

Giant Left Main Coronary Aneurysm without Associated Coronary Lesions

Antonio Merchán, José R. López-Mínguez, Francisco Alonso, Joaquín Fernández de la Concha, Reyes González and León Martínez de la Concha

Servicio de Cardiología y Unidad de Cardiología Intervencionista.
Hospital Regional Universitario Infanta Cristina. Badajoz.

Left main coronary artery aneurysms are very infrequent (0.1%) and the majority are related to atherosclerotic obstructive lesions. Only a few isolated cases without associated coronary lesions have been reported until now. The management of these patients is not well established, thus both conservative and surgical treatments have been postulated. We present a case of giant left main coronary artery aneurysm with a diameter of 27.7x18.6 mm (the biggest reported until now) without coronary tree associated stenosis. It was treated conservatively with oral anticoagulation only and the 5 years evolution has been favourable.

Key words: *Aneurysm. Coronary disease. Angiography.*

Aneurisma gigante del tronco común de la arteria coronaria izquierda sin lesiones coronarias asociadas

Los aneurismas del tronco común de la coronaria izquierda (TCCI) son muy infrecuentes (0,1%) y la mayoría de ellos está relacionada con lesiones obstructivas ateroscleróticas. Sin lesiones coronarias asociadas se han publicado hasta el momento sólo unos pocos casos aislados. El manejo de estos pacientes no está bien establecido y se ha postulado tanto el manejo quirúrgico como el conservador. Presentamos un caso de un aneurisma gigante del TCCI de diámetro 27,7 x 18,6 mm (el mayor publicado hasta la fecha) sin estenosis coronarias asociadas, el cual fue tratado de forma conservadora solamente con anticoagulación oral, presentando en el seguimiento a 5 años una evolución muy favorable.

Palabras clave: *Aneurisma. Enfermedad coronaria. Angiografía.*

INTRODUCTION

Coronary artery aneurysms are very rare, with prevalence rates that vary from 0.25% in Asian populations¹ to 2.6% in white populations,² although most studies show an incidence of 1% to 2%. The aneurysms occur most often in the right coronary artery, followed in frequency by the circumflex and anterior descending arteries.^{4,5} Aneurysms of the main left coronary artery (LMCA) are even more rare; in a study by Topaz et al involving a series of 22 000 coronary angiograms, they were found in only 22 patients (an occurrence rate of 0.1%).⁶

The disease most frequently associated with coronary aneurysms is coronary arteriosclerosis with severe stenosis in the surrounding areas,¹⁻⁵ they have also been reported in Kawasaki disease,⁷ systemic connective tissue disease⁸, infectious disease, and in thoracic trauma.⁹ In an autopsy review of 89 cases of coronary aneurysm by Daoud,¹⁰ 52% were atherosclerotic in origin, 17% were congenital, 11% mycotic-embolic, 11% dissecting, and 4% syphilitic. The Coronary Artery Surgery Study (CASS)⁴ registry, documents only 21 cases of isolated coronary aneurysm. The development of percutaneous cardiac intervention has allowed for more frequent observation of coronary aneurysm secondary to coronary angioplasty¹¹ and also in stent implantation.¹²

As for LMCA aneurysms, all 22 patients in the Topaz et al⁶ study had associated obstructive atherosclerotic disease; in another review of 12 cases published in 1987,¹³ 8 patients also presented with significant arteriosclerosis, one of whom was older

Correspondencia: Dr. A. Merchán Herrera.
Fernando Sánchez Sanpedro, 1, 5º. 06003 Badajoz. Spain.
E-mail: merchanh@inicia.es

Received 29 March 2001.
Accepted for publication 16 August 2001.



Fig. 1. Left coronary angiography with right oblique projection with 15° caudal angle which shows a gigantic LMCA aneurysm without associated atherosclerotic coronary lesions.

than 56 years of age. Only 4 patients had aneurysm with associated heart disease and were younger than 33 years—it was deduced that this type of aneurysm occurs only in younger people and was probably congenital.

Isolated LMCA aneurysm with no associated arteriosclerotic lesions is, therefore, rare, and the ongoing clinical management of these patients is not well established because of the lack of cases documented in the literature. We present a case of a patient with a giant LCMA aneurysm with no adjacent coronary obstruction followed for 5 years and treated with conventional therapy.

CLINICAL CASE

A 38-year-old man was admitted in 1996 for severe precordial pain radiating into his neck and jaw, triggered by physical activity, lasting approximately 1 hour, and accompanied by diaphoresis, nausea, and vomiting. Electrocardiogram revealed slight increase in ascension of the ST segment which then developed into a T-wave inversion anteriorly with no abnormal Q-waves. There was an elevated enzyme curve, with maximum values of CPK 571 U/l, CPK-MB 109 U/l, and LDH 586 U/l, leading to a diagnosis of acute myocardial infarct, without Q-wave, anteriorly. The rest of the biochemical, hematological, and coagulation test results, as well as connective tissue markers, were normal. The patient reported no fever prior to admission, and no history of cardiac disease,

hyperlipidemia, diabetes mellitus, syphilis, thoracic trauma, collagen disease, or vasculitis. He had a history of a grade I-II carcinoma of the papillary vesicle beginning in July 1990 for which he underwent transurethral vesicle resection and subsequent mytomicin chemotherapy. In 1993, metastasis was observed on bone scans, and a new course of chemotherapy was initiated with carboplatin, methotrexate, and vimblastine which was completed in 1995 (one year before the current admission). Cardiopulmonary examination was normal, and apical hypokinesia was observed on echocardiogram. Chest radiography revealed signs of osteopenia in both sides of the ribcage. Coronary angiography revealed a significant saccular LCMA aneurysm, 27.7 mm × 18.6 mm in diameter (Figure 1), with no associated cardiac lesions. Ventriculography showed an apical aneurysm and a 58% ejection fraction.

During his hospital stay the patient did not have another episode of precordial pain, and a stress test prior to hospital discharge was negative. Bone gammagraphy, abdominal echocardiography, and oncology screen showed no worsening of his previous illness. Given his positive clinical course and the accompanying neoplastic illness, the option of surgical intervention was discarded, and treatment with long-term oral anticoagulants was continued. At 5-year follow-up the patient was well, and had experienced no cardiac symptoms; a basal electrocardiogram was within normal limits.

DISCUSSION

LCMA aneurysms are extremely rare, as indicated in the Introduction. Their size varies except the large multiple aneurysms occurring in Kawasaki disease, the largest of which are known to measure 25 mm in diameter,^{3,14,15} which is considerably smaller than the one seen in our patient—27.7 mm—making it the largest LCMA aneurysm reported to date. Although other diagnostic tests such as intracoronary echocardiogram were not used, the projections evident on angiogram appeared to be clear enough to confirm the anatomical relationship of the aneurysm with LCMA.

The majority of aneurysms are associated with proximal, causative severe atherosclerotic coronary stenosis. If we exclude other known causes, such as thoracic trauma, coronary angioplasty, and systemic inflammatory or collagen disease, LCMA aneurysm without associated cardiac lesions is even less usual. In the review by Lenihan et al¹³ of cases published before 1987, only 4 patients did not have coronary lesions and were younger than 33 years of age—therefore, the origin of the LCMA aneurysms was

considered congenital. We found another 5 earlier cases described in the literature,^{3,14-17} all men (50, 69, 77, 27, and 69 years of age, respectively.) All of them, with one exception, were significantly older than the patients in the study by Lenihan et al, although the researchers also attributed the aneurysms to congenital defects because they could not identify any other cause. Our patient, because of his age (38 years) and because he did not present with any condition known to cause coronary aneurysm, may also have had a congenital aneurysm. We cannot, however, discount the possibility that his neoplastic illness was a direct cause of the aneurysm secondary to metastases or neoplastic embolism, or even that the chemotherapy he received might have had a role in its genesis. It must be noted that of the patients included in previous studies, one third who were 77 years of age¹⁶ died of a disseminated malignancy 8 months after the aneurysm was identified. and, although the authors do not give the relevant details, it is reasonable to assume that the illness was already present before the aneurysm was detected, and, given their advanced age, the origin may have been neoplastic rather than congenital, as is possible in our case.

The origin of coronary aneurysms is not well understood. As with other aneurysms, it appears that thrombosis or embolization, or both, with vessel occlusion¹⁸ and the rupture of the aneurysm with sudden cardiac tamponade¹⁹ are the 2 most serious (albeit rare) consequences and can occur in isolated cases of coronary aneurysm without associated cardiac lesions.

While surgical intervention seems clearly indicated for symptomatic patients with severe coronary stenosis and an aneurysm, the course to follow in cases without significant associated cardiac lesions is unclear. While the 5 patients described by Rath et al¹⁸ presented with progression to an acute myocardial infarct due to occlusion of the aneurysmal vessel which was not previously stenosed, the CASS register documents that patients with aneurysms who did not have obstructive disease and received medical treatment had the same survival rate at 5 years as the control patients who did not have coronary atherosclerosis.

Although the small number of existing cases does not allow for establishing a clear course of treatment for these patients, and although in some isolated cases surgical intervention was chosen,^{4,5,15,17} individual medical treatment is a viable option, according to some studies.^{3,6,16,18} It is reasonable to assume that if thrombosis, facilitated by turbulence and local slow fluid, can occur within an aneurysm and produce the vessel occlusion,¹⁸ long-term treatment with oral anticoagulants may be recommended. Outcomes in such cases treated with this therapy have been good,

with no instances of new infarcts or cardiac death during follow-up.^{3,16,20} It should be noted that the cases of acute occlusion described by Rath et al¹⁸ were not treated with anticoagulant therapy.

Our patient had an excellent outcome, both during treatment in the hospital course and at 5-year follow-up which leads us to believe that long-term oral anticoagulant therapy may be the treatment of choice in those isolated cases of aneurysm without significant associated coronary lesions, including when the aneurysm is large and is located in the LCMA.

REFERENCES

1. Wang KY, Ting ChT, Sutton MSJ, Cheng YM. Coronary artery aneurysm: A 25-patient study. *Cathet Cardiovasc Intervent* 1999; 48: 31-38.
2. Syel M, Lesch M. Coronary artery aneurysm: a review. *Prog Cardiovasc Dis* 1997; 40: 77-84.
3. Baretella MB, Bott-Silverman C. Coronary artery aneurysm: An unusual case report and a review of the literature. *Cathet Cardiovasc Diagn* 1993; 29: 57-61.
4. Swaye PS, Fisher LD, Litwin P, Vignola PA, Judkins MP, Kemp HG et al. Aneurysmal coronary artery disease. *Circulation* 1983; 67: 134-138.
5. Burns CA, Cowley MJ, Vetrovec GW. Coronary aneurysms: a case report and review. *Cathet Cardiovasc Diagn* 1992; 27: 106-112.
6. Topaz O, DiSciascio G, Cowley MJ, Goudreau E, Soffer A, Nath A et al. Angiographic features of left main coronary artery aneurysms. *Am J Cardiol* 1991; 67: 1139-1142.
7. Kato H, Akagi T, Sugimura T, Sato N, Kazue T, Hashino K et al. Kawasaki disease. *Coron Artery Dis* 1995; 6: 194-206.
8. López-Gómez D, Shaw E, Alió J, Cequier A, Castells E, Esplugas E. Obstrucción del tracto de salida del ventrículo derecho por un pseudoaneurisma gigante de la arteria coronaria descendente anterior en un paciente con enfermedad de Behçet. *Rev Esp Cardiol* 2000; 53: 297-299.
9. Martí S, Roldán I, Cebolla R, Rincón de Arellano A, Palacios V, Algarra FJ. Aneurisma del tronco común de la arteria coronaria izquierda secundario a traumatismo torácico. *Rev Esp Cardiol* 1996; 49: 381-383.
10. Daoud AS, Pankin D, Tulgan H, Florentin RA. Aneurysms of the coronary artery. Report of ten cases and review of the literature. *Am J Cardiol* 1963; 11: 228-237.
11. Desai PK, Ro JH, Pucillo A, Weiss MB, Herman MV. Left main coronary artery aneurysm following percutaneous coronary angioplasty: A report of a case and review of the literature. *Cathet Cardiovasc Diagn* 1992; 27: 113-116.
12. Voigtländer T, Rupprecht HJ, Stär P, Nowak B, Kupferwaser I, Meyer J. Development of a coronary aneurysm 6 months after stent implantation assessed by intracoronary ultrasound. *Am Heart J* 1996; 131: 833-834.
13. Lenihan DJ, Zeman HS, Collins GJ. Left main coronary artery aneurysm in association with severe atherosclerosis: a case report and review of the literature. *Cathet Cardiovasc Diagn* 1991; 23: 28-31.
14. Nogaki H, Shioi K, Mase T, Aoyama T, Miura A, Nagata Y. Two cases of coronary artery aneurysm including one case of the left main coronary artery aneurysm. *Kyobu Geka* 1998; 46: 513-518.
15. Eguchi M, Kondoh S, Takagi Y, Tsuchihashi K, Abiru M,

- Minatoya Y et al. Giant noninflammatory and nonatherosclerotic coronary arterial aneurysm in the left main trunk assessed by intravascular ultrasound imaging. *Angiology* 2000; 51: 599-602.
16. La Vecchia L, Bedogni F, Ometto R, Mosele GM, Vicenzi M. Aneurysm of the left main coronary artery without obstructive disease: Report of a case presenting with severe unstable angina and treated with systemic thrombolysis. *Cathet Cardiovasc Diag* 1993; 30: 306-309.
 17. Lepojärvi M, Salmela E, Huikuri H, Karkölä P. Repair of an aneurysm of the left main coronary artery. *Ann Thorac Surg* 1996; 61: 1247-1249.
 18. Rath S, Har-Zahav Y, Battler A, Agranat O, Rotstein Z, Rabinowitz B et al. Fate of nonobstructive aneurysmatic coronary artery disease. Angiographic and clinical follow-up report. *Am Heart J* 1985; 109: 785-791.
 19. Iga K, Fujikawa T, Ueda Y, Miki S, Konishi T. Massive hemopericardium as a first manifestation of coronary aneurysm: successful surgical management. *Am Heart J* 1996; 131: 618-620.
 20. Myler RK, Schechtman NS, Rosenblum J, Kollinsworth KA, Bashoru TT, Ward K et al. Multiple coronary artery aneurysms in an adult associated with extensive thrombus formation resulting in acute myocardial infarction: successful treatment with intracoronary urokinase, intravenous heparin and oral anticoagulation. *Cathet Cardiovasc Diagn* 1991; 24: 51-54.