Journal of Cardiology (2009) **54**, 335—338

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JOURNAL of CARDIOLOGY
Official Journal of the Japanese College of Cardiology
www.elsevier.com/locate/jjcc

CASE REPORT

An extraordinary cause of ischemic chest pain in a young man: Congenital ostial atresia of the right coronary artery

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Received 19 December 2008; received in revised form 10 January 2009; accepted 14 January 2009
Available online 25 February 2009

KEYWORDS
Ostial coronary artery atresia;
Coronary anomaly

Summary Chest pain in a young person without cardiovascular risk factors is usually attributed to noncoronary causes; however, if the history suggests ischemic pain, the potential presence of unusual cardiovascular abnormalities should not be disregarded. The present case describes a young man with solitary congenital ostial atresia of right coronary artery, who to our knowledge is only the second case in the medical literature. Manifestation of ischemic symptoms in a relatively advanced age in patients with coronary artery atresia may mislead clinicians to interpret them as signs of atherosclerotic coronary artery disease. Therefore congenital coronary artery atresia should be a part of the differential diagnosis particularly in young patients with ischemic symptoms and no cardiovascular risk factors.

Introduction

Atherosclerotic coronary artery disease (CAD) has a similar clinical picture to congenital coronary artery atresia. Manifestation of symptoms in advanced age may mislead clinicians to interpret them as signs of atherosclerotic coronary artery disease. Therefore, differential diagnosis of congenital coronary artery atresia requires a high degree of suspicion and systematic examination. Herein, we describe a young man with solitary congenital ostial atresia of the right coronary artery, who to our knowledge is only the second case in the medical literature [1].

Case report

A 34-year-old man presented to our outpatient clinic with a history of typical angina pectoris of 10

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0914-5087/$ — see front matter
doi:10.1016/j.jjcc.2009.01.008

years of duration. The patient's past medical history was otherwise unremarkable with no history of hypertension, hypercholesterolemia, or diabetes mellitus. He did not have a history of smoking or drug abuse and his family history was negative for coronary artery disease. His physical examination was completely normal. Laboratory blood examinations including glucose, total cholesterol, low-density lipoprotein, high-density lipoprotein and triglycerides, renal and liver tests, homocysteine, erythrocyte sedimentation rate, C-reactive protein, and coagulation parameters such as partial thromboplastin time (PTT) and prothrombin time (PT) were all within normal limits. His rest electrocardiogram revealed normal sinus rhythm without any ischemic changes.

Exercise TC-99m myocardial perfusion scintigraphy showed reversible myocardial perfusion defects on inferoapical, inferior and inferobasal walls of the myocardium, confirming presence of myocardial ischemia and absence of scar tissue. Consequently he underwent a coronary angiogram which showed the absence of right coronary ostium and filling of the right coronary artery through collateral vessels from the left coronary system (Figure 1). No right coronary ostium was observed in the aortogram (Figure 1). None of the coronary arteries showed any sign of atherosclerosis (Figure 1). Computed tomography (CT) angiogram of the patient revealed absence of the right coronary ostium (Figure 2). CT images also showed hypoplasia of the proximal segment of the right coronary artery which is a characteristic feature of congenital osteal atresia of coronary arteries (Figure 2). The hypoplastic proximal right coronary artery which ends blindly and its relationship with the right coronary cusp was clearly documented (Figure 2). CT images confirmed an atherosclerosis-free coronary

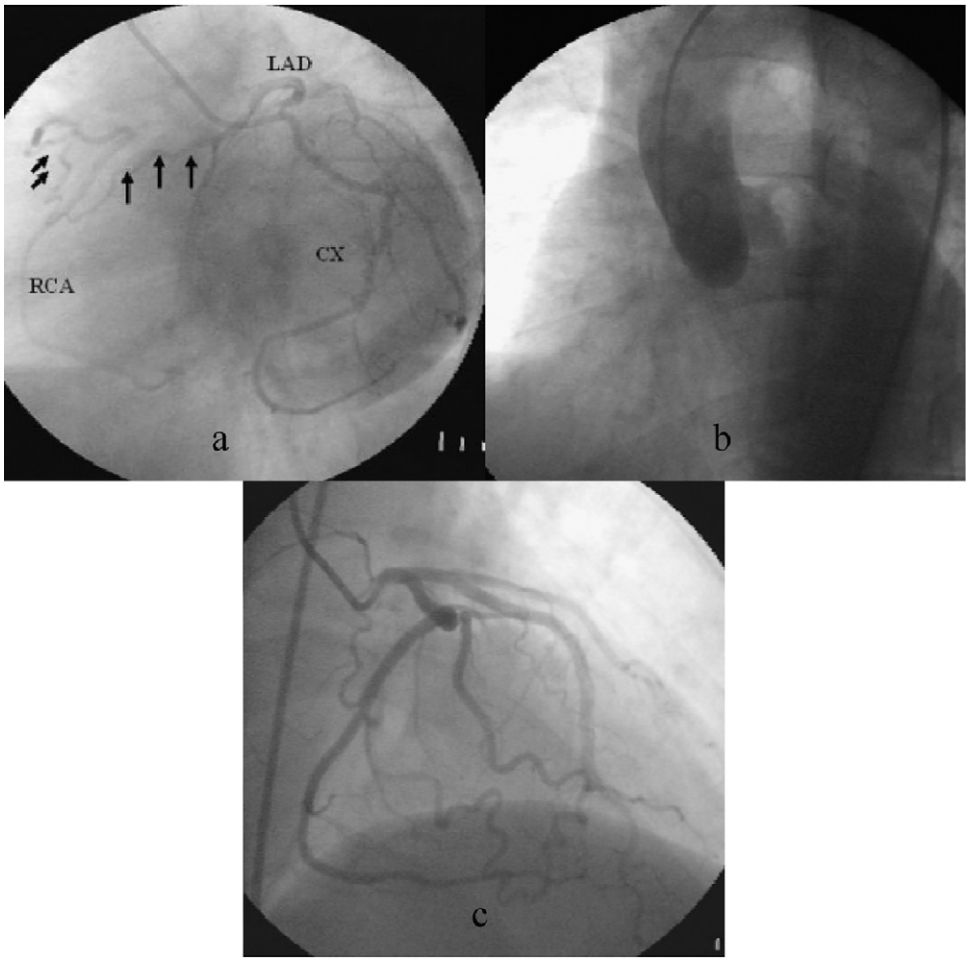

Figure 1 (a) Coronary angiogram revealing the filling of the right coronary artery through collateral vessels from the left coronary system (arrows). (RCA, right coronary artery; LAD, left anterior descending artery; CX, circumflex artery.) (b) No right coronary artery ostium was observed in the aortogram. (c) No sign of atherosclerosis was seen in coronary vessels.

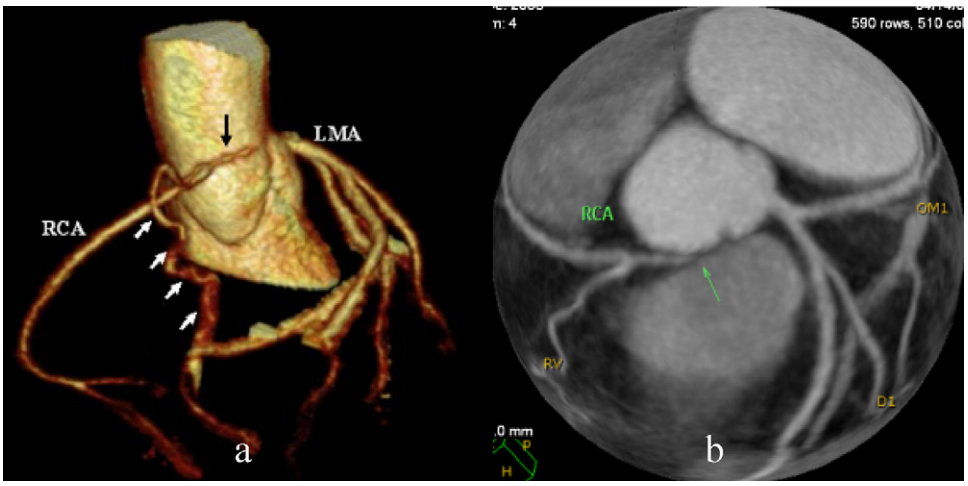

Figure 2 (a) Computed tomography angiogram revealing congenital atresia of the right coronary ostium. A hypoplastic proximal right coronary artery ends blindly (black arrow). Collateral formation from left coronary system to the right coronary artery was observed (white arrows). (RCA, right coronary artery; LMA, left main artery.) (b) Hypoplastic proximal right coronary artery that ends blindly (arrow) and its relationship with the right coronary cusp.

tree. Collateral formation from left coronary system to the right coronary artery was also observed (Figure 2). Coronary calcium analysis, performed by computed tomography, showed ''0'' coronary calcium scoring. Chronic total occlusions caused by atherosclerotic plaque ruptures are well known with their high calcium load. This characteristic calcium deposition was absolutely absent in our case. We believe the absence of calcium is one of the most important convincing features for the diagnosis of coronary atresia rather than an atherosclerotic occlusion. Echocardiogram showed a completely normal left ventricular function without any segmental wall motion abnormalities and absence of valvular heart disease. The patient was also examined to rule out any systemic disease that may lead to ostial occlusion of right coronary artery such as systemic lupus erythematosus, Takayasu's arteritis, and cardiovascular syphilis. All clinical and laboratory examinations were negative for such diseases.

His symptoms showed improvement in response to medical management with metoprolol 100 mg/day, therefore surgery was not considered for the time being.

Discussion

Variations of coronary arteries in origin, course, or distribution are found in 1.3—5.64% of patients undergoing coronary angiography and in 0.3% of autopsied individuals [2]. Congenital abnormalities of the coronary arteries, which lead to nonatherosclerotic coronary obstruction, include anomalous origins from the aorta, contralateral sinus of Valsalva or the pulmonary trunk; the presence of a single coronary artery; ostial atresia; high take-off coronary ostia; coronary artery fistulas or aneurysms and myocardial bridges [3]. Stenosis or atresia of coronary arteries can be solitary anomalies, but they are frequently accompanied by congenital defects such as coronary artery calcification, supravalvular aortic stenosis, Hurler's syndrome, Friedreich's ataxia, homocystinuria, progeria, pseudoxanthoma elasticum, and Marfan's and Ehlers-Danlos syndromes [3]. Congenital atresia or severe coronary ostial stenosis are usually associated with the hypoplasia of proximal segments of the involved coronary arteries. This characteristic finding was also present in our patient.

Almost all reported sporadic cases of coronary artery atresia in the medical literature are regarding the atresia of left main artery. There are a total of three cases of congenital osteal atresia of right coronary artery in the international literature [1,4,5]. The present case is only the second solitary congenital ostial atresia of right coronary artery in the medical literature. In two other cases, ostial atresia of right coronary artery was associated with Marfan syndrome in a 45-year-old woman and aortic-ventricular tunnel in a 6-week-old baby [4,5]. In a review by Musiani et al., the authors found 28 published cases of left main coronary atresia, 15 of which were pediatric; five of these cases had associated cardiac anomalies [6]. While pediatric patients (age: 2 months to 18 years) were

usually overtly symptomatic early in their life, adult patients (age: 38—71 years) began showing symptoms only at an advanced age. Musiani et al. reported that most adult patients with congenital ostial left main artery atresia had a normal and unrestricted childhood and adolescence, and began showing symptoms only at an advanced age.

Surgical revascularization is definitely the procedure of choice in adult patients with left main atresia. However, there is not much experience to comment on the management for right coronary artery atresia. One other patient with solitary right coronary atresia was treated with saphenous vein patch angioplasty [1]. The second case of a 45-year-old woman with Marfan syndrome was treated with reimplantation of right coronary artery [4]. Finally, the third patient who was a 6-week-old baby with an aortic-ventricular tunnel, underwent Piehler's modification for right coronary artery bypass using polytetrafluoroethylene (PTFE) graft [5]. Unfortunately, as all published cases were essentially case reports based on short-term follow-ups, long-term survival and quality of life of these patients are not known. In our case, since our patient was responsive to anti-anginal management, we preferred medical strategy as our initial strategy.

The clinical presentation of atherosclerotic CAD may be similar to that of patients with congenital coronary artery atresia and manifestation of symptoms at a relatively advanced age may mislead clinicians to interpret them as signs of atherosclerotic CAD. We believe many cases of congenital osteal artery atresia are being misdiagnosed as atherosclerotic CAD and the actual incidence of this anomaly could be underestimated. Congenital ostial coronary artery atresia should be a part of the differential diagnosis particularly in young patients presenting with a totally occluded coronary artery and no cardiovascular risk factors.

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