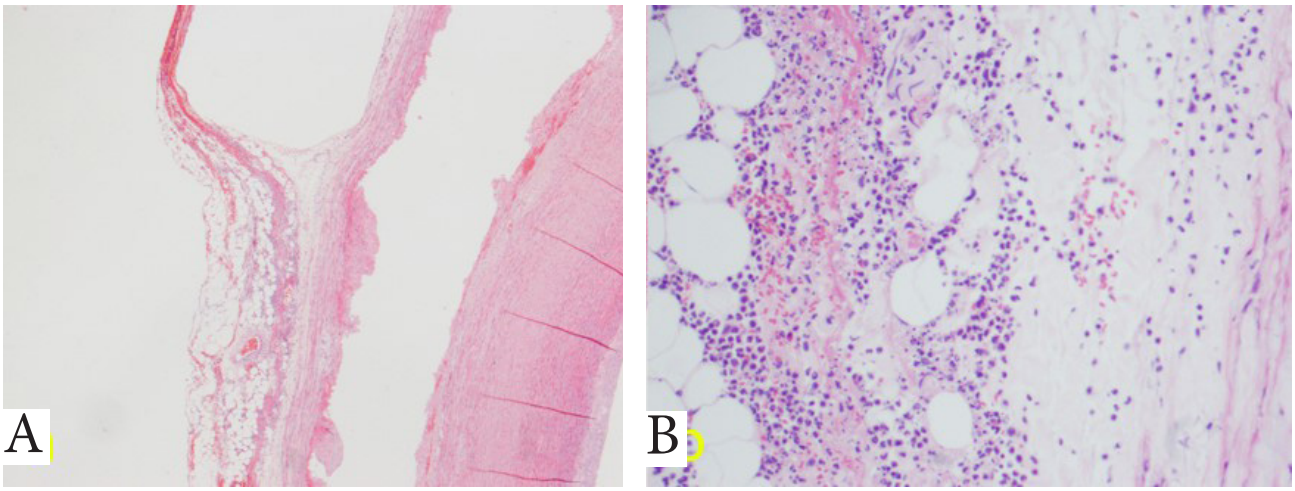

Figure 3A and **Figure 3B**

B admitted acute onset of chest pain. After developing the acute attack of severe chest pain, she died suddenly. At autopsy, a DeBakey type 1 aortic dissection was found and the immediate cause of death was found to be cardiac tamponade secondary to rupture of the aortic dissection.[9]

In these types of inflammatory diseases, when complications such as aortic dissection and aneurysm develop, mortality is high. Therefore, we should be careful about such complications that may be lethal in this patient group and we should follow up the symptoms of patients like chest pain and shortness of breath. When the diagnosis of aortitis is suspected on the basis of clinical presentation, expedient imaging of the entire aorta with an appropriate modality is critical to establish the diagnosis. Modern imaging tools for the aorta include computed tomography angiography (CT), magnetic resonance angiography (MRA), and ultrasonography. Positron emission tomography (PET) scanning has emerged for targeted imaging of vascular inflammation and may be particularly useful when combined with traditional cross-sectional imaging modalities.[10]

An immunosuppressive therapy is the primary treatment of non-infectious aortitis due to vasculitis, and patients are ideally managed by a multi-disciplinary team that includes a rheumatologist and medical and surgical cardiovascular specialists.[11]

In our patient's pathological examination; there was no fibrinoid necrosis on vascular involvement. So we think that the vascular inflammatory disease causing the aortic dissection of the patient is Behcet's disease. However, overlap syndromes and disease interactions should be considered in patients with diffuse vasculitic involvement in which multiple inflammatory diseases coexist as well as in our patients.

Conclusion

In conclusion, vascular complications are the most important predictors of mortality and morbidity in Behcet's disease and also polyarteritis nodosa. Early detection of vasculitis and the need for aggressive treatment are essential for the optimal care of these patients. At the same time, aortic dissection should be kept in mind when these patients are referred to with chest pain.

Conflict of Interests

Authors declare that there are none.

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