# Left main coronary artery aneurysm: a case report and review of the literature

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**Left main coronary artery aneurysm is an uncommon feature of coronary artery disease in adults. We describe the case of a large aneurysm in a 58-year-old patient undergoing cardiac catheterization for effort angina and inducible myocardial ischemia. Specific considerations about the underlying causes and therapeutic options are discussed.**

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The incidence of coronary artery aneurysms on routine angiography in adults varies from 1 to 5%. However, aneurysms of the left main coronary artery are very rare[1]. Aneurysms of the epicardial coronary arteries are more frequently encountered in the right coronary and circumflex artery and are associated with severe atherosclerosis[1-3]. Most reported cases of left main aneurysms were found incidentally during coronary angiography performed in patients with signs or symptoms suggestive of coronary artery disease and myocardial ischemia[1-4]. We describe the case of a large left main aneurysm in a patient undergoing cardiac catheterization at our Institution and discuss the clinical and therapeutic aspects.

## Case report

A 58-year-old obese white female with a 5-year history of non-insulin-dependent diabetes and systemic hypertension was referred to our Institution for exercise-induced chest pain and a recent episode of acute pulmonary edema. The medical history revealed an acute long-lasting febrile illness at the age of 6 and mild combined, obstructive-restrictive pulmonary disease at functional tests. The cardiovascular history dated back to the age of 44 when the patient was hospitalized for acute pulmonary edema associated with uncontrolled systemic hypertension. Thereafter, she complained of effort-induced angina and shortness of breath. A stress perfusion SPECT thallium-201 study revealed a reversible anterior and infero-lateral defect consistent with myocardial ischemia. Medical therapy including transdermal nitrates and β-blockers was started. In the past year, the symptoms became more frequent and more severe, until 4 weeks prior to admission when she suffered another episode of pulmonary edema. The physical examination was unremarkable and the electrocardiogram showed regular sinus rhythm with a heart rate of 60 b/min and Q waves in the inferior leads with persistent ST segment and T wave abnormalities in the inferior and lateral leads. Laboratory tests revealed mild normocytic anemia (hemoglobin 10 mg/dl, mean cell volume 80 $\mu m^3$) with normal iron deposits and normal coagulation and plasma lipid profiles. The day after admission the patient underwent cardiac catheterization and coronary angiography (Figs. 1-3), which revealed a large, severely calcified aneurysm measuring 14.1 × 24.7 mm and entirely involving the left main coronary artery. Severe calcifications were observed in the left anterior descending coronary artery with a significant stenosis in the proximal segment and mild ectasia in the intermediate segment of the left circumflex coronary artery which appeared totally occluded in its distal portion. The right coronary artery was markedly calcified starting from the ostium and chronically occluded in the intermediate segment. The left ventricle was moderately dilated with a severe lowering of the ejection fraction (30%) (Fig. 4). Chest X-rays showed an "egg shell" formation corresponding to the calcified left main aneurysm and signs of localized pulmonary

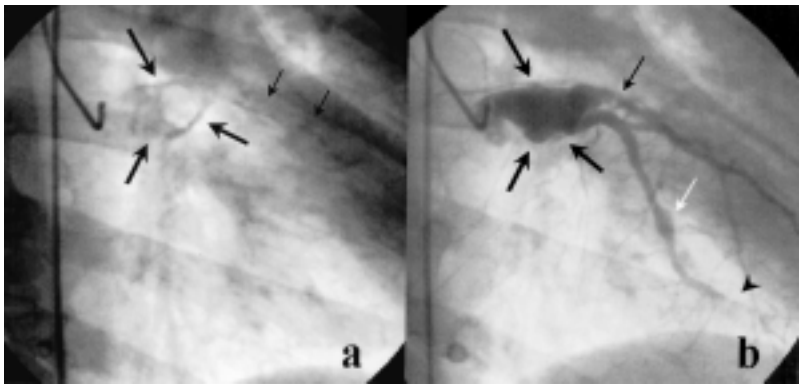


**Figure 1.** *A fluoroscopic angiogram showing ring calcification of the left main coronary artery* (large black arrows) *and calcifications of the left anterior descending coronary artery* (small black arrows) (a); *contrast injection reveals a large calcified aneurysm of the left main coronary artery* (large black arrows), *significant stenosis in the proximal left anterior descending artery* (small black arrow), *mild ectasia in the intermediate left circumflex coronary artery* (white arrow) *and total occlusion of the distal left circumflex coronary artery* (arrowhead) (b) *(15° right anterior oblique projection).*

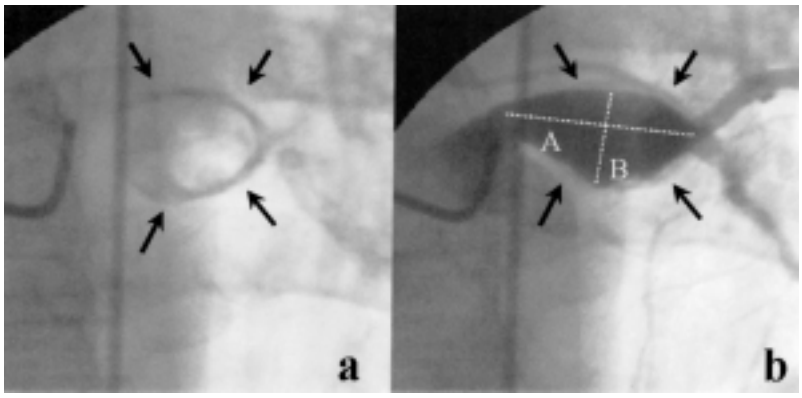


**Figure 2.** *Detail of the fluoroscopic angiogram showing ring calcification of the left main coronary artery* (arrows) (a); *detail of the large calcified aneurysm of the left main coronary artery with maximal longitudinal (A = 24.7 mm) and transverse (B = 14.1 mm) diameters* (b) *(20° cranial anterior oblique projection).*

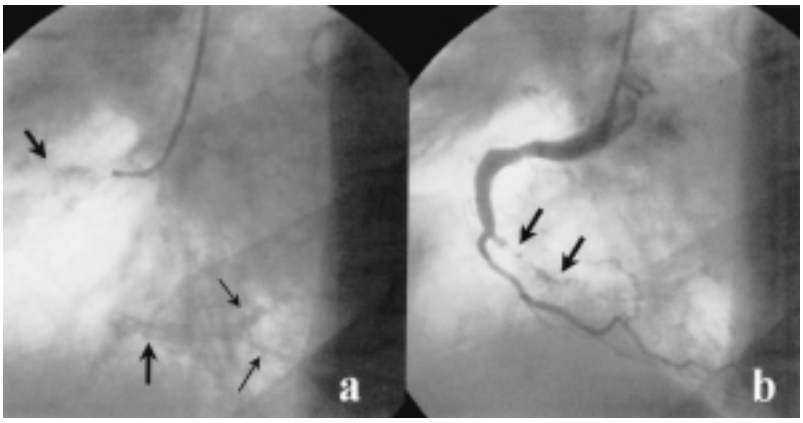


**Figure 3.** *Fluoroscopic angiogram showing severe calcification of the right coronary artery* (arrows) *starting from the ostium* (a) *and total occlusion in the intermediate tract* (arrows) (b) *(30° left anterior oblique projection).*

fibrosis (Fig. 5). Further tests, performed after having diagnosed a left main aneurysm revealed an erythrocyte sedimentation rate of 38 mm/hour and a C-reactive protein concentration of 69 mg/l (normal range < 6 mg/l); screening for hepatitis B revealed high anti-HBsAg and anti-HBcAg titers; however, liver and renal function tests and homocysteine plasma levels were in the normal range. Medical therapy included aspirin (100 mg daily), isosorbide-5-mononitrate (60 mg twice daily), furosemide (50 mg daily), and enalapril (20 mg twice daily). The patient was discharged when asymptomatic and placed on oral anticoagulant therapy with warfarin.

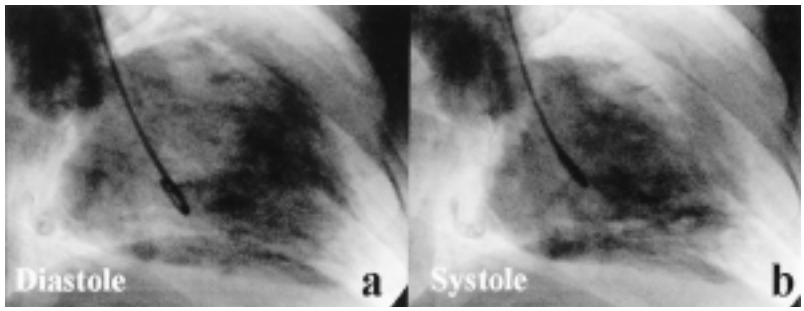


**Figure 4.** *Left ventricular angiography shows moderate dilation of the left ventricle and an ejection fraction of 30% (30° right anterior oblique projection).*

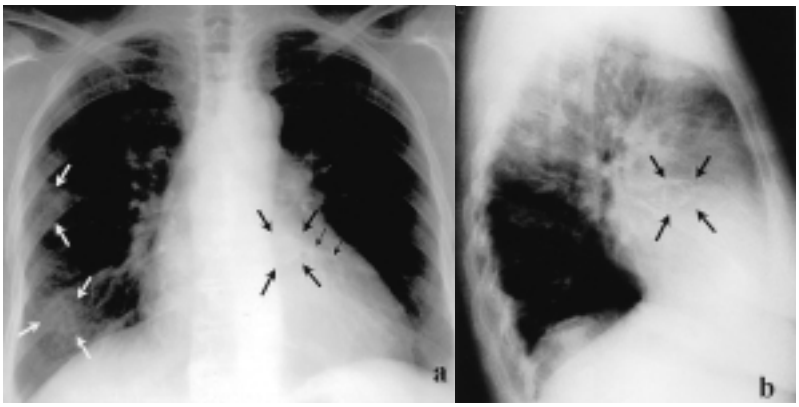


**Figure 5.** *Chest X-rays show coronary calcifications* (black arrows) *and signs of localized pulmonary fibrosis* (white arrows) *in an antero-posterior view* (a). *Calcification of the left main coronary aneurysm* (arrows) *as seen at chest X-rays (lateral view)* (b).

She was advised to return for clinical re-evaluation in view of the possibility of systemic arteritis; coronary bypass surgery was also suggested because of the severity of coronary artery disease.

## Discussion

Left main coronary artery aneurysm is an uncommon feature of coronary artery disease in adults[1]. Most reports in the literature are isolated case reports or autopsy case series. The largest angiographic series was reported by Topaz et al.[4] and comprised 20 322 patients undergoing cardiac catheterization. Among these, a left main aneurysm was diagnosed in 22 patients (0.1%). The authors defined an aneurysm as a significant, permanent, abnormal luminal enlargement of the left main coronary artery equal to or more than twice the diameter of the patient's largest vessel, or 3 times the diameter of a standard coronary catheter. In this series all patients were symptomatic for myocardial ischemia and had angiographically confirmed significant coronary artery disease; an isolated left main aneurysm was present in 10 patients (45%) whereas multiple aneurysms involving even the other coronary branches were found in the remaining 55%. Calcifications were observed in 7 patients (32%). Three-vessel coronary artery disease was diagnosed in 22 (73%) and coronary artery bypass surgery was performed in the majority of them.

**Etiology and differential diagnosis.** Possible etiologies of isolated left main coronary artery aneurysm include Kawasaki disease and other medium size arteritis (such as polyarteritis nodosa)[1,5-7]. Kawasaki disease is an acute febrile illness occurring during infancy (usually before 5 years of age) and is associated with cervical lymphadenopathy, rash and a "strawberry tongue". Sudden cardiac death or a non-fatal acute myocardial infarction (about 1%) due to acute coronary arteritis may occasionally occur[6]. In survivors of Kawasaki disease, even if asymptomatic, a large coronary artery aneurysm may be present and is frequently associated with significant coronary artery disease and coronary occlusions. Several case reports described late cardiac sequelae of Kawasaki disease in adults and suggest that a past medical history including Kawasaki disease should be considered as an important risk factor for ischemic heart disease[6,8,9]. "Classic" polyarteritis nodosa is a rare disease which may occur at any age. It is characterized by systemic arteritis that causes myocardial infarction and/or heart failure in about one third of patients; other clinical manifestations include systemic hypertension, peripheral nervous system and/or muscle involvement, hepatitis, abdominal cramps and rash[10]. Isolated coro-

nary symptoms of polyarteritis nodosa were also reported[7]. Severe atherosclerosis may also cause a left main aneurysm, usually in the setting of diffuse and severe coronary artery disease with multiple occlusions[1-3].

In our patient, the history of an acute febrile illness during infancy may suggest that the episode of acute pulmonary edema at the age of 44 is a late sequela of Kawasaki disease. On the other hand, the persistently elevated erythrocyte sedimentation rate, the increased C-reactive protein levels, anemia due to chronic illness, uncontrolled systemic hypertension and the presence of hepatitis B antibodies may also be suggestive of polyarteritis nodosa[11]. The almost complete absence of risk factors for coronary atherosclerosis at the time of the first clinical manifestation makes severe coronary atherosclerosis an unlikely, although possible, cause.

**Therapy.** Treatment of a left main coronary artery aneurysm may also be controversial. In a follow-up study of 5 patients with a left main aneurysm and without significant coronary stenoses, all experienced acute myocardial infarction with total occlusion of the artery distal to the aneurysm. This suggests thromboembolism as the pathogenetic mechanism[12]. In the series by Topaz et al.[4], 11 patients underwent coronary bypass surgery and 6 received medical treatment, while no information about the remaining 5 was available. According to previous experiences, anticoagulation with warfarin is considered the mainstay of therapy for a left main coronary artery aneurysm, in order to prevent thrombus formation. On the other hand, surgical therapy is reserved for cases with large thrombotic aneurysms or with myocardial ischemia due to significant associated coronary artery stenoses[13]. Intracoronary thrombolytic agents for angiographically evident thrombi have been used[14] but no data comparing antithrombotic therapy with antiplatelet drugs versus warfarin are available in the literature.

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