Case reports

Echocardiographic diagnosis in diffusely aneurysmal coronary artery disease

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Key words:
Aneurysm; Atherosclerosis; Coronary angiography; Echocardiography.

We present the case of a 73-year-old male with diffusely aneurysmal artery disease. Echocardiography revealed a 4.5 cm mass, adjacent to the right side of the heart, suggesting a diagnosis of coronary artery aneurysm. The findings were confirmed at magnetic resonance imaging and coronary angiography. Moreover, in view of the reported strong association between coronary and cerebral artery disease, the patient was submitted to cerebral magnetic resonance imaging, which also demonstrated aneurysmal disease involving the cerebral arteries.

(Ital Heart J 2003; 4 (9): 633-637)

Received February 20, 2003; revision received June 30, 2003; accepted July 22, 2003.

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Introduction

We report the case of diffusely aneurysmal coronary artery disease revealed by echocardiography and confirmed by coronary angiography and magnetic resonance imaging in a patient with a history of hypertension.

Case report

A 73-year-old male patient was referred to our department in May 2002 for gradually increasing fatigue and effort-induced breathlessness (NYHA class III) during the 4 weeks before admission. He had suffered from a previous myocardial infarction. He had a family history of cardiovascular disease, hypertension and diabetes mellitus and a 15-year history of systemic hypertension. He had ceased smoking 4 years previously. There was no prior history of stroke, diabetes or syncope. At the time of admission, physical examination was unremarkable and electrocardiography showed normal sinus rhythm with a heart rate of 82 b/min, Q waves in the inferior leads, R waves in the right precordial leads, and negative T waves in the infero-lateral leads (Fig. 1). Chest X-ray showed cardiomegaly, ectasia of the aorta, diffusely vascular accentuation of the pulmonary fields and, on the right side, scarring due to a previous pleurisy (Fig. 2). Laboratory tests were within normal limits, including antinuclear antibodies, extractable nuclear antigens and antimitochondrial antibodies.

Echocardiography. Transthoracic echocardiography revealed a left ventricle with an aneurysm involving the mid-basal segment of the postero-inferior wall and akinesia of the distal interventricular septum (Fig. 3); the wall thickness and the end-systolic and end-diastolic diameters were only slightly increased. The global ejection fraction was moderately low (40%). Moderate ectasia of the aorta from its root until its descending segment was also visualized (42 mm aortic root and ascending aorta, 29 mm aortic arch, 35 mm thoracic aorta). Moreover, transthoracic echocardiography revealed a round structure with a diameter of 4.5 cm, adjacent to the right heart and compressing the right atrium and ventricle. The mass had an echo-free center with an echogenic layer lining the perimeter, suggestive of thrombus (Fig. 4). Color flow Doppler revealed a narrow, low velocity jet swirling within the mass. The pulsed wave Doppler showed a biphasic flow (systo-diastolic) (Fig. 5). In this setting, it was decided to submit the patient even to transesophageal echocardiography, which revealed a narrow communication between the aorta and the round cystic structure (Fig. 6).

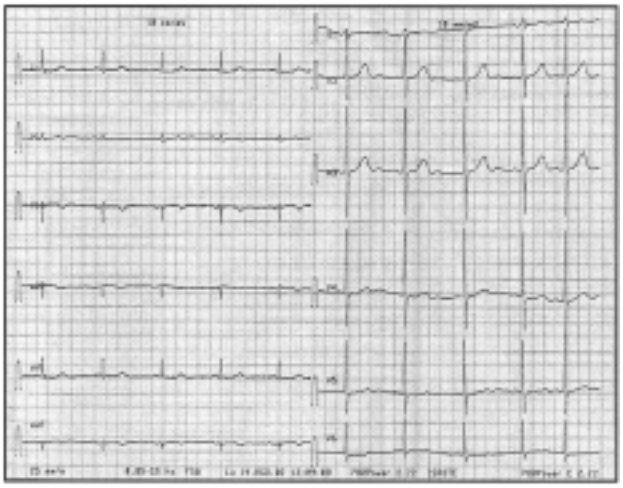


Figure 1. *Basal electrocardiogram.*

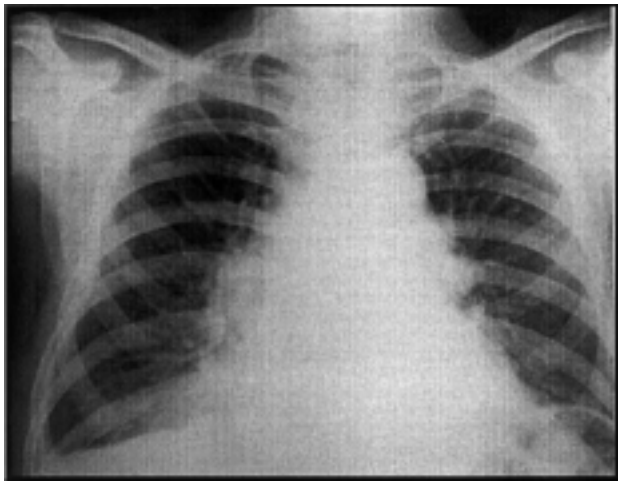


Figure 2. *Chest X-ray film. Note the cardiomegaly, ectasia of the aorta, the diffusely vascular accentuation of the pulmonary fields and, on the right side, the outcomes of a previous pleurisy.*

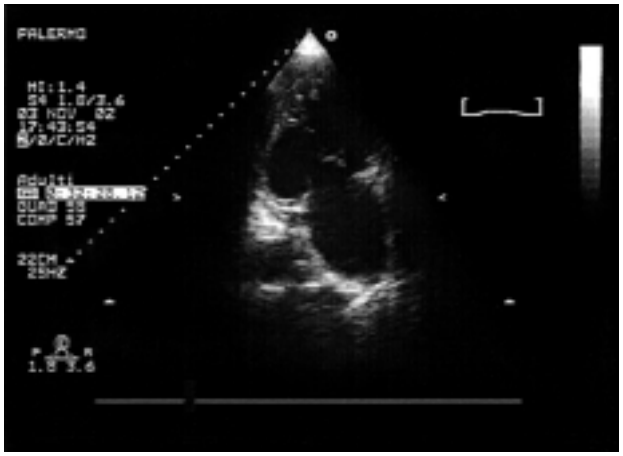
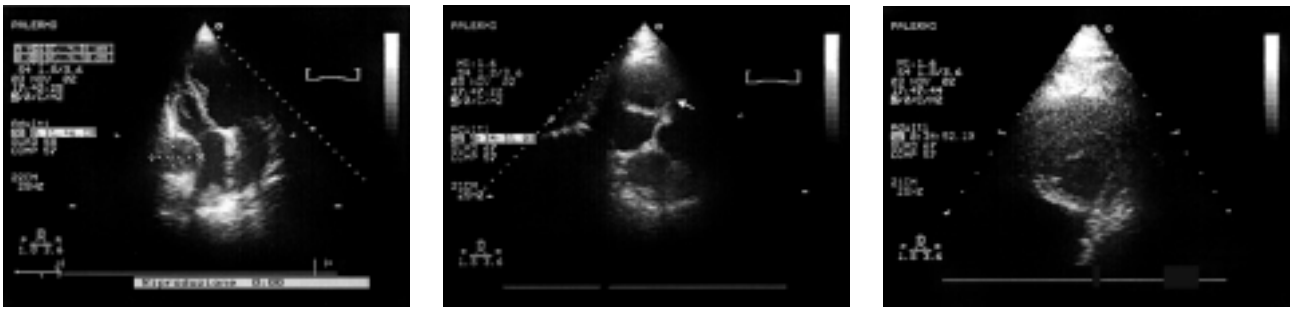


Figure 3. *Transthoracic echocardiography. Note the aneurysm of the left ventricle involving the mid-basal segment of the postero-inferior wall and akinesia of the distal interventricular septum.*

Coronary angiography. In view of his clinical status and owing to the abnormal wall motion as revealed by transthoracic echocardiography, it was decided to submit the patient to coronary angiography.

The coronary angiogram showed diffuse ectasia involving the proximal segment of the left anterior descending artery that was occluded distal to the first diagonal branch and refilled by a collateral system. The circumflex artery and its marginal branches also presented a large saccular aneurysmal structure of their proximal segment (Fig. 7A). The right coronary artery was occluded at its mid segment and refilled by a collateral system. However, selective right coronary injection revealed the presence of a slow swirling opacification of an 80 mm bisaccular and calcified structure located at its mid-proximal segment before the right coronary artery occlusion (Fig. 7B). Thus, the findings demonstrated the presence of diffusely aneurysmal coronary artery disease. Furthermore, aortic angiography was suggestive of moderate dilation of all the aorta, which was occluded at the origin of the left iliac-femoral branch. The left ventricle was mildly dilated with a moderately decreased ejection fraction (40%).

Magnetic resonance imaging. Uehara et al.[1] showed a strong association between cardiac disease and cerebral



A **B** **C**

Figure 4. *Transthoracic echocardiography (*A: *4-chamber view;* B: *subcostal view;* C: *detail of the subcostal view). Note the presence of a round structure (4.5 cm in diameter), adjacent to the right heart and compressing the right atrium and ventricle. The mass has an echo-free center with an echogenic layer lining the perimeter, suggestive of thrombus.*

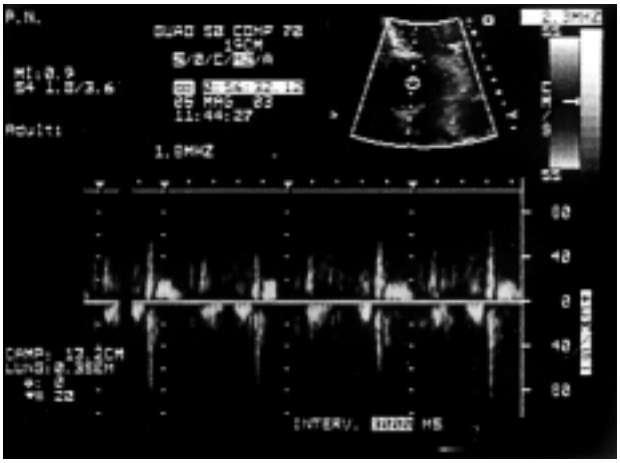


Figure 5. *Pulsed wave Doppler showing the continuous flow (systo-diastolic) into the mass.*

aneurysms. Because of the diffusely vascular aneurysmal disease, it was decided to also investigate the cerebral vascular district. Thus, the patient underwent cerebral and cardiac magnetic resonance imaging, the former, in order to exclude the presence of cerebral aneurysms and to determine the intra and postoperative mortality risk, and the latter, to better clarify the spatial anatomical relations between the great vessels, coronary arteries and the heart, thus confirming, the association between the right coronary artery aneurysms at angiography and the round structure adjacent to the right heart at echocardiography. In this setting, cerebral magnetic resonance imaging confirmed the hypothesis of cerebral artery disease, in particular revealing the presence of an aneurysm located at the apex of the basilar artery and involving the posterior cerebral and superior cerebellar arteries (Fig. 8). A team consisting of consultants in cardiology, and cardiac and cerebral surgery evaluated the patient's clinical status and the magnetic resonance imaging and angiographic findings and concluded that the intraoperative mortality risk for cerebral surgery was too high but that cardiac surgery could be performed[2,3]. The patient refused surgery and was therefore maintained on maximal oral therapy with short interval follow-up visits.

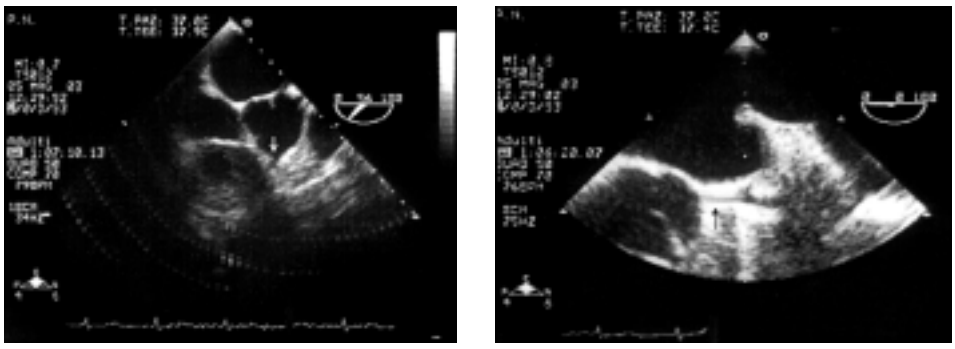


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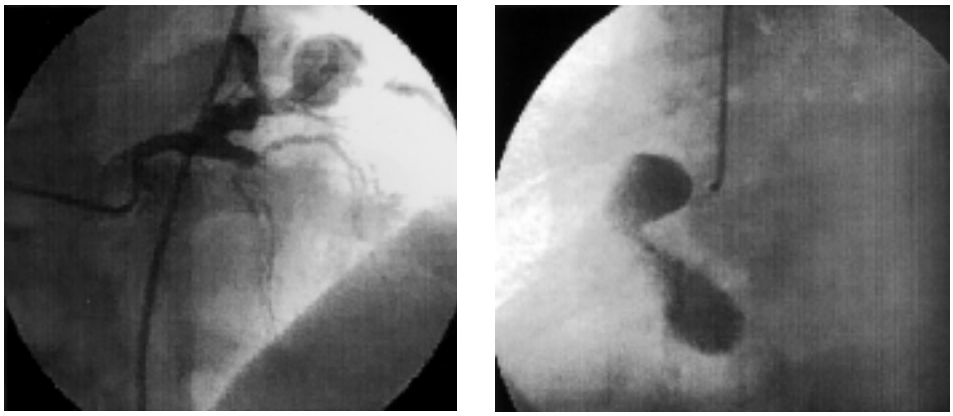
Figure 6. *Transesophageal echocardiography* (A, B)*. Note the presence of a narrow communication between the aorta and the round structure.*



A **B**

Figure 7. A: *coronary angiogram. Note the diffuse ectasia involving the proximal segment of the left anterior descending artery that was occluded after the first diagonal branch and refilled by a collateral system; moreover, the circumflex artery and its marginal branches show a large saccular aneurysmatic structure of their proximal segments.* B: *coronary angiogram. Note the presence of an 80 mm bisaccular and calcified structure of the right coronary artery located at its mid-proximal segment before the site of occlusion and in which the blood flow was slow and turbulent.*

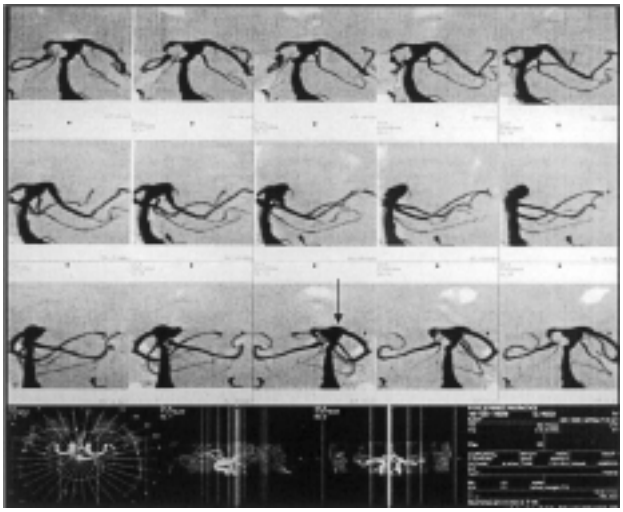


Figure 8. *Cerebral magnetic resonance imaging. Note the presence of the cerebral aneurysm located at the apex of the basilar artery, which involved the posterior cerebral and superior cerebellar arteries.*

Discussion

The most common cause of extensive aneurysmal artery disease in adults is atherosclerosis[4]; other causes include congenital disease, mycotic-embolic disease, mucocutaneous lymph node syndrome (Kawasaki's disease), syphilis, connective tissue disease, and arteritis[5]. Most atheromatous aneurysms are small and thick-walled and have a low risk of spontaneous rupture. Congenital aneurysms which are usually situated on the right coronary artery, are generally large and usually found in young patients[6]. Congenital aneurysms have been reported to rupture into the pericardial cavity[7] or into the right atrium[8].

The incidence of coronary artery aneurysms on routine angiography in adults varies from 1 to 5%[4,9,10]. Aneurysms of the epicardial coronary arteries are frequently encountered in the right coronary and circumflex arteries and are associated with severe atherosclerosis[11,12]. Aneurysms of the left main coronary artery are very rare[4,9]. Recent data revealed a strong association between cardiac disease and cerebral aneurysms which, depending on the patient's gender, ranges between 4 to 15%[1].

In our case, the diffusely vascular aneurysmal disease, the patient's advanced age and his multiple risk factors suggested severe atherosclerosis combined with a genetic connective tissue dysplasia[13] as the most probable cause of the aneurysmal disease.

In this setting, echocardiography allowed us to diagnose a coronary aneurysm demonstrating a round structure adjacent to the right heart and compressing the right atrium and ventricle. The characteristics indicative of aneurysm were the presence of a mass with an echo-free center and an echogenic layer lining the perimeter. Such a picture was suggestive of thrombus and in fact, color Doppler and pulsed wave Doppler confirmed the presence of turbulent and low-velocity blood flow within the mass.

The differential diagnosis of similar masses includes pericardial cysts, aneurysms of the sinus of Valsalva or of the aorta or of the heart wall, post-traumatic pseudoaneurysms of the ascending aorta or the pulmonary trunk, tumors of the heart or pericardium and thymoma[6]. With regard to this, we recorded a continuous flow into the aneurysm. Although normal coronary flow is predominantly diastolic, it is possible that the aneurysm created a low-pressure chamber and attenuated the normal systolic and diastolic differences.

Echocardiography and coronary angiography have a major role in the diagnosis of coronary aneurysms. Color Doppler echocardiography has been reported as an elective technique in the diagnosis of coronary aneurysms. However, coronary angiography is helpful when it is necessary to confirm the presence of multiple coronary aneurysms suspected at echocardiography, i.e. when there is a diffusely aneurysmal disease involving both coronary arteries. The echo findings suggestive of aneurysms of the right coronary artery have been reported in previous articles[6,14,15].

Magnetic resonance imaging generally permits the identification of coronary aneurysms as well as the determination of their dimensions. It also allows one to evaluate the spatial relations between the aneurysm and the great vessels and the heart. It has also been shown to be useful in the diagnosis of cerebral aneurysms, with the advantage over computed tomography of not necessitating ionizing radiation[6,15].

In summary, as the literature suggests, our case confirms that when a diffusely aneurysmatic disease, often with a genetic-familiar etiology, is hypothesized we always have to investigate different vascular districts, because of the frequent and severe simultaneous involvement of the cerebral and coronary artery systems.

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