ESC European Society of Cardiology

European Heart Journal - Case Reports (2018) **2**, 1–5
doi:10.1093/ehjcr/yty091

CASE REPORT

Cardiogenic shock as the first manifestation of large vessel vasculitis in a young patient: case report

Nicolas Isaza[1]*, Adriana Maria Posada[1], Mauricio Eduard Diaz[2,3], and Daniel Isaza-Restrepo[3]

[1]Faculty of Medicine, Pontificia Universidad Javeriana, Carrera 7 No. 40 - 62, Bogotá, Colombia; [2]Faculty of Medicine, Universidad del Bosque, Carrera 9 No. 131A - 02, Bogotá, Colombia; and [3]Department of Cardiology, Fundacion Cardioinfantil Instituto de Cardiologia, Calle 163a No. 13b - 60, Bogotá, Colombia

Received 5 February 2018; accepted 13 July 2018; online publish-ahead-of-print 3 August 2018

Background Cardiogenic shock secondary to coronary involvement in large vessel vasculitis (LVV) is an unsuspected finding, even more, when no other vascular territories are compromised and when it constitutes the initial clinical manifestation. This case report illustrates a case in which a complete diagnostic study uncovered this aetiology.

Case summary A 33-year-old woman with cough and chest pain who was diagnosed with acute bronchitis returned with worsening dyspnoea, chest pain, and developed cardiogenic shock. The initial differential diagnoses included myocarditis and takotsubo cardiomyopathy (TCM) owing to a positive troponin I, and echocardiogram with left ventricular dilation, dyskinesia in mid-ventricular and apical segments, systolic dysfunction, and functional mitral regurgitation. A cardiac magnetic resonance showed contractility abnormalities resembling the pattern of TCM but lacked the characteristic myocardial oedema. Subsequently, a coronary angiography expected to result without obstructions showed a critical narrowing of the left main coronary artery. Surgical management consisted of a pericardium patch grafted in the stenotic ostium to restore adequate perfusion. The surgical specimens were sent to the pathology laboratory that reported findings compatible with LVV. Four days after the surgical intervention the patient was discharged alive with a complete recovery of left ventricular systolic function.

Discussion Chest pain symptoms in a young woman, could be caused by multiple entities, and an ischaemic aetiology from a non-atherosclerotic origin should be kept in mind. A complete study with coronary angiography is crucial to rule out an ischaemic cause even in low-risk groups for atherosclerotic coronary heart disease.

Keywords Cardiogenic shock • Large vessel vasculitis • Ischaemic heart disease • Acute coronary syndrome • Case report

Learning points

- Takayasu arteritis (TA) has been found to be a more common aetiology for ischaemic heart disease in young patients than other more frequently suspected diagnosis and cardiovascular death secondary to coronary involvement in patients with TA is not rare.
- Suspicion and stepwise work up allow its diagnosis and proper management.

* Corresponding author. Tel: (1) 484-821-6241, Fax: (571) 6680221, Email: nicolasisaza93@hotmail.com. This case report was reviewed by Ola Jan Magnus Vedin, Nikolaos Bonaros, Domenico D'Amario, Mark Philip Cassar, and Christian Fielder Camm.

Introduction

Large vessel vasculitis (LVV) includes multiple conditions causing inflammation of large arteries, especially the aortic arch and its branches. The two characteristic entities are giant cell arteritis and Takayasu arteritis (TA).[1–3] Coronary involvement in LVV can occur in a quarter of patients, and the transmural compromise of the ostia and proximal segments is the most common alteration.[2,4–7] Although ischaemic cardiomyopathy is the most common cause of death in the presence of coronary involvement of LVV, acute myocardial infarction is also observed.[1,8] However, ischaemic causes of cardiogenic shock tend to be overlooked in younger patients due to the lower incidence of myocardial infarction.[9]

This report illustrates the case of a young woman that developed cardiogenic shock secondary to coronary involvement of TA. It demonstrates how a thorough approach can help elucidate uncommon causes for cardiogenic shock in young patients.

Timeline

Day	Event
Day 1	Patient consulted the first time the Emergency Department (ED) for cough and chest pain and was discharged with a diagnosis of acute bronchitis
Day 4	Patient consulted the second time to the ED and developed symptoms of cardiogenic shock. Laboratory tests, echocardiography, and chest X-ray were obtained. Transferred to the Coronary Care Unit
Day 5	Cardiac magnetic resonance was performed—differential diagnoses of acute myocarditis and takotsubo cardiomyopathy
Day 6 (AM)	Coronary angiography to confirm the diagnosis showed a critical lesion on the left main coronary artery
Day 6 (PM)	Surgical pericardial patch graft to restore flow in the left coronary artery
Day 10	Repeat echocardiography was performed to assess left ventricular function

Case summary

A 33-year-old woman presented to the emergency department with 1 month of asthenia, non-productive bouts of cough, and associated chest pain. She was discharged with no further work up and a diagnosis of acute bronchitis. Three days later she returned with worsening stabbing, non-radiated chest pain described as pleuritic, however, exacerbated on exertion and improved at rest, and associated to exertional and paroxysmal nocturnal dyspnoea.

Physical examination revealed blood pressure of 112/64 mmHg, heart rate of 116 b.p.m., respiratory rate of 19 breaths/min, oxygen saturation of 98% on room air, no murmurs, fine bilateral crackles in the lung bases, no abdominal abnormalities, and no lower extremity oedema.

A D-dimer and chest X-ray were normal but the electrocardiogram showed 2-mm ST depression in precordial leads, and a positive delta troponin I-hs increasing from 0.06 to 2.25 ng/mL (0–0.026 ng/mL). Other laboratory results such as C-reactive protein and the erythrocyte sedimentation rate were within normal limits. Transthoracic echocardiography revealed a severely dilated left ventricle (LV) with mid-ventricular, apical dyskinesia, severe compromise of the systolic function, left ventricular ejection fraction (LVEF) of 20%, diastolic dysfunction, severe functional mitral regurgitation, moderate functional tricuspid regurgitation, and an estimated pulmonary systolic arterial pressure of at least 80 mmHg (*Figure 1*, Supplementary material online, *Video S1*).

On admission to the Coronary Care Unit, blood pressure was 86/54 mmHg and heart rate was 124 b.p.m., findings consistent with cardiogenic shock. IV norepinephrine was administered for 8 h and then successfully weaned off. The initial work up diagnoses were takotsubo cardiomyopathy (TCM) or acute myocarditis. Therefore, optimal medical therapy was initiated with Enalapril 2.5 mg b.i.d., Spironolactone 25 mg daily (QD), Carvedilol 3.125 mg b.i.d., IV Furosemide 5 mg, and low molecular weight heparin (LMWH). Despite the positive delta troponin, no antiplatelet therapy was initiated due to the non-coronary distribution of the wall motion abnormalities found on echocardiography that led to suspicion of the aforementioned differential diagnoses.

The next day a cardiac magnetic resonance (cMRI) revealed mild LV dilatation, and LVEF of 45%, severe segmental contractility abnormalities in the apical and mid-ventricular walls, with normal basal contractility, which resembled the pattern seen in TCM (*Figure 2*, Supplementary material online, *Video S2*). However, no myocardial oedema, fibrosis, or scarring was observed.

Angiography had not been performed because the troponin elevation was considered to be a leak from the myocarditis, TCM, or heart failure. However, it became necessary to fulfil the diagnostic criteria for TCM and due to the inconclusive results of the cMRI. Results revealed a 99% ostial obstruction in the left main coronary artery with no additional findings (*Figure 3*).

After the coronary angiogram, the patient was emergently transferred to the operating room were severe left coronary ostium stenosis without atheroma plaques was encountered. A pericardium patch was used to widen the ostium of the left main coronary artery. Although arterial grafts were a possible treatment approach, the patch was chosen because of the high long-term risk of occlusion and narrowing with venous or arterial grafts due to the patients' young age. Additionally, a review of the literature suggested the possibility of further inflammation and stenosis at the proximal anastomotic sites of grafts, in the brachiocephalic, subclavian, or internal mammary arteries.[10–12] Surgical specimens were analysed by the pathology laboratory and reported vascular wall architecture distorted by fibrosis, lymphoplasmacytic inflammatory infiltrates, and obliterative vasculopathy, compatible with LVV (*Figure 4*). These findings along with the type of vessels involved and the clinical characteristics of the patient led to a diagnosis of TA.

After 4 days in the hospital wards receiving prednisone 50 mg QD as primary treatment for TA, and Carvedilol 6.25 mg b.i.d., Enalapril 5 mg b.i.d., Spironolactone 25 mg QD, and dual antiplatelet therapy, she was discharged with complete LV function recovery, and no valvular alterations as shown by transthoracic echocardiography.

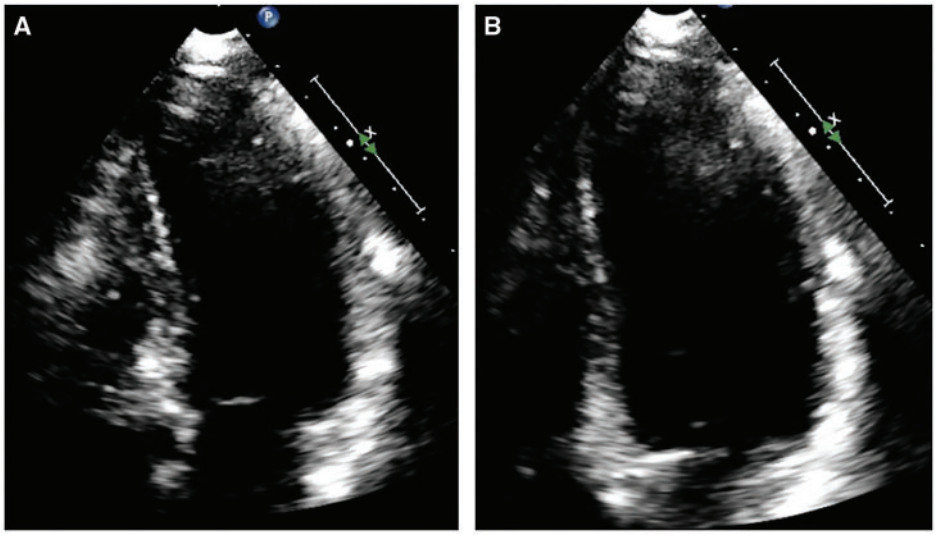

Figure 1 Transthoracic echocardiography: (*A*) apical two-chamber during diastole and (*B*) apical two-chamber during systole.

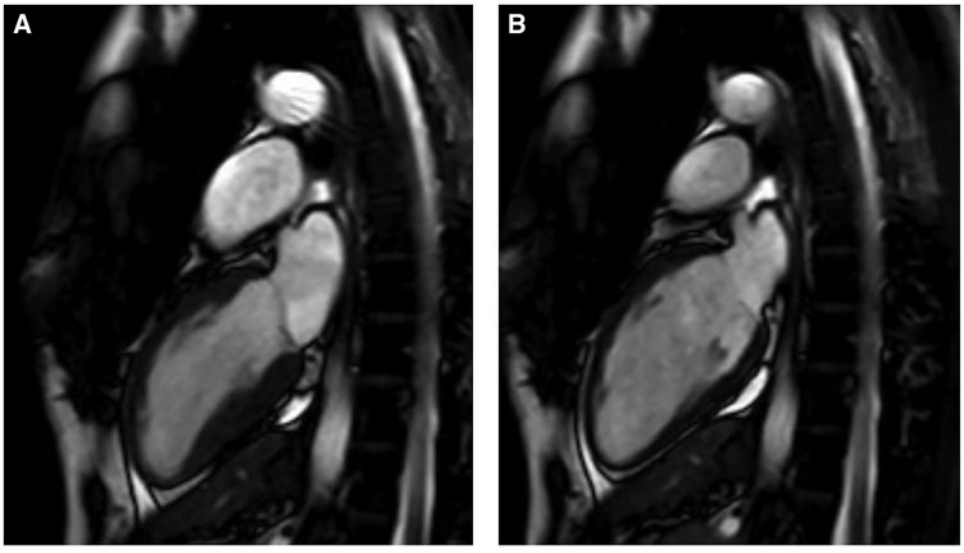

Figure 2 Cardiac magnetic resonance: (*A*) two-chamber during diastole and (*B*) two-chamber during systole.

It was a notable recovery thought to be secondary to ischaemia resolution in the setting of myocardial stunning and lack of scar tissue. Follow-up 2 weeks later with neck, chest, and abdominal computed tomography angiography failed to reveal further vessel compromise, and she denied having chest pain and an improved functional class.

Discussion

In TA, the aortic branches are usually compromised in a transmural manner with subsequent stenosis, or obstruction due to thrombus formation, or aneurysmal thrombosis or rupture.[8] Symptoms relate to end organ hypo-perfusion, hypertension secondary to renal artery stenosis, pulselessness in upper extremities, and neurological symptoms such as postural dizziness and seizures are common.[3,8] Even though the development of cardiogenic shock follows the same principle of end organ hypo-perfusion, its diagnosis is challenging as clinicians are not frequently exposed to it. Additionally, the clinical diagnosis of TA requires further manifestations of symptomatic ischaemia in other vessel groups apart from the arteriographic evidence of narrowing of the aorta or its primary branches.[13] Therefore, in a patient with few symptoms, the diagnosis is

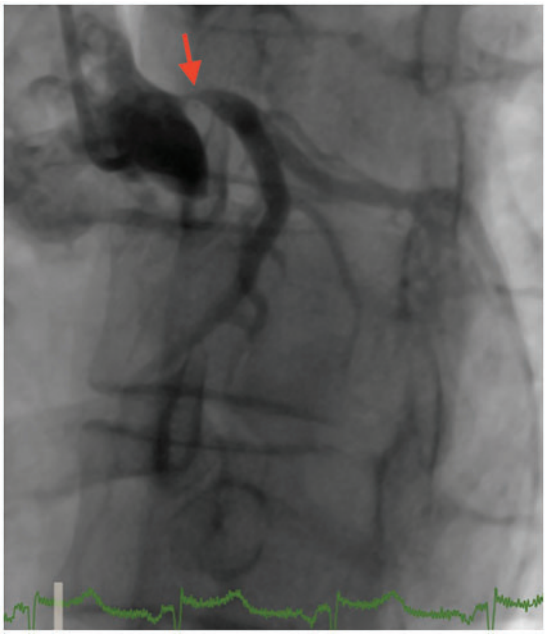

Figure 3 Coronary angiography: right anterior oblique caudal view of left main coronary artery. Red arrow showing critical lesion in the proximal left main coronary artery.

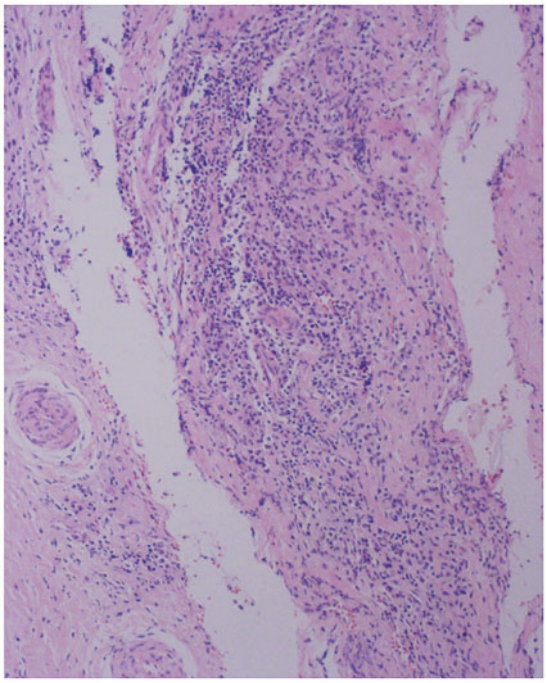

Figure 4 Pathology specimen: vascular wall architecture distorted by fibrosis, lymphoplasmacytic inflammatory infiltrates, and obliterative vasculopahty, with no granulomas, compatible with large vessel vasculitis.

furthermore complicated as it would require pathologic corroboration of the inflammatory vessel compromise.

On the other hand, it is easy to mistakenly minimize symptoms of chest pain in young patients and discharge them without further evaluation. In this case, an ischaemic cause was not initially suspected, and work up was more compatible with myocarditis or TCM.

Nonetheless, a study of 40 young female patients presenting with ischaemic heart disease, revealed that 10% were diagnosed with TA.[14] This proportion was higher than that of other diagnoses such as vasospastic angina, drug abuse, aortic dissection, and TCM. This finding brings attention to TA as a differential diagnosis of ischaemic heart disease, that is easily overlooked as the other aetiologies are thought to be more common.

Furthermore, a series in China revealed that 7.7% of the patients with TA had coronary involvement, concluding that cardiovascular death secondary to coronary involvement is not rare.[15] Taking this into account, clinicians should keep TA as a possible differential diagnosis in young women presenting with chest pain. The approach to these patients requires a stepwise work up with different imaging modalities, coronary, and aortic angiography. Following these steps allowed the team to diagnose a non-suspected aetiology of a severe left main coronary artery lesion in a young patient.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

References

1. Gulati A, Bagga A. Large vessel vasculitis. *Pediatr Nephrol* 2010;**25**:1037–1048.
2. Subramanyan R, Joy J, Balakrishnan KG. Natural history of aortoarteritis (Takayasu's disease). *Circulation* 1989;**80**:429–437.
3. Weyand CM, Goronzy JJ. Medium- and large-vessel vasculitis. *N Engl J Med* 2003;**349**:160–169.
4. Endo M, Tomizawa Y, Nishida H, Aomi S, Nakazawa M, Tsurumi Y, Kawana M, Kasanuki H. Angiographic findings and surgical treatments of coronary artery involvement in Takayasu arteritis. *J Thorac Cardiovasc Surg* 2003;**125**:570–577.
5. Rav-Acha M, Plot L, Peled N, Amital H. Coronary involvement in Takayasu's arteritis. *Autoimmun Rev* 2007;**6**:566–571.
6. Amano J, Suzuki A. Coronary artery involvement in Takayasu's arteritis. Collective review and guideline for surgical treatment. *J Thorac Cardiovasc Surg* 1991;**102**:554–560.
7. Numano F. Differences in clinical presentation and outcome in different countries for Takayasu's arteritis. *Curr Opin Rheumatol* 1997;**9**:12–15.
8. Numano F, Kobayashi Y. Takayasu arteritis—beyond pulselessness. *Intern Med* 1999;**38**:226–232.
9. van Diepen S, Katz JN, Albert NM, Henry TD, Jacobs AK, Kapur NK, Kilic A, Menon V, Ohman EM, Sweitzer NK, Thiele H, Washam JB, Cohen MG. Contemporary management of cardiogenic shock: a scientific statement from the American Heart Association. *Circulation* 2017;**136**:e232–e268.
10. Murashita T, Yoshimoto K, Sugiki H, Yasuda K. Bilateral coronary ostial patch angioplasty with autologous pericardium in Takayasu arteritis: a case requiring replacement of the aortic valve and ascending aorta. *Eur J Cardiothorac Surg* 2004;**26**:866–868.

11. Kumar Venkata R, Agarwal NB, Javali S, Patwardhan AM. Takayasu's arteritis with ostial and left main coronary artery stenosis. *Tex Heart Inst J* 2007;**34**: 470–474.
12. Kawase I, Ozaki S, Yamashita H, Uchida S, Nozawa Y, Matsuyama T, Takatoh M, Hagiwara S. Angioplasty with autologous pericardium for bilateral coronary ostial stenosis in Takayasu disease. *Ann Thorac Surg* 2012;**93**:645–647.
13. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, Fauci AS, Leavitt RY, Lie JT, Lightfoot RW, Masi AT, McShane DJ, Mills JA, Stevens MB, Wallace SL, Zvaifler NJ. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. *Arthritis Rheum 1990* 2010;**33**:1129–1134.
14. Cavalli G, Tomelleri A, Di Napoli D, Baldissera E, Dagna L. Prevalence of Takayasu arteritis in young women with acute ischemic heart disease. *Int J Cardiol* 2018;**252**:21–23.
15. Sun T, Zhang H, Ma W, Yang L, Jiang X, Wu H, Hui R, Zheng D. Coronary artery involvement in takayasu arteritis in 45 Chinese patients. *J Rheumatol* 2013;**40**: 493–497.