# Association of aortic and main left coronary aneurysms with severe aortic insufficiency in Takayasu's arteritis

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## Abstract

Takayasu's arteritis is a granulomatous vasculitis of unknown etiology that affects mainly the aorta and its branches. As a result of intimal fibroproliferation, segmental stenosis, occlusion, dilatation, and aneurysmal formation of the involved vessels may develop. It is an uncommon disease and usually affects young Asian female patients during the second and third decades of life. Coronary arteries are exceptionally affected and coronary aneurysm formation is a very rare finding. We describe a case of a previously healthy 26-year-old Caucasian female whose Takayasu's arteritis presented as a previously undescribed association of aortic and main left coronary aneurysms with severe aortic insufficiency.

## Introduction

Takayasu's arteritis is a granulomatous vasculitis of unknown etiology that affects mainly the aorta and its branches.[1] As a result of intimal fibroproliferation, segmental stenosis, occlusion, dilatation, and aneurysmal formation of the involved vessels may develop.[1] It is an uncommon disease, with an approximate incidence of 2-3 cases per year per million individuals and usually affects young female of Asian ascendance during their second and third decades of life.[2] Coronary arteries are exceptionally affected and coronary aneurysm formation is rare.[3] We describe a case of a previously healthy Caucasian female whose Takayasu's arteritis presented as an association of aortic and main left coronary aneurysms with severe aortic insufficiency.

## Case Report

A 26-year-old Caucasian female was admitted to our hospital with a 3-week history of fatigue, malaise, exertion dyspnea, ortophnea and paroxysmal nocturnal dyspnea. There was a marked difference on the blood pressure measurement between the arms (140/90 mmHg on the right arm and 90/60 mmHg on the left). The brachial pulse could not be felt on the left arm and a systolic murmur was heard on the left infraclavicular area. On cardiac auscultation, a diastolic murmur (++++/VI) was heard on the aortic area, and there were some crackles in the basal regions of both lungs. Her erythrocyte sedimentation rate (ESR) was 64 mm on the first hour and the result of the serum C-reactive protein (CRP) was 24 mg/dL (normal: 0-6 mg/dL). Based on the 1990 *American College of Rheumatology* criteria,[4] a diagnosis of Takayasu's arteritis was made. A high-resolution thorax computed tomography (CT) showed a 4-cm aortic aneurysm spanning the ascending and the proximal descending portions, as well as the aortic arch. Cineangiocoronariography confirmed the findings of the CT and also revealed a severe aortic insufficiency and a large main left coronary aneurysm (Figure 1). A three-day course of intravenous high-dose methylprednisolone (1000 mg) was administered, as well as medications for the management of the heart failure (diuretics, digoxin, angiotensin-converting enzyme inhibitor), which resulted in a remarkable improvement in general symptoms. Methotrexate was started at 15 mg/week as a steroid-sparing medication.

## Discussion

Takayasu's arteritis is primarily a chronic inflammatory vasculitis characterized by stenosis of large and medium sized arteries.[1] The coronary arteries are involved in about 10% of cases, but aneurysm formation, especially affecting the main left coronary, is a very rare finding.[3] Destruction of the elastic fibers in the media of the vessel is the leading pathogenic mechanism of aneurysms formation. In some situations of massive aortic regurgitation, a modified Bentall operation has been proposed.[5] Coronary aneurysms predispose to thrombus formation and acute myocardial infarction, even in patients receiving aspirin and/or warfarin.[6] On the other hand, revascularization under inflammatory circumstances carries a higher risk of complications, such as stenosis, suture line dehiscence and pseudoaneurysm formation.[7] A case of successful surgical resection of a giant right coronary artery has been reported,[8] even though the best surgical timing in Takayasu's arteritis still remains controversial, as long as even when asymptomatic and with no serological evidence of current inflammation (normal ESR), up to 44% of these patients show some degree of histologic active disease.[9]

To our knowledge, this is the first report of an association of aortic and coronary aneurysms with severe aortic insufficiency in a Takayas's arteritis patient. The complexity of this case, allied to the absence of previously described medical interventions specific to this situation, certainly turns it into a therapeutic challenge. Proper follow-up is warranted, to get nearer to an accurate decision about the best moment to choose for a surgical approach, considering its costs and benefits when dealing with vasculitic vessels and its inner complications.

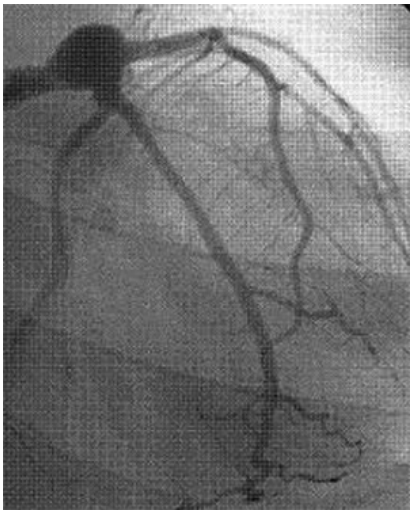

**Figure 1. Angiogram showing a large aneurysm in the main left coronary.**

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## References

1. Johnston SL, Lock JR, Gompels MM. Takayasu arteritis: a review. J Clin Pathol 2002;55:481-6.
2. Watts RA, Scott DG. Classification and epidemiolgy of the vasculitides. Baillieres Clin Rheumatol 1997;11:191-217.
3. Suzuki H, Daida H, Tanaka M, et al. Giant aneurysm of the left main coronary artery in Takayasu aortitis. Heart 1999;81:214-7.
4. Arend WP, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990;33:1129-34.
5. Inoue T, Matsumoto T, Saga T. Massive progression of annuloaortic ectasia in a patient with Takayasu aortitis. Can J Cardiol 2007;23:993-4.
6. Sugahara Y, Ishii M, Muta H, et al. Warfarin therapy for giant aneurysm prevents myocardial infarction in Kawasaki disease. Pediatr Cardiol 2008;29:398-401.
7. Amano J, Suzuki A, Tanaka H, Sunamori M. Surgical treatment for annuloaortic ectasia in Takayasu arteritis. Int J Cardiol 1998;66 Suppl 1:S197-202; discussion S203-4.
8. Kanaan S, Baker C, Starnes V. Resection of giant coronary artery aneurism in a Takayasu's arteritis patient. Ann Thorac Surg 2008;85:1795-6.
9. Kerr GS, Hallahan CW, Giordano J, et al. Takayasu arteritis. Ann Intern Med 1994; 120:919-29.