Coronary CT Angiography Findings in Non-atherosclerotic Coronary Artery Diseases

Z. Zhou [a], L. Xu[a,*], N. Zhang [a], H. Wang [a], W. Liu[b], Z. Sun [c], Z. Fan[a]
a Department of Radiology, Beijing Anzhen Hospital, Capital Medical University, No. Anzhen Rd, Chaoyang District, Beijing, 100029, China
b Department of Cardiology, Beijing Anzhen Hospital, Capital Medical University, No. Anzhen Rd, Chaoyang District, Beijing, 100029, China
c Department of Medical Radiation Sciences, Curtin University, Perth, WA 6845, Australia

1. Introduction

Although acute coronary events mainly result from atherosclerotic lesions in coronary arteries, some cases of myocardial ischemia or infarction may be related to non-atherosclerotic causes, such as congenital abnormalities, vasculitis, and dissection[1].Invasive coronary angiography (ICA) is the established gold standard for the evaluation of coronary artery diseases. However, in recent years, coronary CT angiography (CCTA) has become a non-invasive alternative for assessment of coronary arteries with a lower referral threshold than ICA. Morphological features of coronary diseases can be demonstrated more precisely by CCTA. Currently, most of the studies focus on CCTA in coronary artery disease while very little research is available in the diagnostic assessment of non-atherosclerotic coronary artery diseases by CCTA. This article aims to provide an overview of CCTA findings in non-atherosclerotic coronary artery diseases.

2. Congenital Coronary Abnormalities

Coronary artery anomalies have been reported in approximately 1% among patients undergoing cardiac catheterization, and 0.29% among autopsy specimens[2]. Coronary anomalies comprise a variety of malformations, some of them are benign course with no symptom, and the others related to symptoms such as chest pain or sudden death. Such anomalies may be classified as follows: 1) anomalies of origin and course; 2) anomalies of intrinsic coronary anatomy; 3) congenital fistulas [3]. CCTA is regarded as the first choice for the initial screening of congenital coronary anomalies as it enables excellent visualization of the course of the coronary arteries. The anomalous origin can be classified into four separate subtypes, which are detailed in the following sections.

2.1 Coronary Ostium in Improper Coronary Sinus

Anomalous origin of the coronary artery from the opposite sinus of Valsalva (ACAOS) with an interarterial course is a congenital malformation that might have significant clinical impact[4]. Although ACAOS is often asymptomatic, patients may present with myocardial ischemia or sudden death without symptoms, particularly in patients with left ACAOS2. Potential pathogenesis of stenosis that induces the aforementioned symptoms includes the acute angle of takeoff and kinking of the anomalous coronary artery, mechanical compression of the anomalous coronary artery when it take course within the aortic wall, and spasm of the anomalous coronary artery arising from endothelial injury[4].

Left ACAOS may be further classified into four separate subtypes: 1) The left main coronary artery (LMCA) courses between pulmonary artery and aorta (interarterial) (Fig. 1). 2) The LMCA tracks anteriorly over the right ventricular outflow tract (pre-pulmonic). 3) The LMCA takes course intramyocardially before resurfacing at the proximal portion of the interventricular groove. 4) The LMCA courses posteriorly around the aortic root (retroaortic). CCTA shows that the entire coronary arteries arise from a single ostium in the right sinus of Valsalva. The RCA continues in normal position[5].Right ACAOS may have interarterial, retro-aortic, prepulmonic or septal (subpulmonic) course, the most common being inter-arterial. CCTA shows that both RCA and LCA originate from left coronary sinus. RCA has a slit-like ostium and courses between aorta and pulmonary artery[6].

2.2 Coronary Ostium Outside Aortic Coronary Sinus

Anomalous origin of LCA or RCA from the pulmonary artery (ALCAPA or ARCAPA) is a rare entire affecting 1/300,000 live births. Left-to-right shunt results in coronary steal, and may lead to silent myocardial ischemia, ventricular dysfunction, even sudden cardiac death. Due to its unfavorable prognosis, it is necessary to detect it early before adverse cardiac events occur[7].

CCTA shows that diffusely enlarged and tortuous coronary arteries with RCA or LMCA implantation site on the pulmonary artery (PA) (Fig.2). And sometimes there is a normal origin of the opposite coronary artery with diffuse enlargement and tortuosity.

2.3 Single Coronary Artery

The single coronary artery is defined as only one coronary artery arising with one ostium from the aortic trunk. People with a singlecoronary artery may have a normal life expectancy; however, some may be at a risk for sudden death[8].

CCTA shows the entire coronary circulation arises from the left or right Valsalva sinus with a single ostium. After a common main stem, the artery divides into a dominant LCA or RCA, and finally bifurcates into other branches (Fig. 3).

2.4 Anomalous Location of the Coronary Ostium in the Aortic Root: High, Low or Commissural

Coronary arteries commonly arise from the aortic sinuses rather than from the tubular aorta. However, the coronary ostium may be located above the sinotubular junction, usually by only a few millimeters. Very high takeoff of the coronary ostium occurs in less than 1% of the population[9]. CCTA is useful to define the ostium and the course of the artery, which reveals a high takeoff of the left or right coronary artery from above the coronary sinus with the ostium more than 1 or 2 cm above the aortic valve (Fig. 4) .

2.5 Congenital Fistulas

Coronary artery fistula (CAF) is a connection between the coronary artery and cardiac chamber or great vessel, which bypasses the myocardial capillary bed[10]. CAF is often asymptomatic during childhood but is usually symptomatic in adult patients., The most common findings for symptomatic patients include heart failure resulting from left to right shunting, coronary steal induced ischemia, fistula rupture or thrombosis[11]. CAFs can be classified into 5 types: type 1 coronary artery-cameral fistula, type 2 coronary－pulmonary artery fistula, type 3 coronary - coronary sinus fistula, type 4 coronary arteriovenous fistula, and type 5 bilateral coronary artery fistula.

The most common origin site of fistulas lies in the RCA in about 55% of the patients and about 35% from the LCA. Right ventricle is the most frequent draining chamber in about 45% of the cases, right atrium in about 25% and the pulmonary artery in 15 - 20% with the remaining 7% of the cases drain into the coronary sinus. Left cardiac chambers are the most infrequent draining site[12]. CCTA is superior to ICA in the assessment of exact dimensions, feeding artery, draining site and its anatomic relationships may clearly be defined. CT findings are variable depending on the type of fistula, site of the shunt, shunt volume, and presence of concomitant cardiac abnormalities. The dilation and tortuosity of the fistula may vary (Fig. 5).

3. Coronary Arteritis

Vasculitides are defined as conditions by the presence of inflammation of the vessel wall, which may progressively alter the lumen, resulting in lumen stenosis, occlusion, or aneurysmal dilation. The diseases can be classified into infectious and non-infectious vasculitides[13]. Coronary arteritis is typically associated with polyarteritis nodosa, Kawasaki disease, and Takayasu's arteritis[14]. CCTA is emerging as a reliable examination tool in non-invasive depiction of both luminal and mural lesions in the aorta and its main branches, which may facilitate the detection of early phase of coronary arteritis(Fig. 6).

3.1 Takayasu's Arteritis (TA)

TA is an idiopathic vasculitis characterized by the involvement of the aorta and its main branches. Involvement of coronary artery is found in approximately 10 to 30% of the cases with TA. Three types of pathological features in coronary artery may be observed: type 1, stenosis or occlusion in the proximal segments of the coronary arteries; type 2, diffuse or focal coronary arteritis, which may extend diffusely or may involve focally, so-called skip lesions; and type 3, coronary aneurysm[15]. CCTA demonstrates ostial occlusion of the left main coronary artery and stenosis or occlusion of proximal segments of the coronary arteries, diffuse or focal coronary arteritis and aneurysms of the coronary arteries (most rare)[1] (Fig. 7).

3.2 Behçet's Disease (BD)

BD is a chronic, multisystemic disorder characterized by the inflammation of any size of the blood vessel. The prevalence of arterial occlusion in BD ranges from 0.5 to 1.5% and can involve the coronary, carotid, subclavian , pulmonary, renal, or any peripheral arteries. Coronary artery involvement is extremely uncommon in BD. Coronary lesions can be either occlusive or aneurysmal change, but coronary aneurysms are more frequently seen than stenotic lesions or occlusion induced by adherent thrombus[16]. CCTA shows that the most common lesions are ostial stenosis, non-ostial stenosis or occlusion, and coronary aneurysms[13].

3.3 Coronary Periarteritis

Coronary periarteritis is a rare disease characterized by heavy infiltration of lymphocytes, plasma cells, and neutrophils with destroyed media and marked intimal fibrosis; partly organized thrombus can occur which may obstruct the lumen[17]. This process may further induce chronic inflammation and remodeling, resulting in aneurysm formation and other morphologic changes[18]. CCTA shows that 1) one or more localized or diffused homogeneous nodular lesions, or pseudotumors, along the coronary artery walls or 2) rings of soft-tissue attenuation surrounding the coronary arteries like "Pigs-in-a-blanket" (Fig. 8).

4. Coronary Artery Aneurysm

Coronary artery aneurysm (CAA) is defined as the dilatation of coronary arteries which exceeds the diameter of adjacent normal artery segments or the diameter of the largest coronary artery by 1.5 times. Aneurysms can be classified into true aneurysms (affecting the three arterial linings) and false aneurysms (the arterial wall is damaged, often as a result of injury or iatrogenic cause)[19]. Atherosclerosis is the main cause of the cases of CAA. Other etiologies include congenital abnormality, inflammatory conditions and drug-eluting stents implantation[20].

The RCA was the most commonly affected branch (40-87% of aneurysms), followed by the LCx or LAD. Although most patients are asymptomatic, CAA may lead to myocardial ischemia, myocardial infarction, or sudden death[21]. The main complications include coronary artery rupture, coronary thromboembolism and compression by large CAA, which may lead to serious hemodynamic changes[22].

4.1 Kawasaki Disease

Kawasaki disease (KD), also known as acute febrile mucocutaneous lymph node syndrome, was first reported by Kawasaki in 1967. The disease affects mainly children under age 4 with unknown cause. KD usually affects the RCA and proximal LAD, main stem, more rarely the LCx, and the distal coronary segments[19]. The systemic vasculitis results in formation of CAA[23]. Coronary artery involvement results in much of KD's mortality and morbidity due to myocardial infarction and heart failure[24]. CCTA can accurately detect not only the location, shape, and dimension of the aneurysms but also enable visualization of coronary wall and the presence of luminal stenosis, as well as thrombosis with clinical impact in therapeutic decision-making[21] (Fig. 9) .

4.2 Mycotic Aneurysm

Mycotic aneurysms are very rare accounting for less than 3% of coronary aneurysms. They may result from a complication of infectious endocarditis, or sepsis particularly in immunosuppressed patients. They are subjected to coronary artery bypass grafting (CABG) and repeated catheterizations causing trauma with inflammation of the wall. The LAD and LCx are most commonly involved. Main stem involvement appears to be rare[19]. CCTA can show alterations in both the wall and the lumen of involved artery, and it can demonstrate ectasia or aneurysms affecting the proximal-mid left or right coronary artery.

4.3 Pseudoaneurysm

Pseudoaneurysms of the coronary arteries are most commonly post-traumatic in nature and may result from disruption of the vessel wall including external elastic membrane or transition from a three-layered wall to monolayer outward bulging. The incidence ranges from 0.02% to 0.04% in general and less than 5% found in angiography[22] .On CCTA, a pseudoaneurysm is typically diagnosed as a large, narrow-necked, thin-walled, saccular lesion communicating with the real arterial lumen through the ruptured arterial wall (Fig. 10). Besides arterial wall visualization, CCTA is able to identify the length of lesion, severity of stenosis, the presence or absence of thrombosis, and the distal vessel.

5. Coronary Artery Dissection and Intramural Hematoma

5.1 Spontaneous Coronary Artery Dissection (SCAD)

SCAD is a rare cause of acute myocardial ischemia that commonly results in sudden cardiac death. The reported incidence of SCAD was between 0.1% and 0.28%

in angiographic population[25]. The LAD is most commonly affected, followed by the RCA[19]. The disease has been broadly classified into atherosclerotic and non-atherosclerotic in origin. Non-atherosclerotic causes include the peripartum state, systemic inflammatory conditions or idiopathic[26]. SCAD may also occur either by rupture of a plaque or an intramural hematoma secondary to hemorrhage of vasa vasorum[19]. Its presentation may vary from mild ischemia symptoms to myocardial infarction or even sudden cardiac death.

The diagnosis of SCAD can be made by excluding the more common secondary causes of coronary artery dissection such as blunt chest trauma and coronary interventions. CCTA shows that 1) coronary artery dissection resulting from an intimal tear with medial dissection and false lumen formation, the true and false lumens are separated by the intimal flap; 2) coronary artery dissection resulting in medial dissection and formation of intramural hematoma without intimal tear[25].

5.2 Spontaneous Coronary Intramural Hematoma

Spontaneous coronary intramural hematoma is a subset of SCAD. Hemorrhage of vasa vasorum within the media may lead to the separation of mural layers. The disease usually occurs in young women, particularly in the peri- and postpartum period and with use of oral contraceptive[27]. The symptoms may vary from acute chest pain to cardiogenic shock or sudden death. CCTA shows that the semilunar thickening of the coronary arterial wall, the formation of hematoma is limited to the medial-adventitial layers, with no intimal flaps visualized (Fig. 11).

6. Iatrogenic and Traumatic Abnormalities

Iatrogenic and traumatic coronary artery abnormalities are a rare but devastating condition[28]. This category occurs after endovascular coronary intervention or secondary to thoracic trauma, endomyocardial biopsy, or cardiac surgery. CCTA enables a detailed anatomical study, and therefore plays an important role in the pre-treatment assessment (Fig. 12).

6.1 Iatrogenic Coronary Dissection

Predisposing conditions for iatrogenic coronary dissection may include aortic root calcification, hypertension, aging process and recent myocardial infarction. Once the coronary dissection occurs, it may rapidly propagate in a few minutes[28]. CCTA can demonstrate the dissection into the media with true and false lumen formation without reentry, and reveal a significant stenosis or obstruction of the involved LCA or RCA.

6.2 Iatrogenic Coronary Fistulas

The findings of acquired fistulas after operation or certain medical conditions have been reported in several cases, including mitral valve replacements, percutaneous coronary interventions, CABG and acute myocardial infarctions. In such condition, the sudden postoperative continuous murmur is indicative of the formation of iatrogenic fistula[29]. CCTA can show abnormal connections between the coronary arteries and a cardiac chamber or great vessel after operations or certain medical conditions.

7. Conclusion

CCTA is reliable for coronary artery visualization including assessment of its origin and course, lumen, vessel wall, and adjacent structures. This article highlights the classic imaging signs of non-atherosclerotic coronary artery diseases. Understanding of CCTA findings in these non-atherosclerotic coronary artery diseases will assist timely detection and diagnosis of these pathologies.

Conflicts of interest

None.

References:

1. Araszkiewicz A, Prech M, Hrycaj P, Lesiak M, Grajek S, Cieslinski A. Acute myocardial infarction and rapid development of coronary aneurysms in a young woman - unusual presentation of Takayasu arteritis? *Can J Cardiol* 2007;23（23）:61-63.

2. Lee BY. Anomalous right coronary artery from the left coronary sinus with an interarterial course: Is It Really Dangerous? *Korean Circ J* 2009;39（5）:175-179.

3. Neves PO, Andrade J, Monção H. Coronary anomalies: what the radiologist should know. *Radiol Bras* 2015;48（4）:233-241.

4. Yildiz O, Karabay KO, Akman C, Aytekin V. Anomalous origin of the left main coronary artery from the right coronary artery with a preaortic course. *Tex Heart Inst J* 2015;42（3）:243-245.

5. Marler AT, Malik JA, Slim AM. Anomalous left main coronary artery: Case series of different courses and literature review. *Case Rep Vasc Med* 2013;2013（2013）:380952.

6. Satija B, Sanyal K, Katyayni K. Malignant anomalous right coronary artery detected by multidetector row computed tomography coronary angiography. J Cardiovasc Dis Res 2012;3（1）:40-42.

7. Secinaro A1, Ntsinjana H, Tann O, et al. Cardiovascular magnetic resonance findings in repaired anomalous left coronary artery to pulmonary artery connection (ALCAPA). *J Cardiovasc Magn Reson* 2011;13（1）:27.

8. Jia EZ, Shan QJ, Yang ZJ, et al. Coronary arterial spasm in single right coronary artery. *J Zhejiang Univ Sci B* 2009;10（11）:829-832.

9. Rosenthal RL, Carrothers IA, Schussler JM. Benign or malignant anomaly? Very high takeoff of the left main coronary artery above the left coronary sinus. *Tex Heart Inst J* 2012;39（4）:538-541.

10. Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: Clinical and therapeutic considerations. *Int J Cardiol* 2006;107（1）:7-10.

11. Zhang ZG, Xu XD, Bai Y, et al. Transcatheter closure of medium and large congenital coronary artery fistula using wire-maintaining technique. *J Cardiol* 2015;66（6）:509-513.

12. Lalitha Rudraiah, Gaurav Dhar, Deepak Thatai. Acquired coronary cameral fistula — A report of two cases. *Int J Cardiol* 2008;123（2）:40-42.

13. Enrico Ammirati, Francesco Moroni, Patrizia Pedrotti, et al. Non-invasive imaging of vascular inflammation. *Front Immunol* 2014;5（5）:399-404.

14. Mednick Z, Farmer J, Khan Z, Warder D, Ten Hove M. Coronary arteritis: An entity to be considered in giant cell arteritis. *Can J Ophthalmol* 2016;51（1）:6-8.

15. Wang EL, Sato Y, Takeichi T, Kitamura O. Sudden death of an infant with coronary involvement due to Takayasu arteritis. *Cardiovasc Pathol* 2013;22（1）:109-111.

16. Sonia H, Khaldoun BH, Sylvia M, Faouzi M, Habib G, Mohamed BF. Stenosis and aneurysm of coronary arteries in a patient with Behcet’s disease. *Open Cardiovasc Med J* 2008;2（1）:118-120.

17. Kohout A, Steiner I, Zákravská N. Coronary arteritis with marked fibrous periarteritis: Case report. *Cardiovasc Pathol* 2000;9（5）:297-299.

18. Urabe Y, Fujii T, Kurushima S, Tsujiyama S, Kihara Y. Pigs-in-a-blanket coronary arteries: a case of immunoglobulin G4-related coronary periarteritis assessed by computed tomography coronary angiography, intravascular ultrasound, and positron emission tomography. *Circ Cardiovasc Imaging* 2012;5（5）:685-687.

19. Dehaene A, Jacquier A, Falque C, Gorincour G, Gaubert JY. Imaging of acquired coronary diseases: From children to adults. Diagn Interv Imaging 2016;97（5）:571-580.

20. TK Mishra，SN Routray，B Das，C Satpathy，CK Mishra. Multivessel giant coronary aneurysms: Case report and literature review. *J Indian Coll Cardiol* 2012;2（2）:83-86.

21. De Agustin JA, Gomez de Diego JJ, Rodrigo JL, et al. Right coronary artery aneurysm due to Kawasaki disease: A comprehensive assessment by multislice computed tomography. *Int J Cardiol* 2014;173（2）:12-13.

22. Wang L, Wang J, Cheng TO, et al. Giant left coronary artery aneurysms: Review of the literature and report of a rare case diagnosed by transthoracic echocardiography. *Int J Cardiol* 2015;189（1）:267-271.

23. Baraona F, Valente AM, Porayette P, Pluchinotta FR, Sanders SP. Coronary arteries in childhood heart disease: Implications for management of young adults. *J Clin Exp Cardiolog* 2012;1(8): 006.

24. Subha V Raman, Ashish Aneja, Wael N Jarjour. CMR in inflammatory vasculitis. J Cardiovasc Magn Reson 2012;14（1）:82.

25. Aqe RA, Zoghbi GJ, Iskandrian A. Spontaneous coronary artery dissection, aneurysms, and pseudoaneurysms: A review. *Echocardiography* 2004;21（2）:175-182.

26. Jacqueline Saw. Spontaneous Coronary Artery Dissection. *Can J Cardiol* 2013;29（29）:1027-1033.

27. Antonsen L, Thayssen P, Jensen L. Large coronary intramural hematomas: a case series and focused literature review. *Cardiovasc Revasc Med* 2015;16（2）:116-123.

28. Sarkis A, Maaliki S, Haddad A, Hatem J, Ghanem G. An unusual complication of coronary angiography: Bidirectional dissection of the right coronary artery and the ascending aorta. *Int J Cardiol* 2009;132（132）:20-22.

29. Marios Loukas, Ashley St. Germain, Abigail Gabriel, Alana John, R. Shane Tubbs, Diane Spicer. Coronary artery fistula: a review. *Cardiovasc Pathol* 2015;24（3）:141-148.

Figures Legend

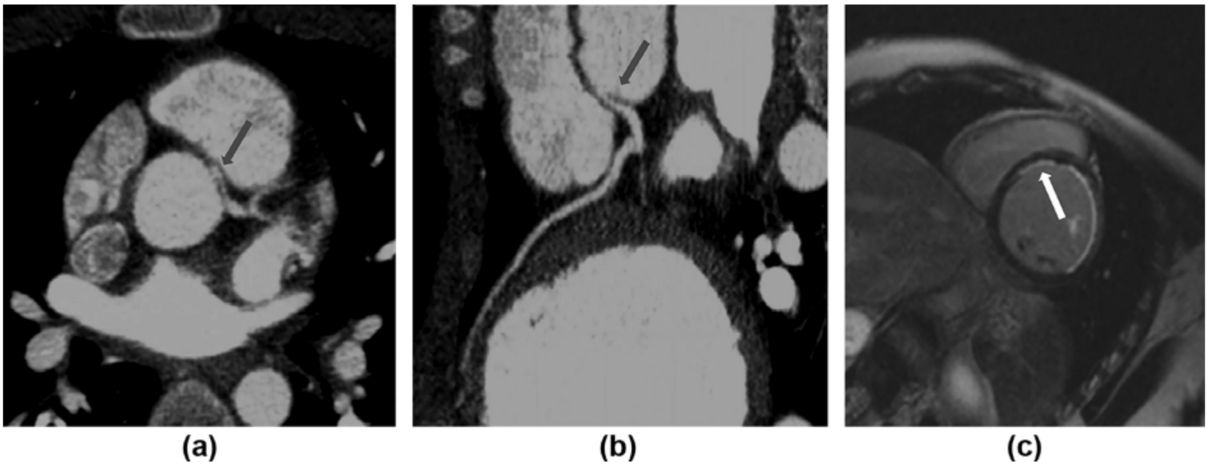


Fig. 1 — 12 -year-old boy with syncope after physical exercise, left coronary artery originating from the right coronary sinus. Axial (A) and CPR (B) images show anomalous origin of the left main coronary artery from the right coronary sinus with an interarterial course (*gray arrow*). MR delayed enhancement shows subendocardial infarction in the left coronary artery territory (C) (*white arrow*).

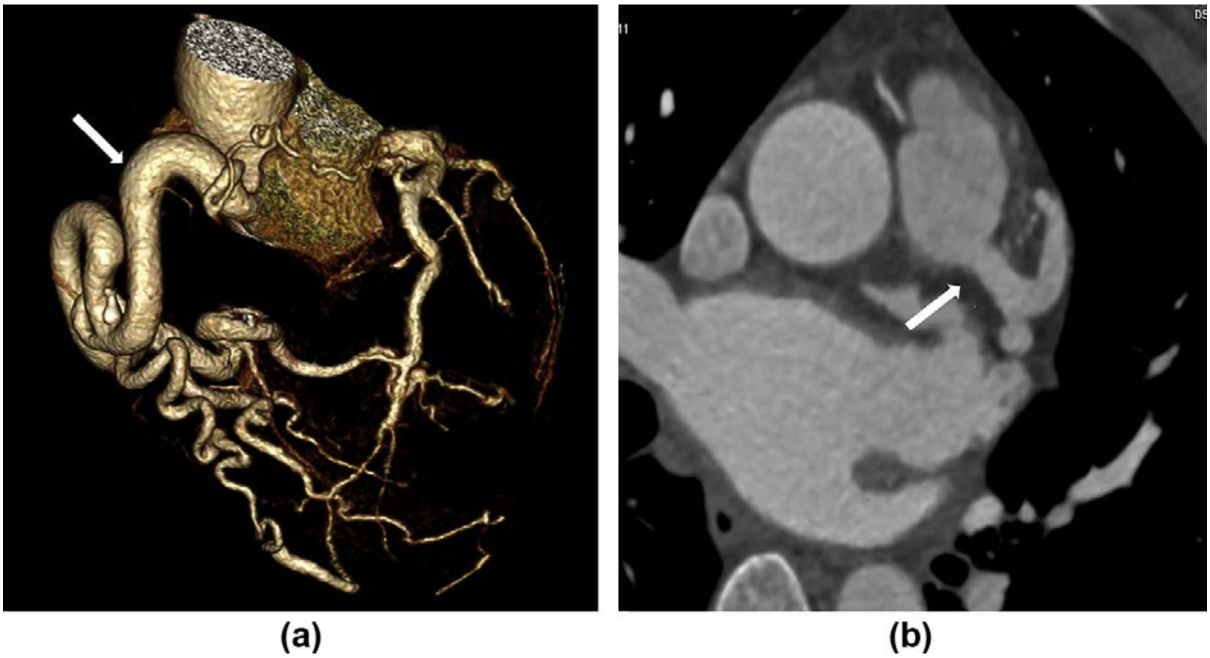


Fig. 2 — 45-year-old female with anomalous origin of left coronary artery from the pulmonary artery. Volume rendering image shows diffusely enlarged and tortuous right coronary artery (A) (*arrow*), anomalous origin of left main coronary artery from the pulmonary artery (B) (*arrow*).

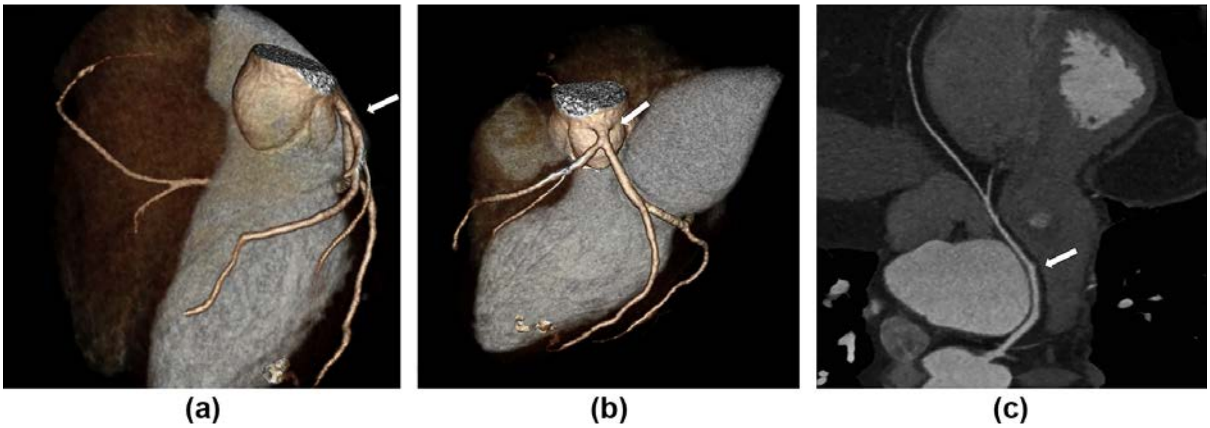


Fig. 3 — 74 -year-old female with single coronary artery. Volume rendering image (A), (B) shows only one coronary artery arising from the left coronary sinus (*arrow*), the left circumflex artery take course to the right atrioventricular groove.

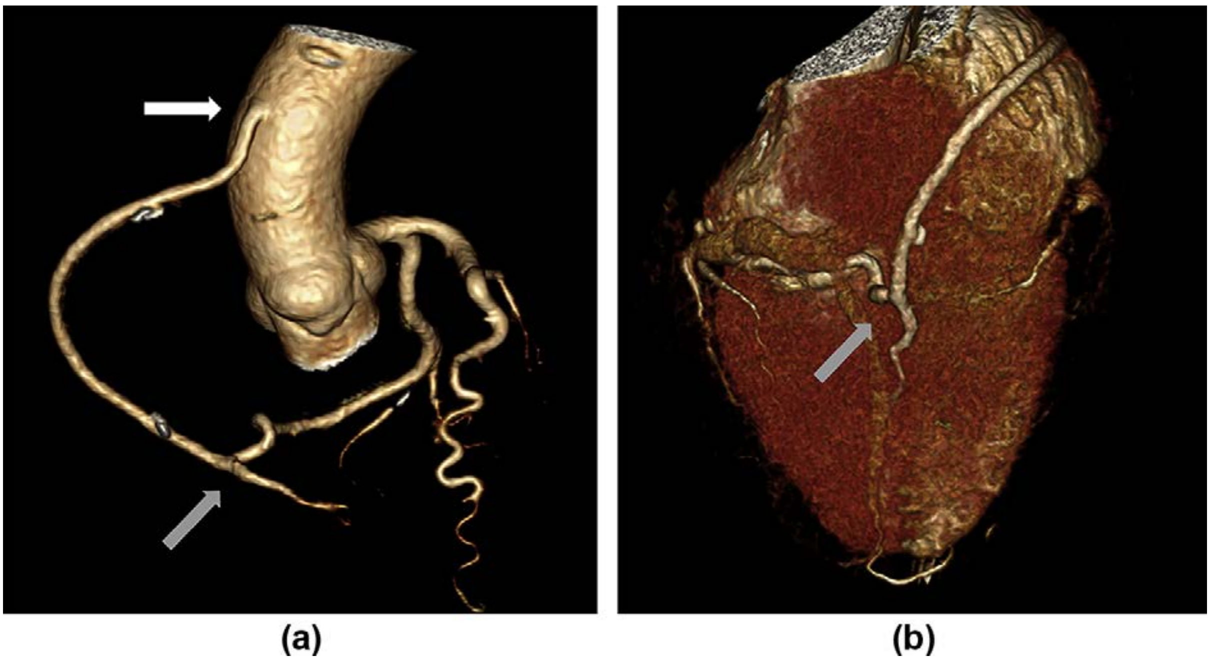
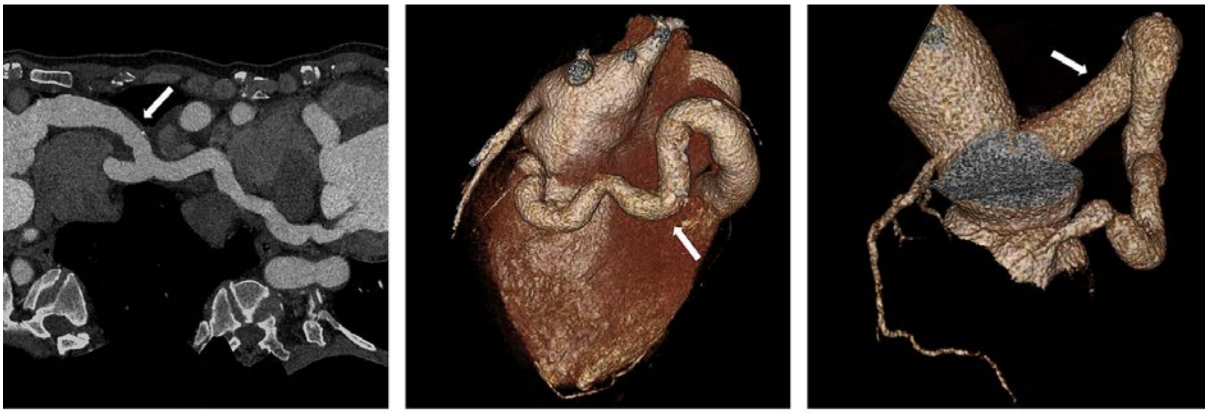


Fig. 4 — 47-year-old male with high-takeoff right coronary artery and direct communication between the right coronary artery and the left circumflex artery. Volume rendering images show a high-takeoff right coronary artery (*white arrow*) with a very rare coronary anomaly circulation: direct communication between the right coronary artery and the left circumflex artery (*yellow arrow*).



(a) (b) (c)

Fig. 5 —53-year-old male with coronary-cameral fistula. Volume rendering images (A, B) show diffusely enlarged and tortuous right coronary artery draining to the left ventricle (*arrow*) .

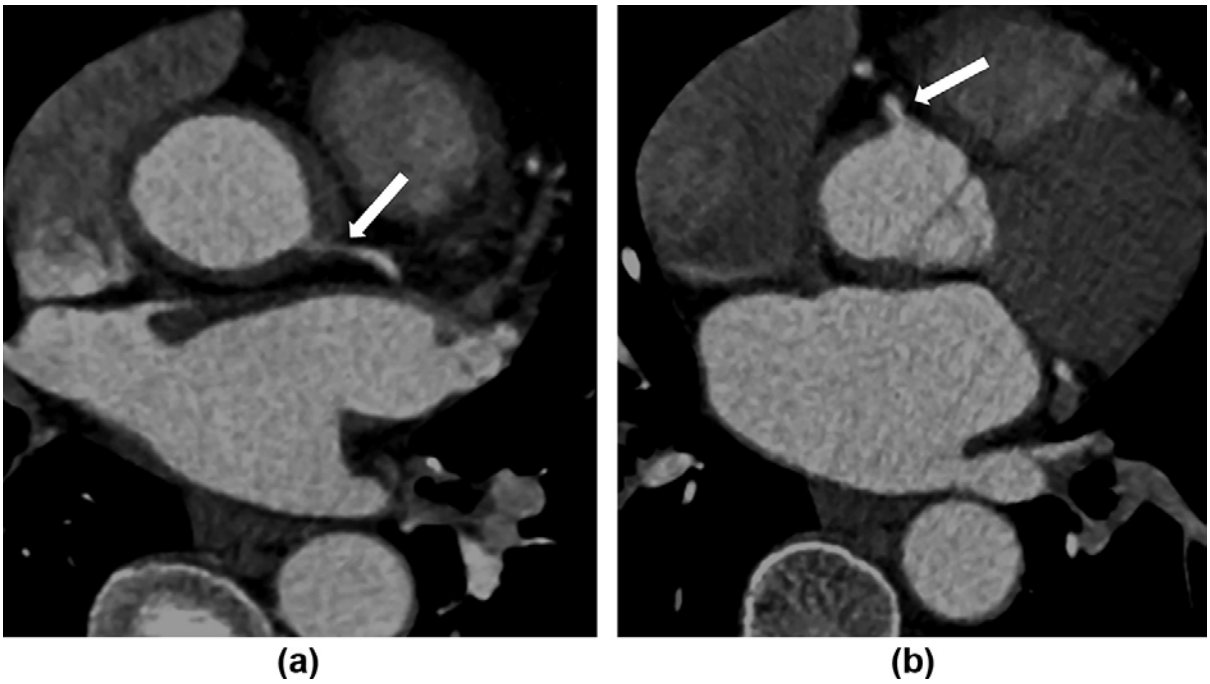


Fig. 6 — 58-year-old male with syphilitic aortitis. Axial images (A, B) show the typical findings: thickening of aortic wall with the involvement of the orifices of the left and right coronary arteries, which cause the stenosis of the coronary ostium (arrow).

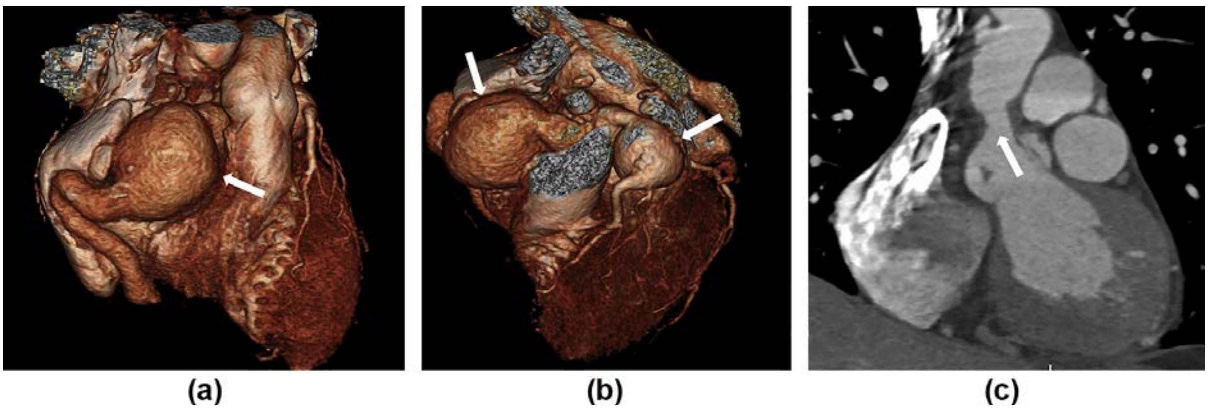


Fig. 7 — 45 -year-old female with Takayasu's arteritis. Volume rendering images (A, B) show aneurysms in the RCA and LAD (*yellow arrow*) , coronal multiplanar

reformatted image (C) shows involvement of the ascending aorta with mural thickening and luminal stenosis (*white arrow*).

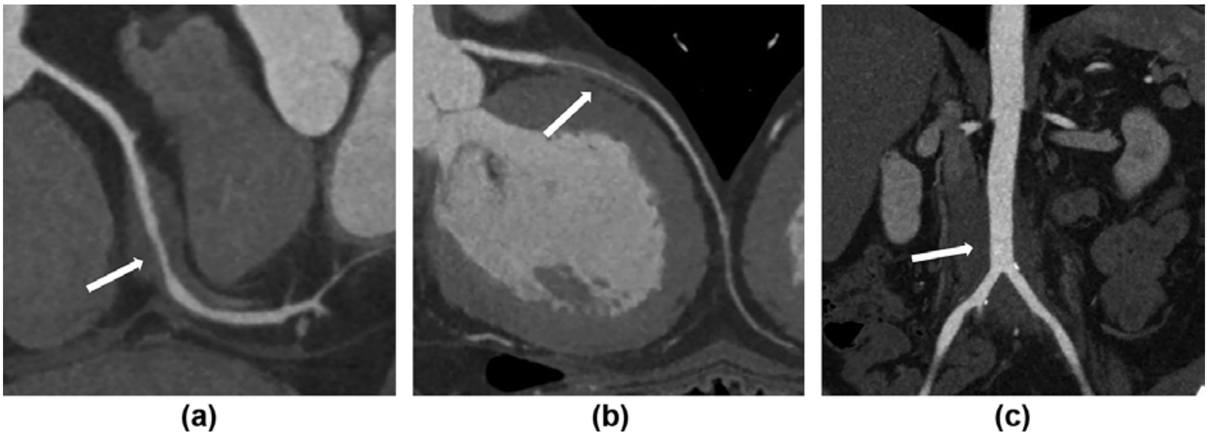


Fig.8 — 47-year-old male with coronary periarteritis. Curved planar reformatted images (A, B) show rings of soft-tissue attenuation surrounding the RCA and LAD (*arrow*), coronal multiplanar reformatted image (C) shows similar involvement of the distal abdominal aorta (*arrow*).

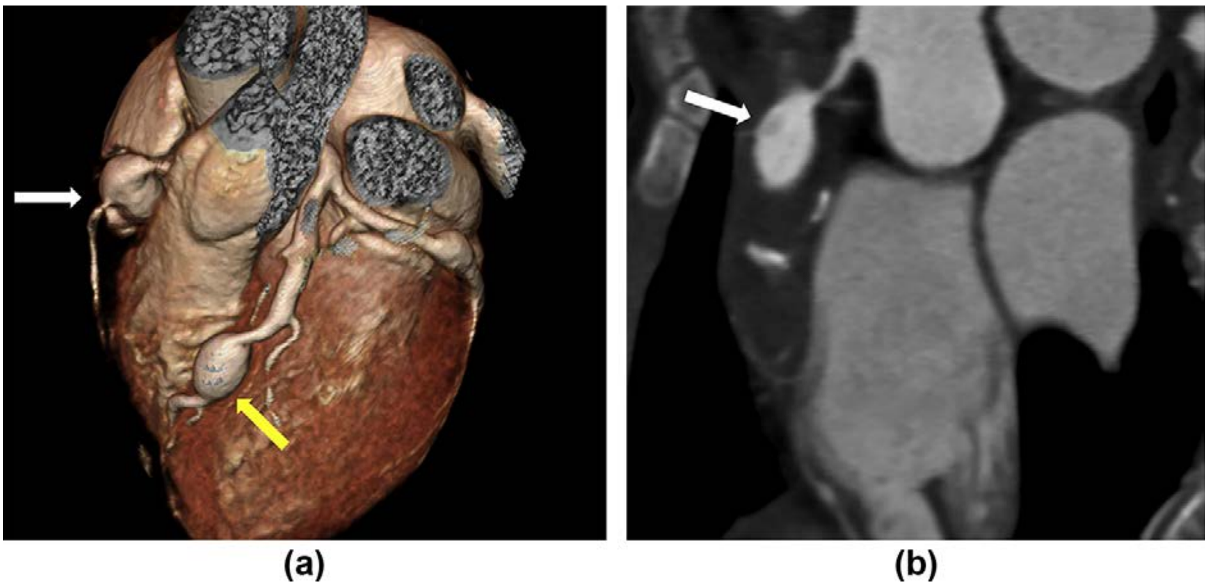


Fig. 9 — 6-year-old boy with Kawasaki disease. Volume rendering image (A) shows aneurysms in the RCA (white arrow) and LAD (yellow arrow), Curved planar reformatted image (B) shows the aneurysm in the RCA with thrombosis. (white arrow).

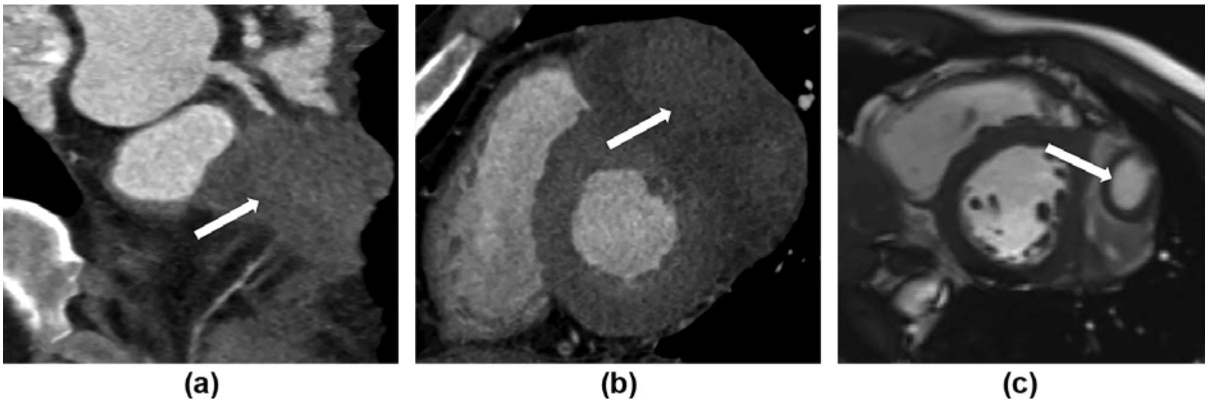


Fig. 10 — 22-year-old male with coronary pseudoaneurysm. Curved planar reformatted image (A) shows mass like soft tissue density in the middle segment of LAD (arrow), Multiplanar reformatted image (B) shows higher density in the center of the mass-like lesion (arrow). MRI shows a lumen within the lesion (C) (arrow).

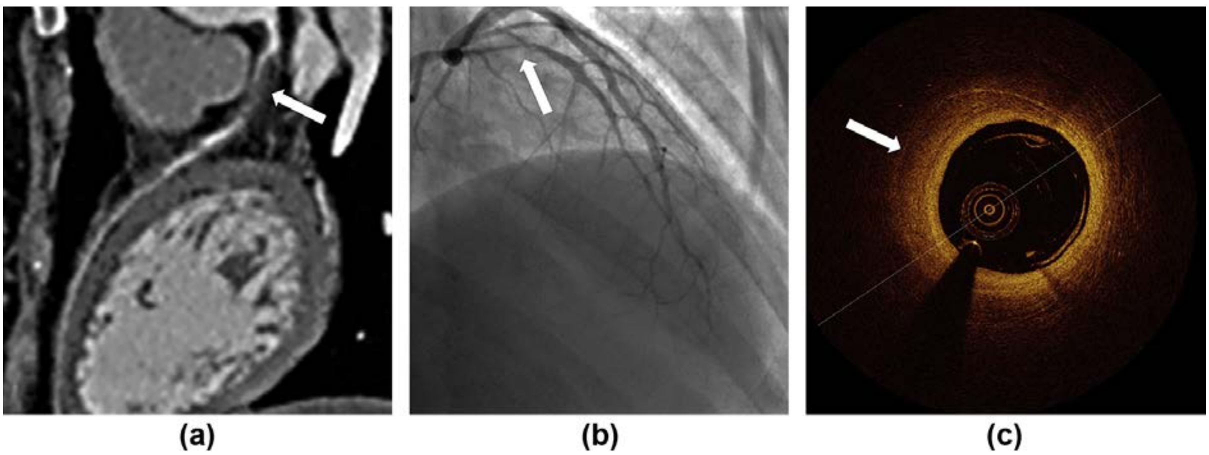


Fig. 11 — 29-year-old female with coronary intramural hematoma. Curved planar reformatted image (A) shows coronary wall thickening and stenosis in the proximal LAD (arrow), invasive coronary angiograph (B) shows the stenosis in the proximal LAD with smooth intima (arrow). (C) Optic coherence tomography shows intramural hematoma without an intimal tear.

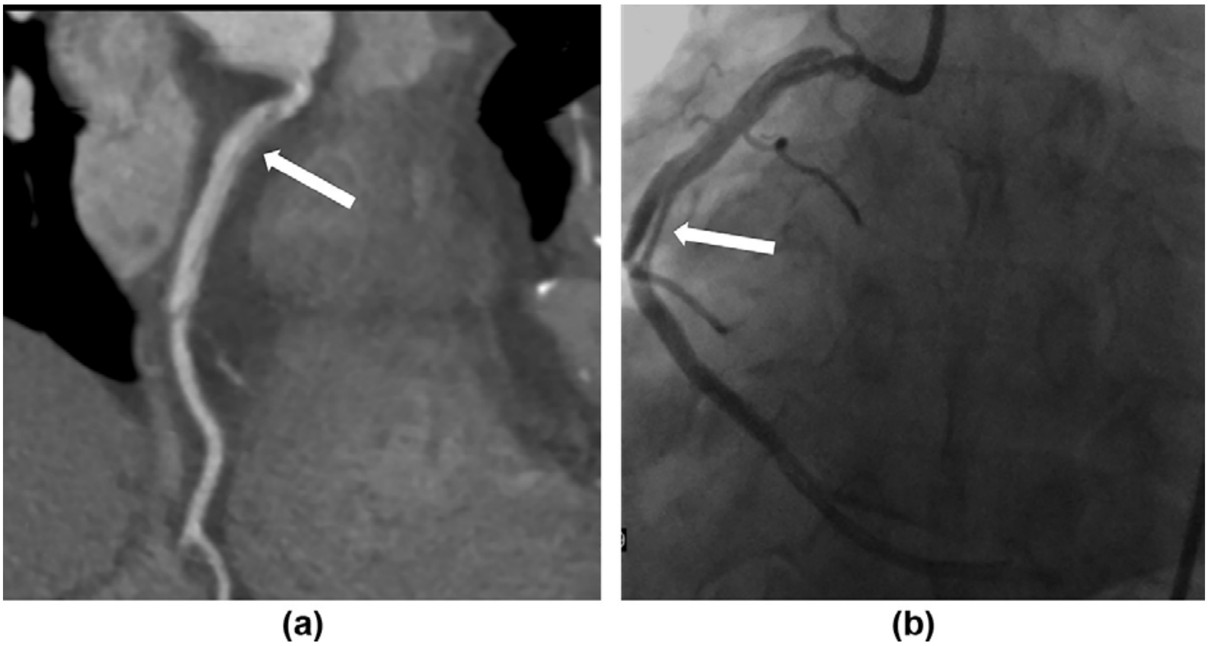


Fig. 12 — 52-year-old male with traumatic coronary dissection. Curved planar reformatted image (A) shows intimal flap within the proximal and middle segment of RCA (arrow), invasive coronary angiograph (B) shows the dissection in the RCA (arrow).