Cardiology in the Young

cambridge.org/cty

Acute coronary syndrome due to multi-vessel coronary artery spasm in an Afghan refugee adolescent mimicking recurrent myocarditis

Yasemin Nuran Donmez[1], Mehmet Erat[2], Ayse Esra Tapci[3] and Hasan Yigit[4]

[1]Pediatric Cardiology, Ankara Training and Research Hospital, Ankara, Turkey; [2]Cardiology, Ankara Training and Research Hospital, Ankara, Turkey; [3]Pediatrics, Ankara Training and Research Hospital, Ankara, Turkey and [4]Radiology, Ankara Training and Research Hospital, Ankara, Turkey

Brief Report

Cite this article: Donmez YN, Erat M, Tapci AE, and Yigit H (2023) Acute coronary syndrome due to multi-vessel coronary artery spasm in an Afghan refugee adolescent mimicking recurrent myocarditis. *Cardiology in the Young* **33**: 2434–2437. doi: 10.1017/S1047951123002573

Received: 6 April 2023
Revised: 25 May 2023
Accepted: 28 June 2023
First published online: 24 July 2023

Keywords:
Chest pain; vasospastic angina; selective coronary angiography; acute coronary syndrome

Corresponding author:
Yasemin Nuran Donmez;
Email: yaseminnurandonmez@gmail.com

Abstract

Vasospastic angina is extremely uncommon for adolescents to experience chest discomfort, which is defined by transitory ST segment elevation or depression and angina symptoms that occur while at rest. It may result in potentially fatal conditions like myocardial infarction, ventricular fibrillation, or even sudden cardiac arrest. To aim of this article is to report a very rare case of a 17-year-old male Afghan refugee who was diagnosed with vasospastic angina after presenting with chest pain, and after receiving calcium channel blocker and nitrates for medical therapy, there were no angina attacks. Our case underlines the value of a thorough evaluation of adolescent's chest pain, the need to diagnose based on the symptoms, and the necessity of performing coronary angiography to rule out coronary causes when there is a high suspicion to a cardiac cause.

Chest pain is often benign in children and adolescents, but it is crucial to recognise cardiac reasons because they carry a significant risk of developing into life-threatening illnesses. To rule out cardiac causes, chest pain characteristics, cardiac enzyme, and electrocardiographic results can be employed.[1] A cardiac cause, such as myocarditis, myocardial ischemia, or structural heart disease, is suspected due to the chest discomfort radiating to the left arm and jaw, the sensation of pressure on the chest, and the concomitant symptoms.[2] Vasospastic angina is a very rare and a difficult-to-diagnose cardiac cause of adolescent chest pain. We report of a 17-year-old Afghan refugee child who had 6 months of myocarditis treatment before being diagnosed with vasospastic angina after experiencing ongoing chest pain.

Case report

A 17-year-old male was referred to our unit with myocarditis suspicion because of chest pain, an increase in cardiac enzymes, and abnormal electrocardiographic results. The patient was an Afghan refugee. His medical history revealed that he had presented with chest pain two more times within a year, that his tests showed elevated cardiac enzymes. He had started treatment for myocarditis with aspirin and metoprolol, and that he had been taking these medications for about 5 months. The characteristics of chest pain was pressure on his chest, no change with respiration nor position, radiating in his left shoulder, left arm, and jaw. The chest pain was discovered to be predominantly in the evening and to last for around 5 minutes when at rest. The patient also did not have any significant family medical history, such as hyperlipidemia or early myocardial infarction. He had never had a fever for Kawasaki disease, any other systemic disease, been overweight, smoked, substance use, or physical inactivity.

Upon examination, he had a heart rate of 60 bpm, a respiratory rate of 20 bpm, and blood pressure of 119/69 mmHg. A grade 2/6 short systolic murmur was audible on the mitral region during cardiac auscultation. *On blood examination*, N-terminal pro-B-type natriuretic peptide 419 ng/L, hs troponin T level 121 ng/L (0–14), HDL 36 mg/dL (40–85), creatine kinase mb 2,5 µg/L (0–4,94), white blood cell count 5,5 × 109/L (4–10), and CRP 0,7 mg/L (<5) were detected. The erythrocyte sedimentation rate, kidney and liver function tests, and other laboratory parameters were within the normal range. *Chest X-ray* was unremarkable. *Electrocardiogram* revealed T wave inversion and pathologic Q wave in the inferior leads, R dominance and ST elevation in the right precordial leads with a rate of 56 bpm. *Echocardiography* revealed posterolateral wall motion abnormality, mild left ventricular posterior wall thickness (PW:12 mm + 2 z score, S:12 mm + 1,5 z score) with a low left ventricular ejection fraction (50%), and mild mitral regurgitation. We first assumed that the results of the electrocardiographic and echocardiogram indicated recurrent myocarditis, and we continued to provide a combination of metoprolol and acetylsalicylic acid as treatment. On the fourth day of his hospitalisation, we had a cardiac MRI for

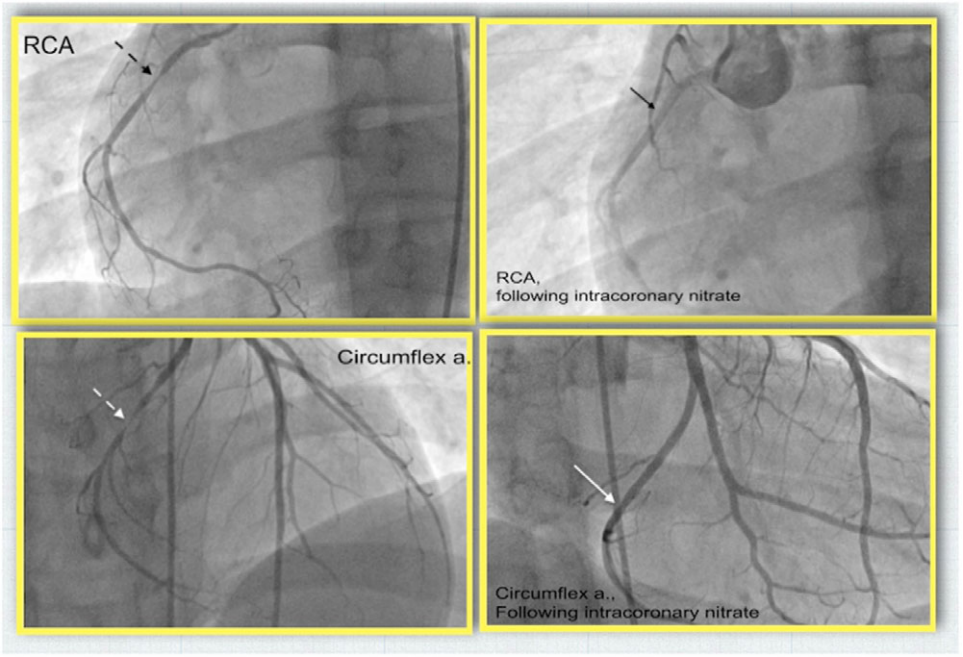

Figure 1. Images from selective coronary angiography showing a spasm in the coronary arteries and how it improved after being treated with intracoronary nitrate. (***a***) Spontaneous vasospasm was seen in the right coronary artery in the left anterior oblique position. (***b***) The spasm resolves after intracoronary nitrate administration. (***c***) A left coronary angiogram's anterior-posterior (AP) cranial view demonstrating circumflex artery vasospasm. (***d***) The spasm resolves after intracoronary nitrate administration.

myocarditis. *Cardiac MRI* revealed an increase in thickness of anteroseptal, inferolateral, and inferoseptal of left ventricle. Subepicardial hypoperfusion on early postcontrast images and subepicardial contrast enhancement on delayed post-contrast images was observed in the left ventricle basal and midventricular inferoseptal, inferior and lateral walls suitable for left circumflex artery and right coronary artery areas (Fig. 1). Hypokinesia was also observed at sites corresponding to this region. Findings were consistent with subacute infarct. *Selective coronary angiography* was planned immediately with the suspicion of infarct in cardiac MRI. Selective coronary angiography showed vasospasm in the distal branch of circumflex and mid right coronary artery (Fig. 2) After the administration of glyceryl trinitrate, opening of vasospasm was observed. The constellation of findings described above established a diagnosis of vasospastic angina. Previous medications were discontinued, and the combination of nitrate and calcium blocker treatment was started. Cardiac enzymes and other laboratory results for the patient were back to normal. Throughout the patient's hospitalisation, there were no angina attacks after maximising doses. He was discharged after 12 days with diltiazem (180 mg daily), isosorbide mononitrate (20 mg daily), statin, and physical activity restriction recommendations. Four months after being discharged, the patient reported no symptoms. Since infarct repair began after 5 months, an MRI was scheduled to be done at a later time.

Discussion

Vasospastic angina (previously known as Prinzmental or variant angina) is coronary artery spasm characterised by anginal symptoms at rest, with transient ST segment elevation or depression and angina that is responsive to nitrates during the time of the episode.[3] It is thought that coronary arteries are more sensitive to vasoconstrictor factors and the pathogenesis is influenced by hyperactive vascular smooth muscles, abnormalities in the autonomic nervous system, magnesium deficiency, microvascular dysfunction, and endothelial dysfunction.[3-5] These factors may cause localised or widespread spasm, which can lead to dynamic coronary blockage.

The clinical presentation is usually with anginal symptoms; however, it may cause syncope, myocardial infarction, arrhythmias (ventricular fibrillation or AV block), or sudden cardiac death if it persists.[4-7] Chest pain is characterised by chronic and recurrent bouts and responds favourably to nitrates. Chest pain can last for more than a year, with the longest recorded period being 36 months.[6] Chest pain is often seen at rest (more so at night or in the morning) and feels like pressure in the chest that spreads to the arm, neck, upper extremities, and back, which does not change with breathing.[5] Chest pain may accompany nausea, palpitations, shortness of breath, and sweating during the anginal episode.[5] For about a year, our patient complained of chest pain that was characterised by pressure on the chest, and spreading to the left shoulder, left arm, and jaw, experienced chest pain during rest and in the evening. He visited the emergency room three times with this complaint.

A significant risk factor for coronary spasm is smoking. Hyperventilation, the cold, exercise, stress, magnesium deficit, and several medications (triptans, ephedrine, beta blockers, ergonovine, and high doses of aspirin) are also known to trigger cardiac vasospasm.[4,5] Our patient had never smoked before, and we believe that his situation as a refugee and his living environment may have been a trigger as a stress factor. Release of vasopressor neurotransmitters such noradrenaline, platelet activation, and serotonin, and abnormal autonomic nervous system function leads to vasospasm in response to stress.[8] Ethnic differences are another risk factor, with Asian groups having a higher incidence.[5]

In the absence of anginal symptoms in vasospastic angina, physical examination, electrocardiography, and laboratory findings are unremarkable. Depending on where the damaged vasospastic coronary artery is located and how long the episodes last, pathologic findings are seen in the electrocardiography. The presence of a brief ST depression or elevation and a negative u wave during vasospastic angina is diagnostic.[3] Selective coronary

https://doi.org/10.1017/S1047951123002573 Published online by Cambridge University Press

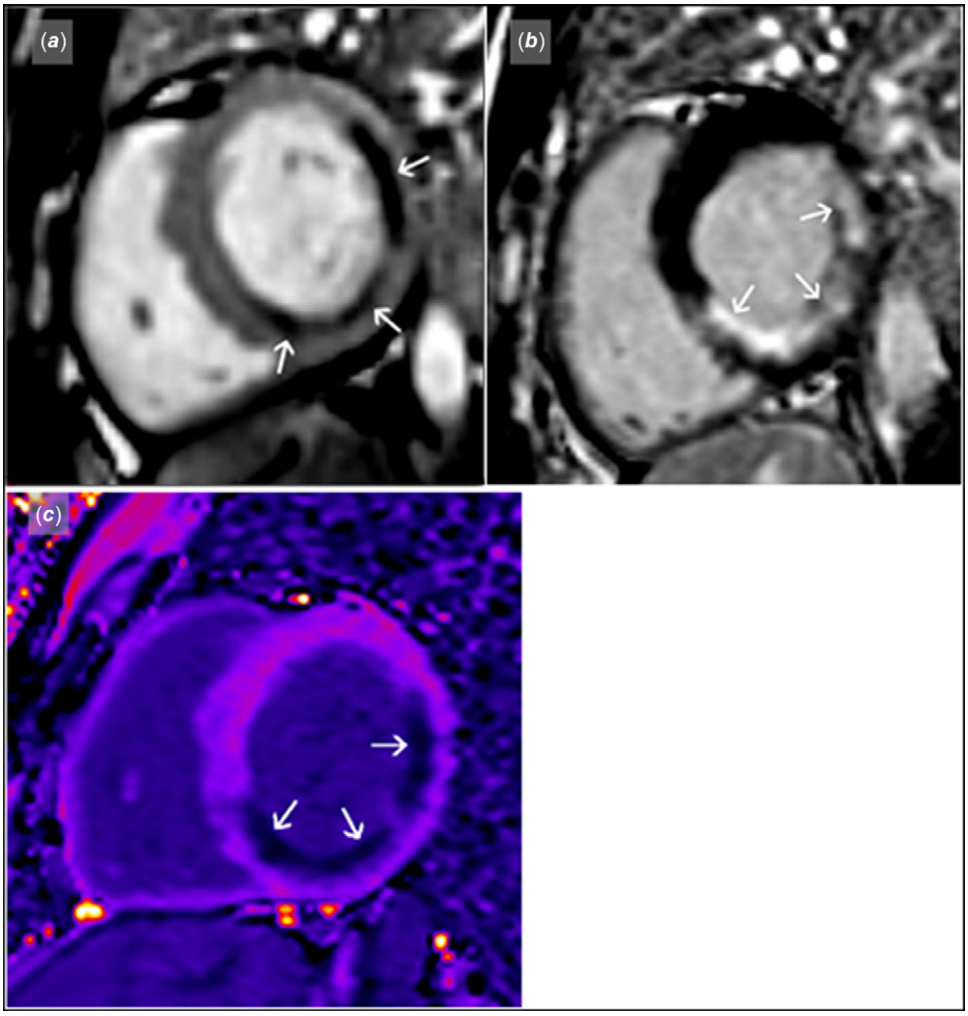

Figure 2. (***a***) Short-axis early postcontrast images shows subepicardial hypoperfusion in Lcx and RCA territory. (***b***) Contrast enhancement suitable for infarct observed on short-axis delayed postcontrast images. (***c***) T1 map.

angiography may be planned on high suspicion of chest pain and spasm may be induced by provocative tests with ergonovine and acetylcholine.[6] Because of the patient's young age, the prevalence of myocarditis, the difficulty in detecting vasospastic angina, the chronic nature of the condition and findings of echocardiography and electrocardiography, we believed that the patient was misdiagnosed with the diagnosis of recurrent myocarditis that he may have had complaints related to multivessel coronary spasm in his previous admissions. Our patient's incorrect diagnosis of myocarditis resulted in him being treated with beta-blockers, which are known to make symptoms worse by causing a stronger vasoconstrictive reaction in cases of vasospastic angina. Selective coronary angiography showed spontaneous coronary spasm and the spasm disappeared with intracoronary nitroglycerine.

Both pharmacological therapy and lifestyle adjustments should be used to control the vasospastic angina. Triggers (stress, alcohol, and cold) should be avoided, and smoking should be discontinued. Improved anginal symptoms and relief from vasospasm are achieved with calcium canal blocker (diltiazem, amlodipine, or long acting nifedipine) and nitrate (isosorbide mononitrate). Statins may be chosen due to their effects on acetylcholine, one of the causes of coronary spasm, as well as their impact on endothelial activities.[7] In refractory cases with persistent symptoms, where Mg deficiency may also be one of the reasons, long-term Mg support may be given.[7] Class I recommendations for calcium channel blockers, class IIa recommendations for long-acting nitrates, and class IIb recommendations for statin therapy are made in the 2013 Japanese Circulatory Society Guidelines for VSA.[8] Following the recommended protocol, we gave our patient these three medications.

In conclusion, the patient we are presenting had already received a diagnosis of myocarditis about a year before, but the chest pain persisted. Although coronary artery disease in teenagers is extremely rare, it is crucial to maintain a high threshold for suspicion when chest pain persists and there are pathologic echocardiography and electrocardiography signs indicative of cardiac conditions. The results of the patient's cardiac MRI were also very useful in confirming the diagnosis, and we were able to establish vasospastic angina by selective coronary angiography based on the MRI infarct findings. Angina episodes resolved with calcium channel blocker and nitrate treatment.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human medical regulations and with Helsinki Declaration of 1975, as revised in 2008.

Informed consent. Written informed consent was obtained from all patient's parents.

https://doi.org/10.1017/S1047951123002573 Published online by Cambridge University Press

References

1. Chen L, Duan H, Li X, Yang Z, Jiao M, Sun K, Jin M. The causes of chest pain in children and the criteria for targeted myocardial enzyme testing in identifying the causes of chest pain in children. Front Cardiovasc Med 2021; 8: 582129.
2. Collins SA, Griksaitis MJ, Legg JP. 15-minute consultation: a structured approach to the assessment of chest pain in a child. Arch Dis Child Educ Pract Ed 2014; 99: 122–126. DOI: 10.1136/archdischild-2013-303919.
3. Beltrame JF, Crea F, Kaski JC, et al. International standardization of diagnostic criteria for vasospastic angina. Eur Heart J. 2017; 38: 2565–2568. DOI: 10.1093/eurheartj/ehv351.
4. Tandon V, Mosebach CM, Kumar M, Joshi S. Refractory vasospastic angina: when typical medications don't work. Cureus 2019; 11: e4134. DOI: 10.7759/cureus.4134.
5. Picard F, Sayah N, Spagnoli V, Adjedj J, Varenne O. Vasospastic angina: a literature review of current evidence. Arch Cardiovasc Dis. 2019; 112: 44–55. DOI: 10.1016/j.acvd.2018.08.002.
6. Sueda S. Young vasospastic angina patients less than 20 years old. Circ J. 2019; 83: 1925–1928. DOI: 10.1253/circj.CJ-19-0433.
7. Stern S, Bayes de Luna A. Coronary artery spasm: a 2009 update. Circulation. 2009; 119: 2531–2534. DOI: 10.1161/CIRCULATIONAHA.108.843474.
8. Group JCSJW. Guidelines for diagnosis and treatment of patients with vasospastic angina (Coronary Spastic Angina) (JCS 2013). Circ J. 2014; 78: 2779–2801. DOI: 10.1253/circj.cj-66-0098.

https://doi.org/10.1017/S1047951123002573 Published online by Cambridge University Press