

# Juvenile Takayasu's arteritis presenting as acute coronary syndrome in a 10 year-old girl

Młodzieńcze zapalenie tętnic Takayashu objawiające się jako ostry zespół wieńcowy u 10-letniej dziewczynki

Santosh Kumar Sinha, Dibbendhu Khanra, Lokendra Rekwai, Puneet Aggarwal,  
Chandra Mohan Varma, Vinay Krishna

Department of Cardiology, LPS Institute of Cardiology, G.S.V.M. Medical College, Kanpur, India

## Abstract

A 10 year-old girl was admitted to hospital for evaluation of sudden onset chest pain of three days' duration. There were no co-morbidities or family history of coronary artery disease. There was a disparity of blood pressure in the upper limbs, while pulses in the lower limbs were barely palpable. Her electrocardiogram revealed ST segment elevation in II, III, and aVF leads. Laboratory tests showed elevated troponin T (0.8 ng/mL), total leukocyte count (13,800/ $\mu$ L), C-reactive protein (94 mg/L), and erythrocyte sedimentation rate (56 mm in first hr). Echocardiography showed mildly impaired left ventricular systolic function with an ejection fraction of 58%. Aortogram showed total occlusion of the brachiocephalic trunk, diffuse thoraco-abdominal coarctation, and osteo-proximal stenosis (80%) of the left renal artery. Coronary angiography revealed total occlusion of the left circumflex artery and tubular lesion of the proximal left anterior descending artery with 70% stenosis. Based on these findings, she was diagnosed to be in the active phase of juvenile Takayasu's arteritis having acute inferior wall myocardial infarction. Her treadmill test for reversible ischaemia (modified Bruce protocol) was negative, and she was discharged on medication (antiplatelets, corticosteroids, and immunosuppressive agents).

Key words: juvenile Takayasu's arteritis, acute coronary syndrome, diffuse coarctation

Folia Cardiologica 2019; 14, 1: 79–82

## Introduction

Takayasu's arteritis (TA) is a chronic systemic idiopathic inflammatory large vessel vasculitis that usually afflicts the aorta and its main branches, and pulmonary arteries. It leads to stenosis, dilatation, or aneurysm formation of affected vessels [1]. Coronary artery involvement as an initial presentation is rare, though its involvement is seen in 22% of cases which occur in the later stage.

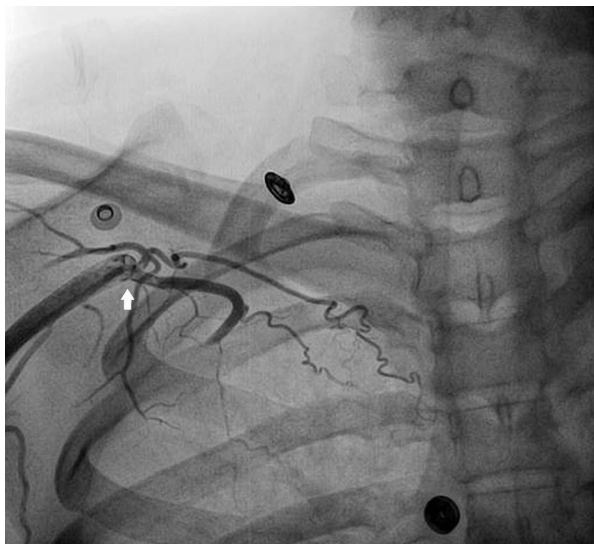
Conventionally, C-reactive protein (CRP), and erythrocyte sedimentation rate (ESR) are the markers of disease activity. The disease can be monitored with 18-fluorodeoxyglucose

positron emission tomography ( $^{18}$ FDG-PET), magnetic resonance imaging (MRI), computed tomography (CT), and pentraxin level monitoring. However, the best therapeutic approach in cases of acute coronary syndromes (ACS) has not been well established because the results of percutaneous transluminal coronary angioplasty (PTCA) and/or coronary artery bypass grafting (CABG) are variable.

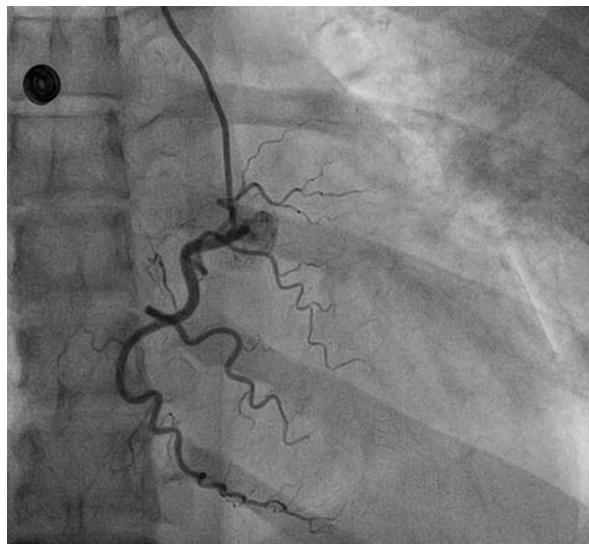
## Case report

A 10 year-old girl was admitted to hospital for evaluation of sudden onset chest pain of three days' duration. There

Address for correspondence: Santosh Kumar Sinha MD, FAESC, Asst. Professor, Department of Cardiology, LPS Institute of Cardiology, G.S.V.M. Medical College, G.T. Road, Kanpur, Uttar Pradesh 208002, India, fax +91 0512 255 61 99/255 65 21, e-mail: fionasan@rediffmail.com



**Figure 1.** Coronary angiogram through right transradial route showing total occlusion of brachiocephalic trunk



**Figure 2.** Coronary angiogram through left transradial route showing normal right coronary artery

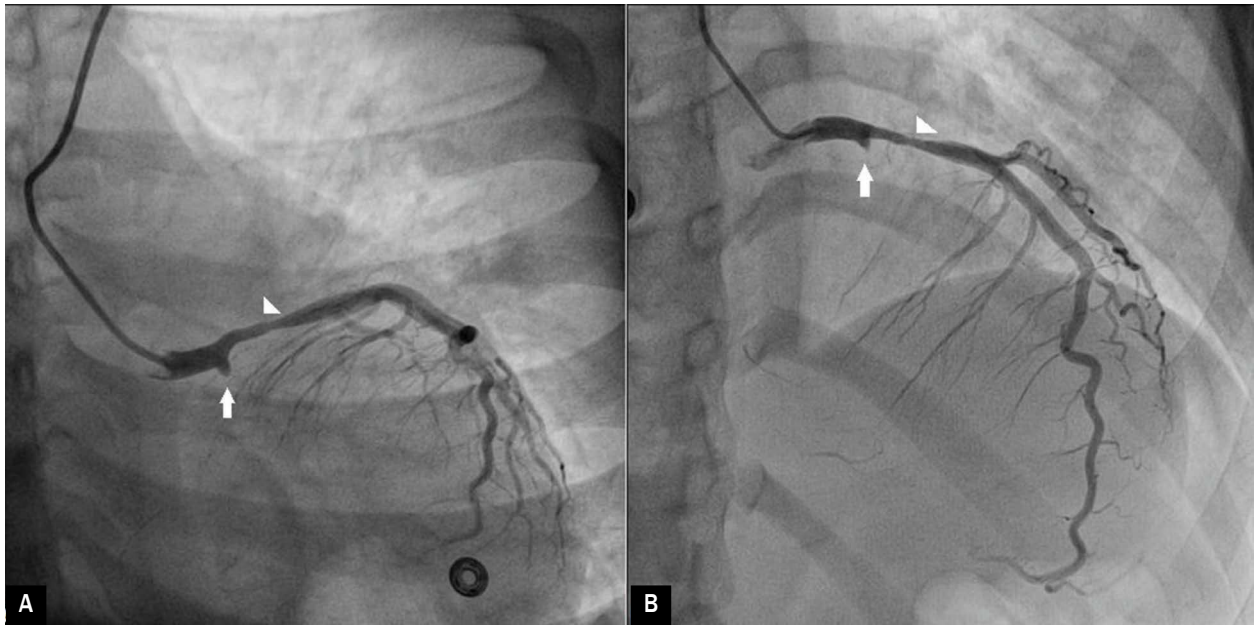
were no co-morbidities or family history of coronary artery disease. On examination, blood pressure in the left arm was 124/80 mm Hg, in the right arm was 82/68 mm Hg, while pulses in both lower limbs were weak. Her electrocardiogram revealed ST segment elevation in II, III, and aVF leads, and reciprocal changes in anterior leads. Laboratory tests showed elevated troponin T (0.8 ng/mL), total leukocyte count (13,800/ $\mu$ L), CRP (94 mg/L), and erythrocyte sedimentation rate (56 mm in first hr). Chest radiography was normal. Echocardiography showed mild concentric left ventricular hypertrophy and mildly impaired systolic function with an ejection fraction of 58%. Other forms of vasculitis such as polyarteritis nodosa, Kawasaki's disease, giant cell arteritis (GCA), and Cogan's syndrome were ruled out based on age, presentation, and pattern of vascular involvement. Rheumatoid factor (RA), anti-nuclear antibody (ANCA), anticardiolipin, and lupus anticoagulant titre were negative, thereby ruling out secondary forms of vasculitis. Coronary angiogram performed through the right transradial route failed as it showed total occlusion of the brachiocephalic trunk (Figure 1). Through the left transradial route, it revealed normal right coronary artery, total occlusion of left circumflex artery, and tubular lesion of the proximal left anterior descending artery with 70% stenosis (Figures 2, 3). Aortogram through the left transradial and right transfemoral routes showed total occlusion of the brachiocephalic trunk, diffuse thoraco-abdominal coarctation, and osteo-proximal stenosis (80%) of the left renal artery (Figure 4).

Based on the clinical presentation, angiographic findings, and having ruled out other possible causes, she was

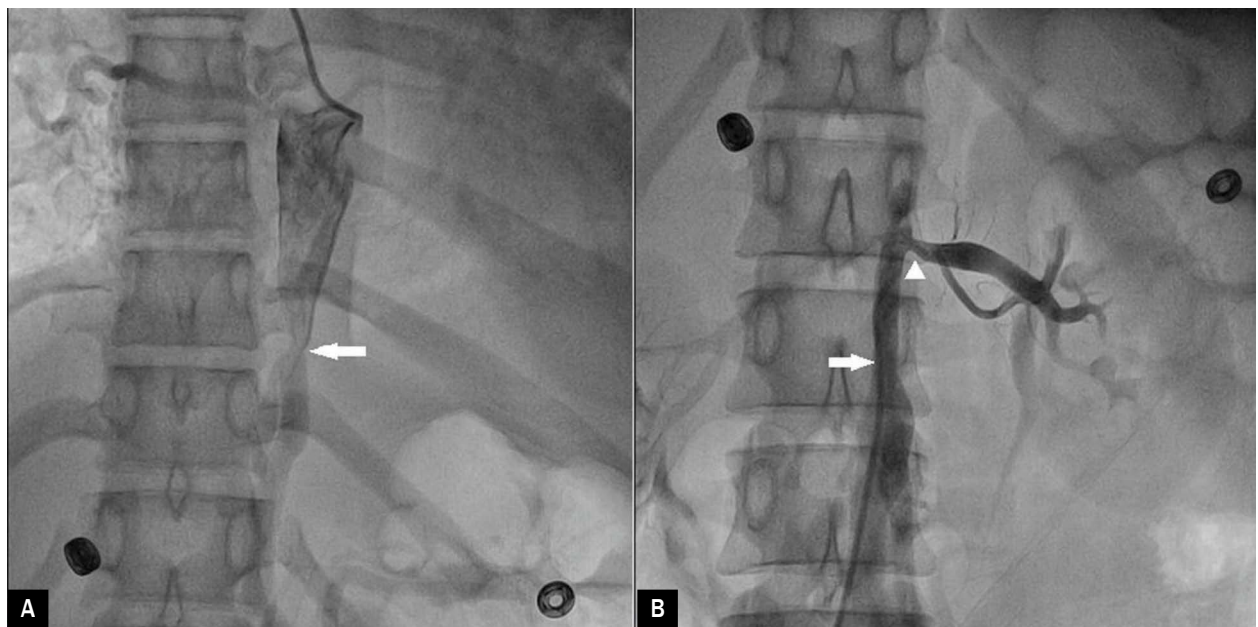
diagnosed to be in the active phase of juvenile Takayasu's arteritis having acute inferior wall myocardial infarction as supported by the EULAR (European League against Rheumatism)/PRES (Paediatric Rheumatology European Society) criteria [2]. Her treadmill test for reversible ischaemia (modified Bruce protocol) was negative, and therefore no intervention was performed. Because this was the active form of the disease, she was treated with intravenous cyclophosphamide pulse therapy (15 mg/kg/day in 3-weekly intervals), prednisolone 1 mg/kg/day, ecospirin 75 mg, clopidogrel 75 mg, amlodipine 5 mg, ramipril 5 mg, and hydrochlorothiazide 12.5 mg, and discharged.

## Discussion

Cardiac involvement in the form of coronary artery disease is seen in up to 30% of patients with TA, a disease which affects females more than males [3]. The usual age of involvement is late adulthood. Cardiac affliction occurs in two stages – in the initial acute stage as arteritis, and in the delayed chronic phase with vascular damage [3]. The temporal sequence of cardiac involvement is still not clear. Its spectrum spans from chronic stable angina to acute coronary syndrome. Usually, acute coronary syndrome is seen in the initial stages when conventional biomarkers of inflammation like CRP and ESR are elevated. In the acute stage with cardiac involvement, immunosuppressive therapy should be used to prevent the untoward complication and its sequelae [4]. Acute ischaemia is one of the main causes of death in TA, accounting for mortality of up to 50% at five years because of critical lesions including left



**Figure 3A, B.** Coronary angiogram through left transradial route showing total occlusion of left circumflex artery (A) and tubular lesion of proximal left anterior descending artery with 70% stenosis (B)



**Figure 4A, B.** Aortogram through left transradial and right transfemoral route showing total occlusion of brachiocephalic trunk, diffuse thoraco-abdominal coarctation, and osteo-proximal stenosis (80%) of left renal artery

main coronary artery [3]. Because TA primarily affects the aorta and its branches, the ongoing inflammation may engulf the coronaries as well as an extension of intimal proliferation and contraction of the fibrotic media and adventitia from the ascending aorta. It can be confirmed by catheter-based conventional angiography, CT angiography, and MRI [4]. In symptomatic cases, it can be treated with percutaneous coronary intervention (PCI)

implanting either bare-metal stents or drug-eluting stents, or CABG depending on comorbidities, age, and extent of disease. PCI is associated with a high risk of in-stent restenosis which can be mitigated by immunosuppressive treatment. Some authors have opined that drug-eluting stents serve as a useful bridge while immunosuppression is optimised and until definitive CABG can be performed [5]. Issues involving CABG are also complex because TA

commonly involves subclavian arteries, making the internal mammary artery the second choice to a saphenous vein graft, unlike most CABG procedures. Our patient had been asymptomatic since admission, and there was no identifiable ischaemia. This was why she was discharged with antiplatelets and immunosuppressive therapy. This was also in accordance with occluded artery trial (OAT), though in our patient the left circumflex artery was the infarct-related artery [6]. The active phase of TA in our case was based on raised acute phase reactants such as ESR and CRP. However, in one study by Soto et al involving 32 patients with TA, they were found to be elevated in only 78% of the study population [3]. This has been further

confirmed by histopathological studies wherein Lagneau et al demonstrated that in 40% of their patients with TA who were in clinical remission on the basis of acute phase reactants, active vasculitis was demonstrated in surgically obtained specimens [7].

Our case demonstrates the malignant nature of TA in the Indian sub-continent especially among juveniles, and also that the decision regarding intervention in the active stage can be judged using a non-invasive modality.

### Conflict(s) of interest

The authors declare no conflict of interest.

### Streszczenie

Dziewczynka w wieku 10 lat została przyjęta do szpitala w celu wyjaśnienia przyczyny nagłego bólu w klatce piersiowej, który wystąpił 3 dni wcześniej. Pacjentka wcześniej nie chorowała, a wywiad rodzinny w kierunku choroby wieńcowej był ujemny. Stwierdzono różnicę w wartościach ciśnienia tętniczego między prawym a lewym ramieniem, a tętno na kończynach dolnych było ledwo wyczuwalne. W zapisie elektrokardiograficznym widoczne było uniesienie odcinka ST w odprowadzeniach II, III i aVF. W badaniach laboratoryjnych wykazano podwyższone stężenie troponiny T (0,8 ng/ml), liczby leukocytów (13 800/ $\mu$ l), stężenia białka C-reaktywnego (94 mg/l) i wskaźnika opadania erytrocytów (56 mm w 1. godz.). Badanie echokardiograficzne ujawniło łagodne zaburzenie czynności skurczowej lewej komory z frakcją wyrzutową 58%. W aortogramie stwierdzono całkowitą okluzję pnia ramiennie-głowowego, rozsiane zwężenia w aorcie piersiowej i brzusznej oraz zwężenie ujścia i proksymalnego odcinka (80%) lewej tętnicy nerkowej. Koronarografia uwidoczniała całkowitą okluzję gałęzi okalającej i cewkowate zwężenie (70%) proksymalnego odcinka tętnicy przedniej zstępującej. Na podstawie wyników badań u chorej rozpoznano aktywną fazę młodzieńczego zapalenia tętnic Takayasu z ostrym zawałem ściany przedniej serca. Wynik przeprowadzonego u chorej ograniczanego objawami testu wysiłkowego na bieżni ruchomej (wg zmodyfikowanego protokołu Bruce'a) był ujemny. Chorą wypisano do domu i zalecono odpowiednią terapię (leki przeciwplateletowe, kortykosteroidy i leki immunosupresyjne).

Słowa kluczowe: młodzieńcze zapalenie tętnic Takayasu, ostry zespół wieńcowy, rozsiane zwężenia aorty

Folia Cardiologica 2019; 14, 1: 79–82

### References

1. Rav-Acha M, Plot L, Peled N, et al. Coronary involvement in Takayasu's arteritis. *Autoimmun Rev.* 2007; 6(8): 566–571, doi: [10.1016/j.autrev.2007.04.001](https://doi.org/10.1016/j.autrev.2007.04.001), indexed in Pubmed: [17854750](https://pubmed.ncbi.nlm.nih.gov/17854750/).
2. Ozen S, Ruperto N, Dillon MJ, et al. EULAR/PReS endorsed consensus criteria for the classification of childhood vasculitides. *Ann Rheum Dis.* 2006; 65(7): 936–941, doi: [10.1136/ard.2005.046300](https://doi.org/10.1136/ard.2005.046300), indexed in Pubmed: [16322081](https://pubmed.ncbi.nlm.nih.gov/16322081/).
3. Soto ME, Espinola-Zavaleta N, Ramirez-Quito O, et al. Echocardiographic follow-up of patients with Takayasu's arteritis: five-year survival. *Echocardiography.* 2006; 23(5): 353–360, doi: [10.1111/j.1540-8175.2006.00238.x](https://doi.org/10.1111/j.1540-8175.2006.00238.x), indexed in Pubmed: [16686616](https://pubmed.ncbi.nlm.nih.gov/16686616/).
4. Endo M, Tomizawa Y, Nishida H, et al. Angiographic findings and surgical treatments of coronary artery involvement in Takayasu arteritis. *J Thorac Cardiovasc Surg.* 2003; 125(3): 570–577, doi: [10.1067/mtc.2003.39](https://doi.org/10.1067/mtc.2003.39), indexed in Pubmed: [12658199](https://pubmed.ncbi.nlm.nih.gov/12658199/).
5. Lee HK, Namgung J, Choi WH, et al. Stenting of the left main coronary artery in a patient with Takayasu's arteritis. *Korean Circ J.* 2011; 41(1): 34–37, doi: [10.4070/kcj.2011.41.1.34](https://doi.org/10.4070/kcj.2011.41.1.34), indexed in Pubmed: [21359067](https://pubmed.ncbi.nlm.nih.gov/21359067/).
6. Hochman JS, Lamas GA, Knatterud GL, et al. Occluded Artery Trial Research Group. Design and methodology of the occluded artery trial (OAT). *Am Heart J.* 2005; 150(4): 627–642, doi: [10.1016/j.ahj.2005.07.002](https://doi.org/10.1016/j.ahj.2005.07.002), indexed in Pubmed: [16209957](https://pubmed.ncbi.nlm.nih.gov/16209957/).
7. Lagneau P, Michel JB, Vuong PN. Surgical treatment of Takayasu's disease. *Ann Surg.* 1987; 205(2): 157–166, indexed in Pubmed: [880571](https://pubmed.ncbi.nlm.nih.gov/880571/).