CASE REPORT
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Primary percutaneous coronary intervention in a patient with anomalous origin of the left coronary artery from the opposite sinus of Valsalva and left main coronary artery occlusion

Primarna perkutana koronarna intervencija kod bolesnika sa anomalnim ishodištem leve koronarne arterije iz suprotnog sinusa Valsalva-e i okluzijom glavnog stabla leve koronarne arterije

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Abstract

Introduction. Congenital coronary anomalies are detected in about 5% of all performed coronarographies. Coronary artery (CA) anomalies (CAA), considered to be of great risk, are the ones where the CA arises from the opposite sinus (anomalous origination of CA from opposite sinus, ACAOS) of Valsalva. These anomalies are detected in about 1% of cases. This report shows a unique case of a patient with anterior wall ST-elevation myocardial infarction (STEMI) caused by left main CA (LMCA) occlusion, which arose from the right coronary cusp and had an interarterial course, successfully treated with primary percutaneous coronary intervention (PCI). **Case report.** A 46-year-old male patient was admitted to the hospital due to STEMI of the anterior region. On admission, the patient was hypertensive (150/100 mmHg) in sinus rhythm (heart rate 70/min), Killip I. After the initial examination and admitting dual antiplatelet therapy, the patient underwent urgent coronarography. Coronarography was performed using the transradial approach. The right CA had no significant stenosis and was easily cannulated, whereas the left CA could not be cannulated at the usual position. Attempts to cannulate the left CA with multiple catheters of various curves were unsuccessful. The conclusion was that there was a CA anomaly, and the cannulation of the anomalous left CA, which arose from the opposite (right) coronary cusp (anomalous aortic origin of the left CA, AAOLCA), was successfully performed with a Multipurpose catheter. Moreover, the LMCA was occluded in the distal segment. Two drug-eluting stents (DES) were implanted, but the patient developed the no-reflow phenomenon and cardiogenic shock. After the patient was stabilized, computed tomography (CT) coronarography was performed, and AAOLCA with an interarterial course was registered. During the follow-up period, single photon emission computed tomography (SPECT) was performed, and in the staged procedure, a stent was implanted into the proximal circumflex artery using the T and protrusion (TAP) technique. **Conclusion.** Patients with STEMI and the anomalies of CAs are very rare. As such, these patients represent a great challenge for revascularization. Possessing the knowledge of anatomic varieties is paramount when it comes to these patients to treat them adequately with primary PCI.

Key words:
computed tomography angiography; coronary angiography; coronary occlusion; coronary vessel anomalies; percutaneous coronary intervention; sinus of valsalva; st elevation myocardial infarction.

Apstrakt

Uvod. Kongenitalne anomalije koronarnih arterija (KA) otkrivaju se kod oko 5% svih izvedenih koronarografija. Veoma rizičnim se smatraju anomalije kod kojih KA potiču iz suprotnog sinusa (*anomalous origination of coronary artery from opposite sinus,* ACAOS) Valsalva-e i registruju se kod oko 1% slučajeva. Prikazan je jedinstven slučaj bolesnika sa infarktom prednjeg zida miokarda sa ST elevacijom (*ST-elevation myocardial infarction*, STEMI) izazvanog okluzijom glavnog stabla leve koronarne arterije (LKA) porekla iz desnog koronarnog kuspisa sa interarterijskim pravcem pružanja, koji je uspešno lečen primarnom perkutanom koronarnom intervencijom (PKI). **Prikaz bolesnika.** Bolesnik, starosti 46 godina, primljen je kao hitan slučaj zbog kliničkih i elektrokardiografskih znakova za STEMI anteriorne regije**.** Po prijemu je bio hipertenzivan (150/100 mmHg), u sinusom ritmu

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(frekvencija srca 70/min), Killip I. Posle prvog pregleda i uvođenja dvojne antiagregacione terapije bolesnik je podvrgnut urgentnoj koronarografiji. Koronarografija je urađena transradijalnim pristupom. Desna KA koja je bila bez značajnih suženja je lako kanulisana, dok leva koronarna arterija nije mogla da se kanuliše. Pokušano je sa više katetera različitih krivina, ali bez uspeha. Zaključeno je da se radilo o koronarnoj anomaliji i tek sa *Multipurpose* vodič-kateterom uspešno je kanulisana leva KA čije je ishodište bilo iz desnog koronarnog kuspisa (*anomalous aortic origin of the left coronary artery,* AAOLCA). Arterija je bila i okludirana u svom distalnom segmentu. Urađena je implantacija dva stenta obložena lekom, ali je kod bolesnika došlo do zastoja koronarnog protoka i kardiogenog šoka. Nakon stabilizacije stanja bolesnika, izvedena je koronarografija uz pomoć kompjuterizovane tomografije kojom je potvrđen AAOLCA, a registrovano je i da postoji potencijalno rizični interarterijski pravac pružanja leve KA. Nakon pregleda izvršenog tehnikom kompjuterizovane tomografije sa jednom fotonskom emisijom (*single photon emission computed tomography*, SPECT), tokom perioda praćenja, u sledećoj etapi je urađena implantacija stenta u proksimalnu cirkumfleksnu arteriju uz pomoć *T and protrusion* (TAP) tehnike. **Zaključak.** Bolesnici sa STEMI i anomalijama KA se relativno retko sreću i predstavljaju izazov za revaskularizaciju, te je poznavanje anatomskih varijeteta neophodno kako bi ovi bolesnici mogli biti adekvatno tretirani primarnom PKI.

Ključne reči:
angiografija, tomografska, kompjuterizovana; angiografija koronarnih arterija; koronarna okluzija; koronarni krvni sud, anomalije; perkutana koronarna intervencija; sinus valsalvae; infarkt miokarda sa st elevacijom.

Introduction

Congenital coronary artery (CA) anomalies (CAA) are detected in around 5% of all performed coronarographies [1]. In the literature, there are 66 different anomalies described, while the ones considered of great risk are anomalous CAs from the opposite sinus (ACAOS) of Valsalva. These anomalies are detected in around 1% of cases. Anomalous aortic origin of the right CA (AAORCA) arising from the left coronary cusp is detected more often, whereas the anomalous aortic origin of the left CA (AAOLCA) arising from the right coronary cusp is detected in 0.15% of the cases [1, 2]. With both CAA, there are several different courses, such as prepulmonic, retroaortic, subpulmonic (septal), or interarterial courses, where the CA is placed between the aorta and the pulmonary trunk. The interarterial course is the only course that can cause sudden cardiac death and, therefore, is considered malignant [3]. ST-elevation myocardial infarction (STEMI) is rarely detected in patients with CAA and is demanding to treat. The very identification and cannulation of the culprit artery with anomalous origin can be quite challenging.

Herein we presented a unique case of a patient with anterolateral wall STEMI, caused by an occlusion of the left CA arising from the right coronary cusp with interarterial course, successfully treated with the primary percutaneous coronary intervention (PCI).

Case report

A 46-year-old patient was admitted to the hospital due to anterior wall STEMI (Figure 1). Chest pain started 2 hrs prior to admission to the hospital. The patient had no prior medical history, and the only risk factor for CA disease was smoking. On admission, the patient was hypertensive (150/100 mmHg) in sinus rhythm (heart rate 70/min), Killip I. After the initial examination, dual antiplatelet therapy was introduced, and the patient was transferred to the catheterization unit for urgent coronarography.

Coronarography was performed using a transradial approach. The right CA was easily cannulated with diagnostic catheter Tiger (Terumo, Japan), and it had no significant stenosis, whereas the left CA could not be cannulated in the left coronary cusp. Cannulation of the left CA was attempted with multiple catheters, such as EBU of various curves, JL 4.0, and Mach 1™ Amplatz, but these attempts were unsuccessful.

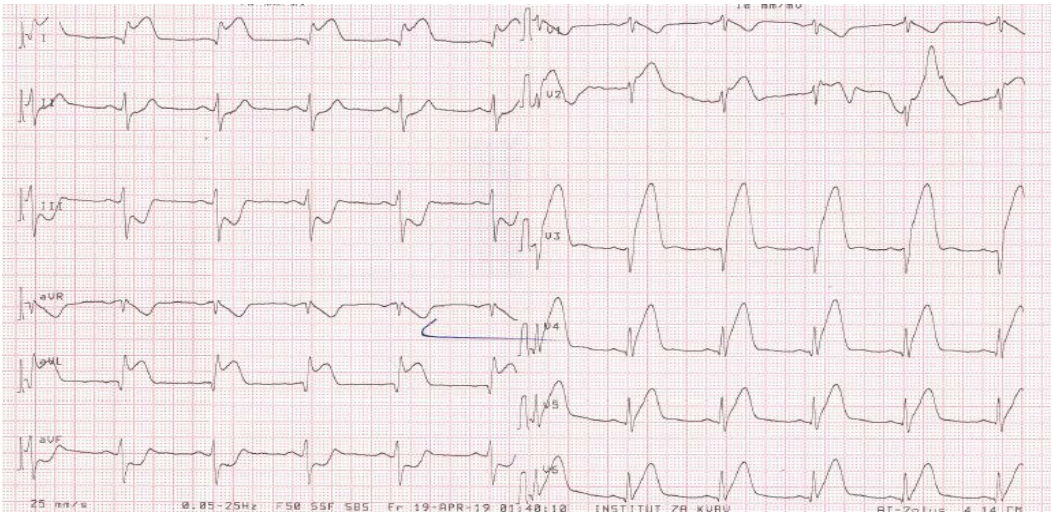


Fig. 1 – Electrocardiographic (ECG) finding of ST-elevation myocardial infarction on admission.

The operator concluded there was an existing CAA, and the cannulation of the AAOLCA from the right coronary cusp, which was occluded in its distal segment, was successfully cannulated with a Mach 1™ Multipurpose catheter (Boston Scientific, USA) (Figure 2A). After passing the guidewire through the occlusion, the antegrade flow was achieved, followed by ventricular fibrillation and prolonged cardiopulmonary resuscitation. Finally, the restoration of spontaneous circulation (ROSC) was established. Due to the progression of heart failure to the state of cardiogenic shock, the patient was intubated and put on mechanical ventilation, and vasopressor support was introduced in the treatment.

Because of the loosing of the position of the guiding catheter and clinical instability, crossover to the right femoral access was performed. This time, the left CA was cannulated with a Mach 1™ JR 4.0 guiding catheter (Boston Scientific, USA). After passing the guidewire, thrombus aspiration was performed due to the high thrombus burden. Subsequently, predilatation was performed, and after that, TIMI 2 flow was established. A trifurcation lesion of the left main CA (LMCA) was detected. The plaque was propagating from distal LMCA to the proximal left anterior descending artery (LAD) and circumflex (Cx), while the gracile Ramus had no significant lesion at the ostia (Figure 2B). Two drug-eluting stents (DES) were implanted, one DES 3.5 × 23 mm (Xience Xpedition, Abbott, USA) from the LMCA to the LAD. The second DES 3.5 × 15 mm (Xience Xpedition, Abbott, USA) was implanted in the LMCA, with a short overlap with the previously implanted stent. Furthermore, a proximal optimization technique (POT) with a semi-compliant balloon, 4.0 × 12 mm (Sprinter, Medtronic, USA), with high-pressure inflation, was performed. After POT, a no-reflow phenomenon developed (Figure 2C), and GP IIb/IIIa antagonist (tirofiban) was admitted intracoronary. Furthermore, an intra-aortic balloon pump (IABP) was implanted (Figure 2D), and the patient was transferred to the coronary care unit (CCU). After 48 hrs in the CCU, the patient was clinically stable, and both vasopressor support and IABP were removed. The patient was extubated after 72 hrs. Echocardiography registered an ejection fraction of 40% with akinesia of the apex and all apical segments of the left ventricle and hypokinesia of the medial anterolateral and inferolateral wall.

On the seventh day of hospitalization, the patient was stabilized and transferred to the ward. Computed tomography (CT) coronarography was performed for detailed analyses of the CAA. CT registered a slit-like AAOLCA from the right

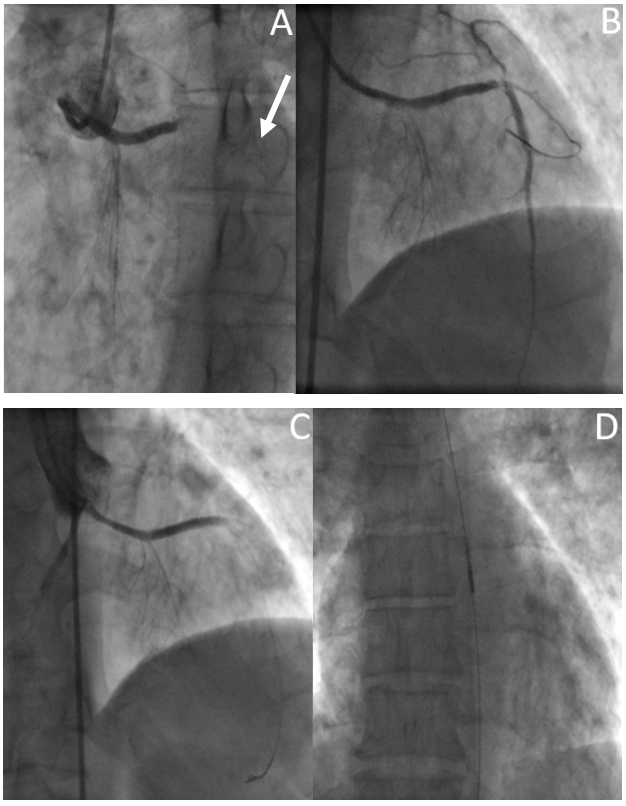


Fig. 2 – A) The arrow points to the occlusion site of the distal segment AAOLCA, LAO 46, CAU 3; B) Anterograde flow is established, and a trifurcation lesion is detected, RAO 2, CRA 26; C) No-reflow at the end of the procedure, RAO 14, CRA 26; D) Positioning of IABP, AP 0. AAOLCA – anomalous aortic origin of the left coronary artery; LAO – left anterior oblique view; CAU – caudal view; RAO – right anterior oblique view; CRA – cranial view; IABP – intra-aortic balloon pump; AP – anteroposterior view.

coronary cusp with the interarterial course (placed between the aorta and pulmonary trunk). Furthermore, the length of the LMCA was 50 mm. Interestingly, the artery at the point of 7.2 mm from the orifice at the aorta enters the heart muscle and passes through up to trifurcation in all its lengths. The stents were patent, and at the Cx ostia, a significant lesion was registered (Figure 3). On the fourteenth day of the hospitalization, the patient was discharged in good general condition. During the six-month follow-up period, the patient had symptoms of stable angina. Single photon emission computed tomography (SPECT) was performed, and it showed significant ischemia in the irrigational area of Cx.

Ten months after the STEMI, repeat coronorography was performed and, this time, a left Amplatz 1 guiding catheter was used, and successful cannulation of LCA and RCA was achieved. Coronography registered significant stenosis of the ostial Cx, and the previously implanted stents were patent. As a result, DES 2.5 × 15 mm (Xience Pro, Abbott, USA) was implanted on the ostium of Cx with the T and protrusion (TAP) technique (Figure 4).

Discussion

The report presented a case of a STEMI patient with AAOLCA from the right coronary cusp with an interarterial course. Besides, the patient had LMCA occlusion in the distal segment. To the best of our knowledge, this is a unique case in the literature.

The usage of noninvasive technologies, such as CT and MRI, enables detecting and registering coronary anomalies

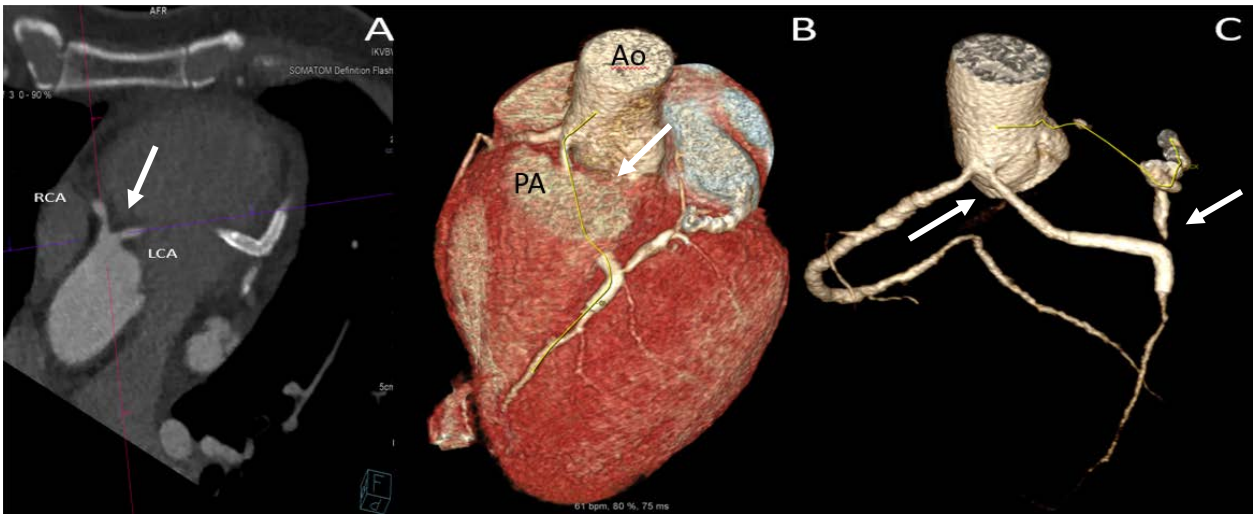


Fig. 3 – A) The arrow points to the anomalous origin of LCA from the opposite sinus of Valsalva; B) 3D reconstruction registers the interarterial course of AAOLCA, the arrow points to the part of the artery which is inside of the muscle (Ao – aorta, PA – pulmonary artery); C) The left arrow points to the slit-like orifice of AAOLCA, the right arrow points to significant stenosis of ostial circumflex (Cx). LCA – left coronary artery; AAOLCA – anomalous aortic origin of the left coronary artery.

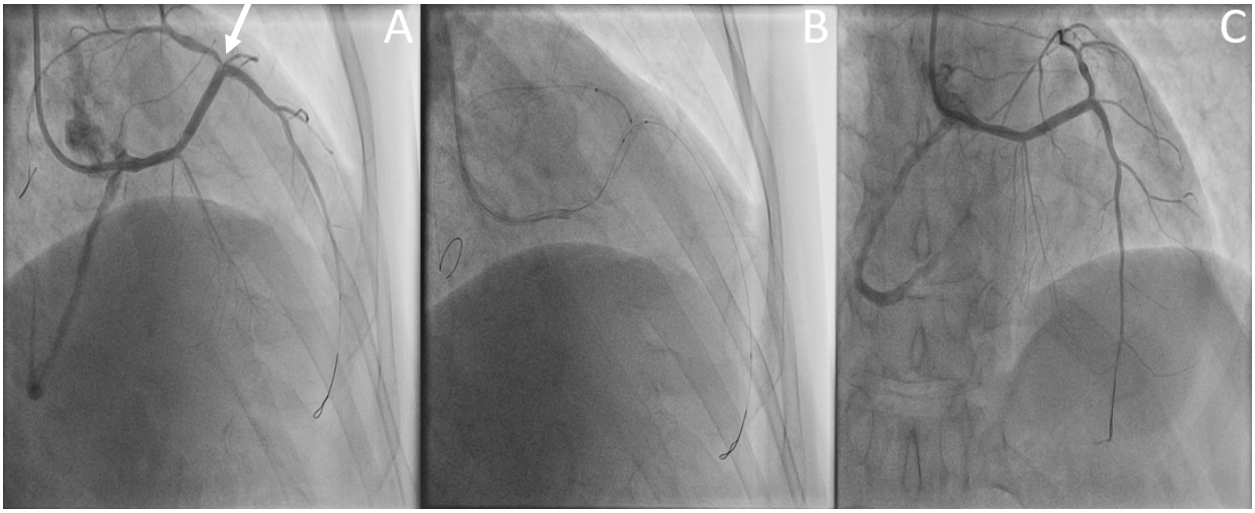


Fig. 4 – A) The arrow points to a lesion at the circumflex (Cx) ostia RAO 46, CRA 30; B) T and protrusion (TAP) technique, DES 2.5 × 15 mm in Cx and NC balloon, 5 × 12 mm in LMCA, LAD 3, RAO 46, CRA 30; C) Final result, LAO 7, CRA 25. RAO – right anterior oblique view; CRA – cranial view; DES – drug-eluting stent; NC – non-compliant; LMCA – left main coronary artery; LAD – left anterior descending artery; LAO – left anterior oblique view.

more frequently [3, 4]. The detection of CAA deserves special attention. Even though most of these anomalies are benign, CA arising from the opposite sinus of Valsalva with interarterial course presents a risk of sudden cardiac death. It can be caused by many various factors, such as a slit-like orifice or tangential passage of origin, or it might result from extreme physical activity, which can lead to the compression of the anomalous coronary artery with an interarterial course. Extreme physical activity can result in increased blood flow through the aorta and pulmonary artery leading to compression, which causes ischemia. It is crucial to emphasize that these anomalies present a great risk for the younger population, which is exposed to extreme physical activity. That mostly refers to sportsmen, athletes, and military recruits. Sometimes, the first manifestation can be sudden cardiac death (SCD); however, very often, there are symptoms present during physical activity, such as angina-like symptoms, arrhythmia, presyncope, and syncope [5, 6].

The literature provides us with a different frequency of atherosclerotic disease in anomalous CAs compared to normal. Recent studies indicate a slightly higher incidence of CA disease (CAD) in anomalous CAs than what was shown in earlier studies which indicated an equal incidence of CAD [7]. However, STEMI patients with coronary anomalies are quite rare. According to the research conducted by Marchesini et al. [8], only 5 out of 1,015 STEMI patients (0.4%) had an anomaly of the CA at the same time.

Performing primary PCI in STEMI patients and CAA is challenging for every operator, mostly because of the cannulation problems of the anomalous culprit artery. However, a problem might arise during the procedure in terms of balloon and stent deliverability in the culprit lesion area. In this case report, the cannulation of the LCA presented a problem, considering that it could not be detected in the left coronary cusp. After exchanging multiple catheters, the operator decided to use a Multipurpose catheter and search for the left CA in the right coronary and posterior cusp. Luckily, in this case, only the cannulation was challenging, considering the anomalous origin. In both procedures, there was no problem with the device deliverability.

The occlusion of the LMCA is a disastrous event, and most patients have a fatal outcome on the way to the hospital. De Luca et al. [9] registered an incidence of 0.8% in patients with myocardial infarction and LMCA occlusion. Their study showed high intrahospital mortality, which was 58% of all patients and 80% of patients who developed cardiogenic shock or had no-reflow at the end of the procedure. Certain factors indicated that they contribute to the higher survival rate of these patients, such as dominant RCA, the existence of hetero-collateral circulation, and fast revascularization [10–12]. In our case, RCA was dominant, and hetero-collateral showed the very periphery of LAD. However, the existence of hetero-collateral circulation did not imply the anomalous origin of LCA. The operator's experience in assuming the presence of coronary anomaly and the adequate selection of catheter allowed successful revascularization of LMCA and proximal LAD.

Despite the loss of around 15 minutes until the cannulation of anomalous LCA, guidewire passage, and establishing anterograde flow through the infarction artery, cardiogenic shock as well as no-reflow at the end of the procedure developed. No-reflow developed as a consequence of a high thrombus burden and aggressive post-dilatation during POT, which led to distal embolization with thrombus masses and debris from the lesion, causing a microvascular obstruction. Treatment of no-reflow was challenging because nitroglycerin, adenosine, or intracoronary adrenalin could not be given due to hemodynamic and rhythmic instability. The decision to administer GP IIb/IIIa antagonist (tirofiban) with implantation of mechanical circulatory support with IABP and aggressive treatment of cardiogenic shock with the support of invasive mechanical ventilation led to fast stabilization. It is important to emphasize that the veno-arterial extracorporeal membrane oxygenation (VA-ECMO) was considered. However, due to the no-reflow and admission of GP IIb/IIIa antagonist, there was a high risk of bleeding; thus, it was decided to wait and monitor the patient's status. As satisfactory hemodynamical stabilization was achieved promptly during the treatment at the CCU, the implantation was no longer needed.

Since 3D reconstruction is not possible, angiography has a limited sensitivity when it comes to adequate diagnostics of anomalies of CAs. Therefore, to diagnose the type of anomalies, their course, and their placement in relation to great vessels, it is necessary to use multi-slice computed tomography or magnetic resonance imaging. Applying these methods prior to coronarography can facilitate the cannulation of the anomalous CA, and this option is manageable in elective and stable patients [13].

A surgical approach to AAOLCA and AAORCA anomalies is considered in patients under 35 years of age, who were exposed to high physical efforts, and who had experienced some of the symptoms which led to the diagnosis of this type of anomaly. When it comes to older patients, the type of treatment should be carefully considered, the need for surgical intervention in particular. Whether these patients are asymptomatic is also of great importance. Tests for determining ischemia are advised in these patients [14]. Surgery treatment was not taken into consideration for the patient presented in this case report since the patient did not have any symptoms before the coronary incident and was not exposed to great physical efforts. SPECT showed ischemia only in the Cx area, which was sub-occluded at the origin and was revascularized by stent implantation.

Conclusion

STEMI patients with CAA are rare and represent a challenge for revascularization; thus, knowing the anatomical varieties is essential to treating these patients with PCI.

Conflict of interest

The authors declare no conflict of interest.

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