Coronary Aneurysm With Stent Dislocation Leading to the Diagnosis of Marfan Syndrome

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A 50-year-old male patient presented with unstable angina in our chest pain unit. He had a history of successful resuscitation after sudden cardiac arrest attributable to acute myocardial infarction 3 years ago. Coronary angiography at that time demonstrated acute proximal occlusion of the right coronary artery (Figure 1A) treated by balloon dilatation and implantation of 2 bare metal stents (Figure 1B). The second overlapping stent was necessary because of insufficient coverage of the diseased ostium after the first stent.

The current coronary angiography showed a coronary aneurysm located between the longitudinally dislocated proximal and distal stents (Figure 2). Coronary multislice computed tomography (SOMATOM Definition Flash, Siemens Medical Systems, Erlangen, Germany) confirmed the stent dislocation and displayed the complete extent of the partially thrombosed coronary aneurysm (Figures 3 and 4). We suspected a connective tissue disorder owing to further clinical signs including the patient’s height (2.04 m) and a marked pectus excavatum with a dilated ascending aorta of 48 mm in the computed tomography scan (Figure 5). Genetic analysis confirmed the diagnosis of Marfan syndrome by identifying a point mutation of intron 8 coding for fibrillin-1 gene. The patient was treated conservatively with dual-antiplatelet and antihypertensive medication, and genetic counseling and annual clinical reevaluations were recommended.

Giant coronary artery aneurysms are rare with an incidence of 0.02% to 0.04% in the general population. Often diagnosed incidentally during angiography, they can be linked to stent fractures with stent dislocation or to the polymer coating of drug-eluting stents. Only a few case reports of coronary artery aneurysms or acute dissections caused by connective tissue disorders are described. Marfan syndrome is a disorder of connective tissue structure and function with a reported incidence of ≈1 in 5000 individuals. Clinical manifestations include skeletal, ocular organs, and the cardiovascular system with a particular involvement of the aorta. Genetic testing has become an important tool in the diagnosis of Marfan syndrome and other related genetic connective tissue diseases.

We here report on an unusual case of a coronary artery aneurysm with stent dislocation leading to the diagnosis of Marfan syndrome. This case highlights the potential need to consider a connective tissue disease in patients with coronary artery aneurysms. Further studies and follow-up on this group of patients are needed to optimize diagnostic and treatment options.

Disclosures

None.

References

Figure 1. Coronary angiography with acute occlusion of the proximal right coronary artery (A) and result after implantation of stents (arrows) in the left anterior oblique view (B).

Figure 2. Coronary angiography of the right coronary artery with proximal coronary aneurysm (arrows) in left anterior oblique view (A) and in the right anterior oblique view (B).

Figure 3. Coronary multislice computed tomography displayed the complete extent of the coronary aneurysm (arrows) located between the 2 stents (arrow heads) with eccentric thrombosis (asterisk).

Figure 4. Three-dimensional volume rendering images demonstrate the anatomic relation between the stents, the right coronary ostium, and the coronary aneurysm, and the further course of right coronary artery, as well.

Figure 5. Thoracic computed tomography demonstrates a pectus excavatum (arrow) together with dilated ascending aorta with a lumen of 48 mm (asterisk) in comparison with the normal descending aorta (#).
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