Images in cardiovascular medicine
Isolated distal coronary dissection in Marfan syndrome

Dominick J. Angiolillo, Raul Moreno, Carlos Macaya

Interventional Cardiology Unit, Cardiovascular Institute, San Carlos University Hospital, Madrid, Spain

(Ital Heart J 2004; 5 (4): 305-306)

Received August 18, 2003; revision received December 4, 2003; accepted December 10, 2003.

Address:

Dominick J. Angiolillo, MD, FESC

Interventional Cardiology Unit
Cardiovascular Institute
San Carlos University Hospital - planta 2N
Plaza Cristo Rey s/n
28040 Madrid
Spain
E-mail: dominickjangiolillo@hotmail.com

A 44-year-old female with Marfan syndrome and Braunwald class IIIC unstable angina was referred to our hospital for cardiac catheterization. The diagnosis of Marfan syndrome was made at a young age because of her phenotypic features rather than because of her symptoms. In particular, the patient presented the following phenotypic features: pectus excavatum, facial skeletal abnormalities, including malar hypoplasia, joint hypermobility, a high arched palate, an increased arm span-to-height ratio (> 1.05), scoliosis, an increased axial length of the ocular globe, and miosis. No pathologic findings were ever observed at chest radiography or electrocardiography. The patient's cardiovascular history began at the age of 23 when she started complaining of fatigue following small efforts. At physical examination a systolic murmur of the mitral valve was identified and serial echocardiographic imaging revealed mitral valve prolapse associated with severe regurgitation. No pathologic findings of the aorta or of the aortic valve were identified. The patient subsequently underwent mitral valve replacement (metallic prosthesis) due to severe mitral insufficiency secondary to mitral valve prolapse. Afterwards the patient remained asymptomatic on coumarin treatment until the age of 44 when a new hospitalization was required for a non-Q wave myocardial infarction of the inferolateral wall. At the time of arrival in hospital, she was asymptomatic with minor electrocardiographic changes and therefore a conservative medical approach was chosen (aspirin, nitroglycerin, atenolol and low-molecular-weight heparin). An increase in creatine kinase up to 809 IU/l was observed. Following hospital admission the patient presented several episodes of rest angina and was therefore referred for cardiac catheterization. At angiography, a long (20 mm), spiral dissection in the distal segment of the left circumflex artery was documented (Figs. 1 and 2); no other significant angiographic findings were observed and the aortic dimensions were within the normal range. The global left ventricular systolic

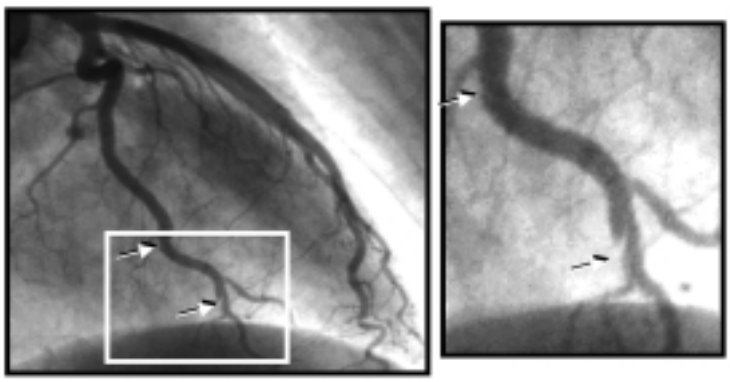

Figure 1. *30º right oblique anterior, 30º cranial projection.*

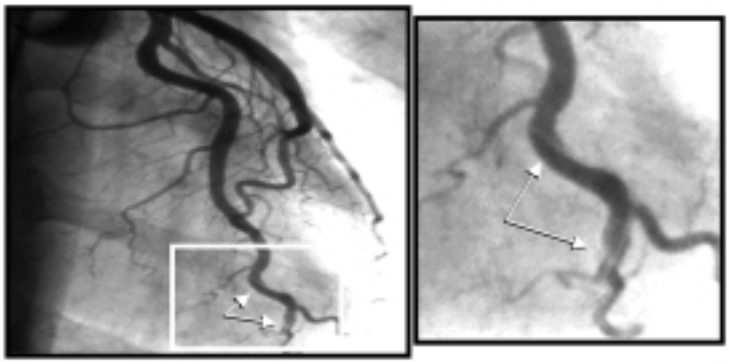

Figure 2. *Anteroposterior projection.*

function was preserved, although an area of hypokinesia of the diaphragmatic wall was documented. Since the coronary dissection was in the distal segment of a small diameter (2 mm) vessel, a conservative medical treatment strategy was preferred. The patient was discharged and 6 months later is doing well.

The Marfan syndrome is a heritable and generalized disorder of the connective tissue with major clinical features involving the skeletal, ocular and cardiovascular systems[1]. Although proximal coronary dissections leading to myocardial infarction are not infrequent findings in Marfan syndrome patients with proximal aortic dissections, isolated dissections (in the absence of aortic dissections) are extremely rare and occur in the proximal segments of the coronary arteries[2]. However, to date isolated dissections of the distal segments of the coronary arteries have not been reported in Marfan syndrome patients and therefore should be warranted.

Coronary artery dissections are commonly due to extension from a proximal aortic dissection, cardiac catheterization, percutaneous coronary interventions, cardiac surgery, and cardiopulmonary resuscitation[3]. Spontaneous coronary artery dissections are much less common than secondary dissections and although they are an extremely rare cause of ischemic heart disease, they frequently lead to myocardial infarction and sudden cardiac death[4]. The majority of cases of spontaneous coronary artery dissections appear to be idiopathic but dissections have been associated with Marfan syndrome, Kawasaki disease, systemic lupus erythematosus, blunt chest trauma, atherosclerotic disease, the use of contraceptives, and with cocaine abuse[5]. In particular, spontaneous coronary artery dissections most frequently (75% of cases) occur in young (mean age 40 years), otherwise healthy women, especially in the *peripartum* or early *postpartum* period. Dissections most frequently involve the proximal and mid segments of the coronary arteries[1-5]. Although their optimal management is still uncertain, both urgent coronary artery bypass grafting and coronary stenting are frequently carried out in this life-threatening clinical setting[5]. However, we describe an extremely unusual case of myocardial infarction due to a dissection in a distal coronary segment, in the absence of pathologic aortic findings, in a young female patient with Marfan syndrome in whom a conservative treatment strategy was chosen and proven to be safe.

Acknowledgments

Dr. Angiolillo was supported by the Working Group on Interventional Cardiology and Coronary Pathophysiology of the European Society of Cardiology.

References

1. Pyeritz RE, McKusick VA. The Marfan syndrome: diagnosis and management. N Engl J Med 1979; 300: 772-7.
2. Virmani R, Forman MB, Robinowitz M, McAllister HA Jr. Coronary artery dissections. Cardiol Clin 1984; 2: 633-46.
3. Wellford LA, Kelly TM. Survival with acute primary coronary artery dissection: a case report and review of the literature. J Emerg Med 1994; 12: 193-8.
4. Mohamed HA, Eshawesh A, Habib N. Spontaneous coronary artery dissection: a case report and review of the literature. Angiology 2002; 53: 205-11.
5. Vara Manso J, Barriales Alvarez V, Moris de la Tassa C. Spontaneous coronary artery dissection. Int J Cardiol 1998; 67: 263-4.