Archives of the Balkan Medical Union
vol. 53, no. 1, pp. 129-134
March 2018

CASE REPORT

SPONTANEOUS CORONARY ARTERY DISSECTION AS A CAUSE OF ACUTE MYOCARDIAL INFARCTION

Lucia Cojocaru[1,2], Luminița Matei[1,2], Irinel R. Parepa[1,2], Andrei C. Rusali[1,2], Elvira Craiu[1]

[1] „Ovidius" University of Constanta, Faculty of Medicine, Constanta, Romania
[2] Emergency Clinical Hospital of Constanta, Cardiology Clinic, Constanta, Romania

Abstract

Spontaneous coronary artery dissection is a rare diagnosis in a postmenopausal woman. We report a case of a 55-year-old postmenopausal woman, who presented with acute myocardial infarction due to spontaneous long dissection of left anterior descending coronary artery, complicated with repetitive ventricular tachycardia, rapid formation of left ventricular aneurysm with thrombus, pulmonary edema and cardiogenic shock, with subsequent improvement after successful angioplasty and intensive care. We also present a short review of the literature.

Key words: coronary artery, spontaneous dissection, acute myocardial infarction.

Abbreviations:
ACS = acute coronary syndrome;
AMI = acute myocardial infarction;
BMSs = bare-metal stents;
bpm = beats per minute;
CAGB = coronary artery bypass grafting;
CV = cardiovascular;
ECG = electrocardiogram;
LAD = left anterior descending coronary artery;
LVEF = left ventricle ejection fraction;
PCI = percutaneous coronary intervention;
SCAD = Spontaneous coronary artery disease;

Résumé

Dissection spontanée de l'artère coronaire chez une femme ménopausée comme cause d'infarctus aigu du myocarde

La dissection spontanée de l'artère coronaire est un diagnostic rare chez une femme ménopausée. Nous présentons le cas d'une femme ménopausée âgée de 55 ans présentant un infarctus aigu du myocarde dû à une longue dissection spontanée de l'artère coronaire descendante antérieure gauche compliquée de tachycardie ventriculaire répétitive, de formation rapide d'un anévrisme ventriculaire gauche avec thrombus, d'œdème pulmonaire et de choc cardiogénique, avec amélioration après une angioplastie réussie et des soins intensifs. Nous présentons également une brève revue de la littérature.

Mots-clefs: artère coronaire, dissection spontanée, infarctus aigu du myocarde.

Corresponding author: Lucia Cojocaru
Emergency Clinical Hospital of Constanta, Cardiology Clinic, Tomis Boulevard No 145, Constanta, Romania
phone +40722370635; e-mail address: bostanlucia@yahoo.com

Introduction

Spontaneous coronary artery dissection (SCAD), a non-traumatic and non-iatrogenic separation of the coronary wall[1], is a rare yet increasingly recognized cause of acute coronary syndrome, especially in younger female patients without traditional vascular risk factors[2].

Since its first description in 1931[3] more cases are now identified due to increased awareness and earlier use of angiography[4]. Despite that, the pathophysiology of SCAD remains poorly understood[5-8], the clinical features and prognosis are insufficiently characterized and the management of patients remains highly controversial[5].

It is important to recognize SCAD, as patient characteristics and management differ from typical ACS.

Case presentation

We present the case of a 55-year-old female, heavy smoker (more than 40 pack-year) and drinker, postmenopausal (menopause at 50 years old), known with a spontaneous abortion (at 38 years of age, age of pregnancy – 6 months), with a family history of cardiovascular (CV) disease (three brothers with acute myocardial infarction under 55 years old) and without past medical history. She arrived at the hospital by ambulance, three hours after the onset of epigastric and retrosternal pain, followed by palpitations. During the transport by ambulance, she presented an episode of ventricular tachycardia responsive to antiarrhythmic medication (amiodarone).

On the arrival at the hospital, she was alert and oriented, afebrile, in sinus rhythm, with a heart rate of 80 bpm, blood pressure of 120/80 mmHg, respiratory rate of 18 breaths per minute, an oxygen saturation of 98% while breathing ambient air, with normal findings at the physical exam. The electrocardiogram (ECG) at admission revealed sinus rhythm, 83 bpm, +30 degree axis, Q waves in V3-V6, elevation of the ST segment in V1-V5, negative T waves in DI, aVL, V3-V6 and an corrected QT interval of 376 ms (Figure 1). Transthoracic echocardiography revealed normal cardiac chambers size, normal aspect of the ascending aorta and arch, mild degenerative mitral insufficiency, mild functional tricuspid insufficiency, no indirect signs of pulmonary hypertension, type I diastolic dysfunction, apical aneurysm with mobile thrombus, moderate systolic dysfunction of the left ventricle (LVEF=35%) and no pericardial fluid (Figure 2). Laboratory tests showed high values of myocardial necrosis markers, mild sideropenic anemia, dyslipidemia with low high-density lipoprotein cholesterol and high triglyceride values, high values

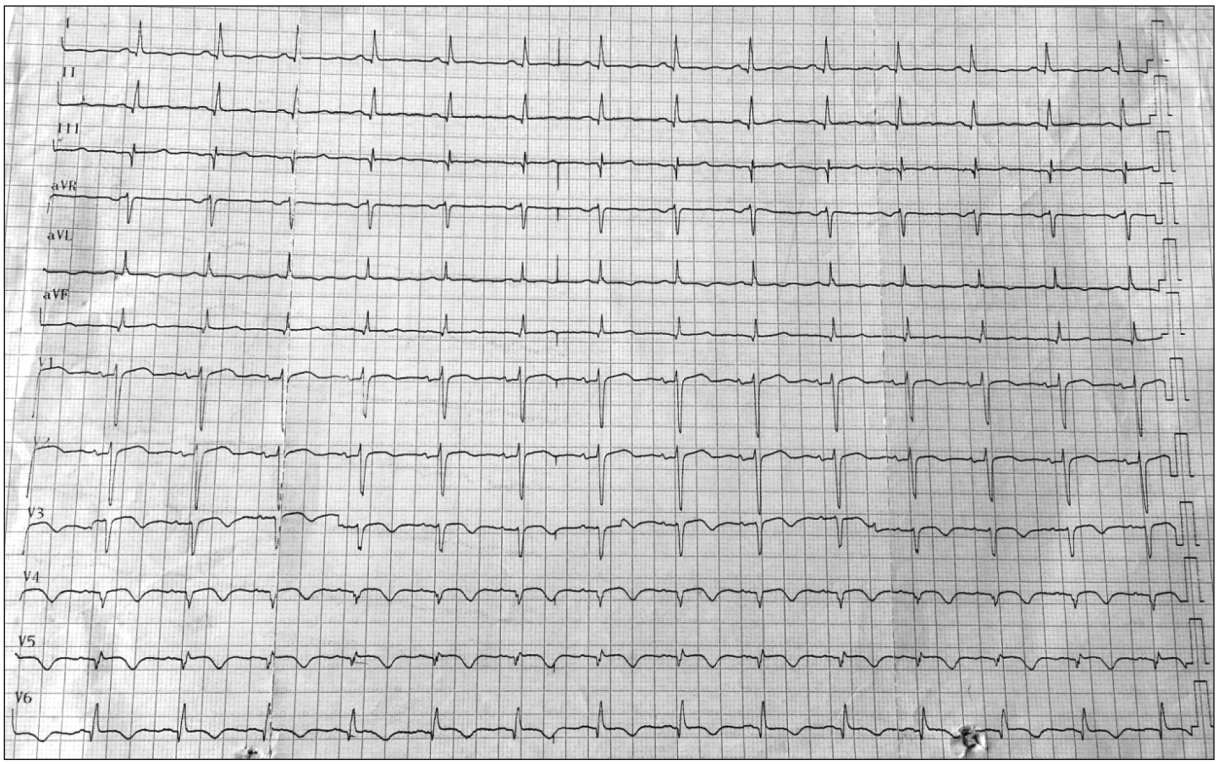

Figure 1. ECG on admission: sinus rhythm, 83 bpm, +30 degree axis, Q waves V3-V6, elevation of the ST segment in V1-V5 and negative T waves in DI, aVL, V3-V6. The corrected QT interval is 376 ms.

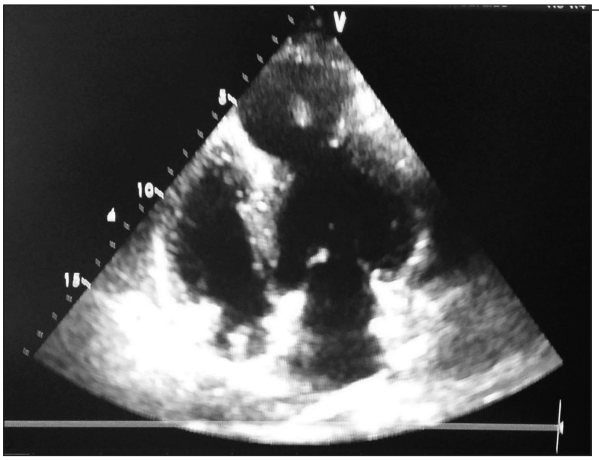

Figure 2. Transthoracic echocardiography (apical 4 chamber view) showing apical aneurysm with mobile thrombus.

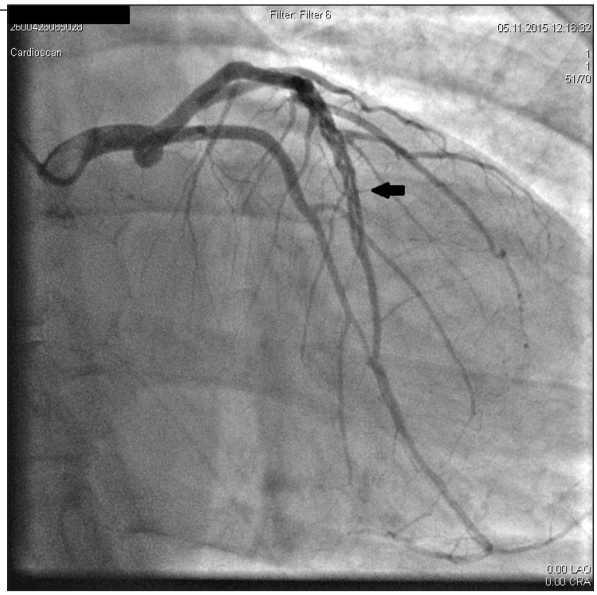

Figure 3. Coronarography (5 November 2015): long dissection flap (longitudinal filling defect – black arrow) in the mid and distal segment of LAD.

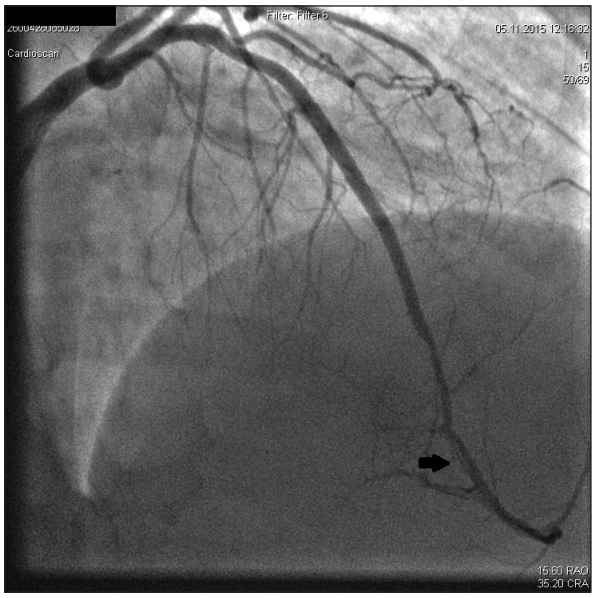

Figure 4. Angioplasty with implantation of two BMSs in the mid segment of LAD. Residual small dissection in the distal segment of LAD (black arrow).

of thyroid stimulating hormone and free T4 (thyroxine).

The patient was referred to the cath lab for emergency coronarography, which revealed a long type 1 dissection, from the mid to distal segment of the left anterior descending coronary artery (LAD) (Figure 3). Given the severity of the dissection and its clinical manifestations, the decision was made to perform an angioplasty with stenting. Two bare-metal stents (BMSs) were successfully implanted in the mid segment of LAD with TIMI3 flow (Figure 4). Twelve hours after angioplasty, the patient presented another episode of ventricular tachycardia with hemodynamic instability, treated by electrical cardioversion, followed by pulmonary edema and cardiogenic shock, that required orotracheal intubation and ventilator support, inotropic and vasopressor support. After another twelve hours of intensive care, the patient was stabilized, alert and oriented, with spontaneous respiration, oxygen saturation of 100%, heart rate of 98 bpm and blood pressure 120/80 mmHg without inotropic support. After treatment with anticoagulant, double antiplatelet therapy, antiarrhythmic drugs (beta-blocker and amiodarone), angiotensin converting enzyme inhibitor, loop diuretic, aldosterone blocker, statin and thyroid replacement hormone, the clinical evolution was good and the patient was discharged hemodynamically stable, without angina.

After discharge from the hospital, the patient was lost to follow-up for two years, until she presented another episode of palpitations with retrosternal pain and dyspnea. At the admission, the patient had ventricular tachycardia with hemodynamic instability, and she was successfully treated by electrical cardioversion. The electrocardiogram post-cardioversion revealed sinus rhythm, with a heart rate of 63 bpm, Q waves in V3-V6, negative T waves in DI, aVL, V3-V6 and a corrected QT interval of 520 ms (under venous infusion of amiodarone). The transthoracic echocardiography was similar to the previous one, but without the thrombus in the apical aneurysm. An emergency coronarography was performed, that revealed permeable stents at the level of mid segment of LAD and the previous dissection in the distal segment of LAD, without other coronary lesions (Figure 5). Although previous studies showed that the majority of patients treated conservatively had spontaneous angiographic healing[4], our patient's dissection in the distal segment of LAD was unchanged. Given

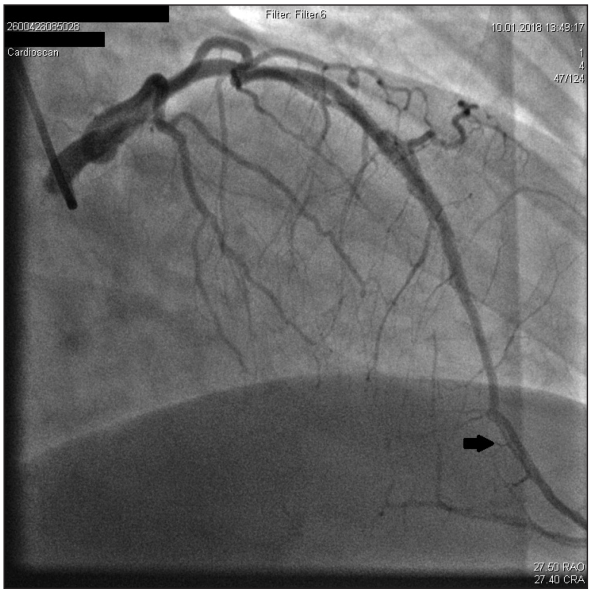

Figure 5. Coronarography (10 January 2018): permeable BMSs in the mid segment of LAD. Residual small dissection in the distal segment of LAD (black arrow).

the stable character of the remaining coronary dissection during these two years and the distal location without impairing of the blood flow, a conservative approach was preferred. Since fibromuscular dysplasia is an associated and possibly causal factor[9], the patient underwent a screening computed tomographic angiography, that showed normal renal, iliac and cerebral vessels. During the hospitalization, the patient repeated an episode of sustained ventricular tachycardia (unresponsive to amiodarone and lidocaine, that required electrical cardioversion) and short episodes of unsustained ventricular tachycardia. She was referred to the cardiac surgery department for left ventricular reconstructive surgery.

Discussion

SCAD is a sudden separation between the layers of a coronary artery wall, that creates an intimal flap or intramural hematoma[9]. The underlying mechanism is not fully understood, but an intimal tear or bleeding of vasa vassorum with intramedial hemorrhage has been suggested[1, 10].

Retrospective registry studies have reported SCAD detection in 0.07% to 1.1% of all coronary angiograms performed[11]. In the general population, SCAD is the cause of acute coronary syndrome (ACS) in 0.1% to 0.4% of cases, affecting especially women, SCAD being the cause of 24% of cases of ACS in women under 50 years of age[1]. Although classically believed to affect young women, SCAD is now increasingly recognized to occur in older, postmenopausal women[1], as in our patient. In a study of 168 patients, the mean age was 52 years and 92% were women, with 62% being postmenopausal[12].

In the vast majority of patients, the cause of SCAD remains uncertain, although numerous associated conditions have been identified[9]. The commonest identified predisposing factors were fibromuscular dysplasia, pregnancy, multiparity (≥4 births), postpartum status, connective tissue disorders (Marfan's syndrome, Ehlers-Danlos syndrome – type IV), systemic inflammatory conditions (periarteritis nodosa, systemic lupus erythematosus, eosinophilia) and hormonal therapy[1,9,11,12]. Most patients with SCAD do not have traditional risk factors for atherosclerosis, although there is a weak association with hypertension and smoking[13]. Precipitating factors (that increase cardio-circulatory stress) can provoke the SCAD (especially if a predisposing arteriopathy exists)[1]. These stressors include labor and delivery, intense Valsalva-type activities (coughing, retching, vomiting), intense emotional stress, extreme physical exertion, sympathomimetic drugs (cocaine, amphetamines), aggressive hormonal therapy[1,4]. In our patient we did not find a predisposing or precipitating factor, but she had an important risk factor for atherosclerosis (smoking).

Patients with SCAD are rarely asymptomatic[14] and usually present with signs and symptoms characteristic of AMI[1]. Life-threatening ventricular arrhythmias and sudden cardiac death are known early complications[4].

The diagnosis of SCAD requires a high degree of suspicion and a careful angiographic study[9]. SCAD can be classified into three types based on the coronary angiographic appearance[1,15]:

- Type 1 – occurs in 29% of cases and consists in the presence of a longitudinal filling defect, representing the radiolucent intimal flap; there is often staining of the arterial wall with appearance of a double lumen[4];
- Type 2 – the most common type, consists in the presence of diffuse long smooth tubular lesions, due to intramural hematoma, with no visible dissection plane, that can result in complete vessel occlusion[4];
- Type 3 – occurs in 4% of cases and consists in multiple focal tubular lesions due to intramural hematoma that mimic atherosclerosis[4].

If the diagnosis is not certain, imaging of the vascular wall with intravascular ultrasound or optical coherence tomography may be helpful (especially for the diagnosis of type 2 and 3).

The optimal treatment strategy for patients with SCAD remains uncertain, due to the limited clinical experience[1], and may vary based on the type and severity of presentation[11]. Although comparative

studies between treatment modalities (conservative management, thrombolysis, percutaneous coronary intervention and coronary artery bypass grafting) do not exist, the conservative management is preferred in stable patients with normal flow in the affected coronary artery[11,16], based on reports of documented angiographic resolution of most dissected segments[5,11].

Patients with AMI who have ongoing ischemia or hemodynamic instability should be considered for revascularization, preferable with percutaneous coronary intervention (PCI)[1,12,16]. PCI in patients with SCAD is technically challenging, due to fragility of the vessel wall, the difficulty of retaining the guidewires within the true lumen and to the fact that any instrumentation can propagate dissection and occlude side branches[1,4]. The reported success rates are less than 50%[12,16], thus PCI should only be pursued when there is a strong clinical indication, in centers with on-site cardiac surgery[4], ideally with intracoronary imaging guidance to optimize stenting, and a drug-eluting stent should be used if a long stent is anticipated[17]. When the revascularization strategy is pursued, but the percutaneous option is unsuccessful or not technically feasible, coronary artery bypass grafting (CAGB) is the alternative. The patients who underwent CAGB as an initial strategy fared well in the short term, but a high rate of late bypass graft occlusion was reported[11].

In our patient, we opted for a hybrid approach – we treated the dissection in the mid LAD segment by angioplasty and stent implantation (due to the big caliber of the affected coronary artery, severity of the dissection with impairment of distal flow, unlikely to resolve with medical treatment alone, and due to recurring chest pain and sustained ventricular tachycardia) and the dissection of the distal segment of LAD was managed with a „conservative" therapeutic strategy (due to its distal location without impairing of the blood flow).

Although thrombolytic agents have been used[5,11,18] in SCAD patients, there is increased concern that fibrinolytic therapy could propagate the extension of intramural hematoma[4]. If there is a high index of suspicion for SCAD, if it is possible, it is prudent to transfer the patient to a facility with catheterization capacity[19].

Medical recommendations in SCAD are similar to standard acute coronary syndromes, based on opinion[4]. The recommended antiplatelet therapy consists of aspirin, if the patient is treated conservatively, and aspirin plus clopidogrel (for 1 year)[1] after stent placement, with no data on the role of ticagrelor or prasugrel[4]. Anticoagulants (unfractionated heparin or low-molecular-weight heparin) are advised in the first days after dissection[13,20]. Glycoprotein IIb/IIIa inhibitors have been used without complications, but they could potentially delay healing of the intramural hematoma and lead to dissection extension[4] so more data are needed. Nitrates can be used to relieve chest pain and long-acting nitrates are usually effective for potential coronary vasospasm, with a lower risk of hypotension than calcium channel blockers[9]. Beta-blockers are recommended in all patients, with the potential to reduce arterial shear stress, facilitate healing and reduce long-term recurrence[4]. Angiotensin-converting-enzyme inhibitors and angiotensin II receptor blockers are of particular interest, because they may inhibit the expression of matrix metallo-proteinases and stabilize the vessel wall[13]. The benefit of statins in SCAD is unknown, so they are recommended only when dyslipidemia is present (goal: low-density-lipoprotein cholesterol level < 100mg/dL)[1,9].

Due to susceptibility of recurrence, it is recommended that SCAD patients stay at least one week of observation in the hospital[13]. During the hospital stay, the patient must be evaluated for possible causes of SCAD, especially fibromuscular dysplasia.

The 10-year risk of dissection recurrence is up to 20%, predominantly in women, so close long-term follow-up is needed[9,11]. Due to risk of new or worsening dissection of coronary arteries in patients with SCAD, it is recommended to avoid repeated coronary angiography solely for monitoring purposes[9]. Instead, functional testing or coronary computed tomographic angiography can be used[9]. Cardiac rehabilitation should be recommended to all patients with SCAD. Moderate intensity aerobic physical activity should be encouraged, with the avoidance of weightlifting and bodybuilding, competitive racing or athletic pursuits at high levels [9]. Although there are no reports of recurrent SCAD in successive pregnancies[17], pregnancy in these patients is not recommended, even in those who did not experience peripartum SCAD[9].

In general, the prognosis of patients with SCAD is good, if they survive the acute phase[13]. On long-term follow-up, the patients with SCAD had a significantly better survival than those with other acute coronary syndromes, but with similar rates of major adverse cardiac events[11]. Current estimates of 1 year and 10 year survival rates are 98.9% and 93.3%, respectively[2]. The estimated 10-year rate of death, heart failure, myocardial infarction or dissection recurrence was 47%[1,11].

Conclusions

SCAD is a rare, life-threatening condition. It affects predominantly women, classically being an important cause of acute coronary syndromes in

young women without traditional cardiovascular risk factors, but it also occurs in older, postmenopausal women. Although the underlying cause is still unknown, the association with pregnancy or postpartum period and fibromuscular dysplasia must be noted. Its presentation may vary from asymptomatic to sudden cardiac death, most cases presenting as acute coronary syndromes. The diagnosis is mainly based on coronary angiography, but intravascular imaging may be needed for confirmation, so it is important to maintain a high index of suspicion in relevant clinical situations. The preferred treatment strategy is based on the clinical presentation and coronarography findings, with the conservative approach being the choice in uncomplicated cases. PCI is the reperfusion strategy of choice, but it is associated with high rates of technical failure and complications, so it must be performed by an experienced operator. If the patient survives the acute phase, the long-term prognosis is excellent, but the risk of recurrent SCAD is significant and close follow-up is needed.

Acknowledgements

We thank all members of the medical team and the patient.

Compliance with Ethics Requirements:

„The authors declare no conflict of interest regarding this article"

„The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from the patient included in the study"

References

1. Douglas PS, Saw J. Spontaneous coronary artery dissection. https://www.uptodate.com (Accessed on November 30, 2017).
2. Wonnacott D, Berringer R. Spontaneous coronary artery dissection. Case report and review of literature. *Canadian Family Physician* 2016;62(12): 994-996.
3. Pretty H. Dissecting aneurysm of coronary artery in woman aged 42: rupture. *BMJ* 1931;1: 667.
4. Shahid A. Spontaneous coronary artery dissection. E-Journal of Cardiology Practice Vol.14, N°38 – 22 Feb 2017. https://www.escardio.org (Accessed November 30, 2017).
5. Alfonso F, Paulo M, Lennie V et al. Spontaneous coronary artery dissection. Long-term follow-up of a large series of patients prospectively managed with a „conservative" therapeutic strategy. *JACC: Cardiovascular Interventions* 2012;5(10): 1062-1070.
6. DeMaio SJ Jr, Kinsella SH, Silverman ME. Clinical course and long-term prognosis of spontaneous coronary artery dissection. *American Journal of Cardiology* 1989;64: 471– 4.
7. Motreff P, Souteyrand G, Dauphin C, Eschalier R, Cassagnes J, Lusson JR. Management of spontaneous coronary artery dissection: review of the literature and discussion based on a series of 12 young women with acute coronary syndrome. *Cardiology* 2010;115: 10–18.
8. Vrints CJ. Spontaneous coronary artery dissection. *Heart* 2010;96: 801–808.
9. Hayes S. Spontaneous Coronary Artery Dissection (SCAD): New insights into this not-so-rare condition. *Texas Heart Institute Journal* 2014;41(3): 295-298.
10. Alfonso F. Spontaneous coronary artery dissection: new insights from the tip of the iceberg? *Circulation* 2012;126: 667-670.
11. Tweet MS, Hayes SN, Pitta SR, et al. Clinical Features, management, and prognosis of spontaneous coronary artery dissection. *Circulation* 2012;126(5): 667-670.
12. Saw J, Aymong E, Sedlak T, et al. Spontaneous coronary artery dissection: association with predisposing arteriopathies and precipitating stressors and cardiovascular outcomes. *Circulation. Cardiovascular interventions* 2014;7(5): 645-655.
13.Tanis W, Stella PR, Pijlman AH, Kirkels JH, Peters RHJ, de Man FH. Spontaneous coronary artery dissection: current insights and therapy. *Netherlands Heart Journal* 2008;16(10):344-349.
14. Dakik HA, Nader GA, Arja WA, Sawaya J, Gharzuddine W. Asymptomatic spontaneous coronary artery dissection. *Clinical Cardiology* 2010;33(7): E40-E42.
15. Saw J. Coronary angiogram classification of spontaneous coronary artery dissection. *Catheterization and cardiovascular interventions* 2014;84(7): 1115-1122.
16. Tweet MS, Eleid MF, Best PJ, et al. Spontaneous coronary artery dissection: revascularization versus conservative therapy. *Circulation. Cardiovascular interventions* 2014;7(6): 777-786.
17. Adlam D, Cuculi F, Lim C, Banning A. Management of spontaneous coronary artery dissection in the primary percutaneous coronary intervention era. *The Journal of Invasive Cardiology* 2010;22(11): 549-553.
18. Jovi Z, Obradovi S, Djeni N, et al. Does thrombolytic therapy harm or help in ST elevation myocardial infarction (STEMI) caused by the spontaneous coronary dissection? *Vojnosanitetski Pregled* 2015;72(6): 536-540.
19. Bergen E, Huffer L, Peele M. Survival after spontaneous coronary artery dissection presenting with ventricular fibrillation arrest. *The Journal of Invasive Cardiology* 2005; 17(10): E4-6.
20. Sarmento-Leite R, Machado PRM, Garcia SL. Spontaneous coronary artery dissection: stent it or wait for healing. *Heart* 2003; 89(2): 164.