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Images in Cardiovascular Medicine

Left Main Coronary Artery and Bilateral Mammary Artery Aneurysms in a Patient With Extensive Aortopathy

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Case Description

A 54-year-old woman presented with cardiac arrest. Her medical history was significant for extensive vascular aneurysms involving her aorta, celiac artery, bilateral carotid arteries, bilateral iliac arteries, and bilateral internal mammary artery (IMA) (Fig. 1 and Fig. 2). She has had 2 prior type B aortic dissection repairs and 1 recent ascending type A aortic dissection repair with bioprosthetic aortic valve replacement, reimplantation of the right coronary artery, and a single-vessel coronary artery bypass graft with left IMA to the left anterior descending coronary, with ligation of an aneurysmal left main coronary artery (Fig. 3).

After successful resuscitation, she underwent repeat angiography, which did not reveal acute findings but confirmed the presence of her aneurysmal left IMA graft (Fig. 3). Eventually, she received implantable cardioverter-defibrillator placement for secondary prevention and was discharged home on amiodarone, with plans for ventricular tachycardia ablation as an outpatient.

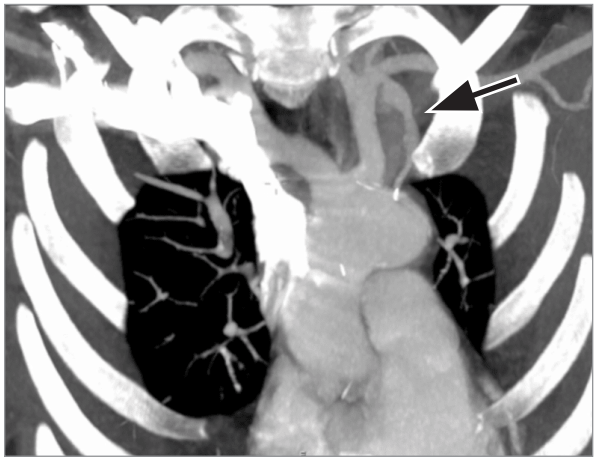

Fig. 1 Multiplanar reconstruction of computed tomography chest angiography shows an aneurysmal left internal mammary artery (arrow) 5 years before its use for grafting to the left anterior descending artery.

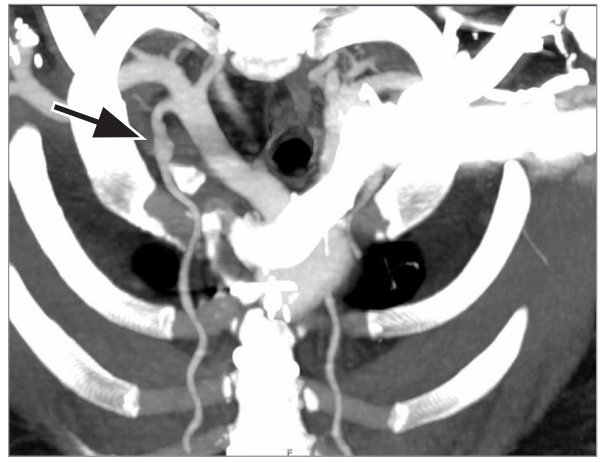

Fig. 2 Multiplanar reconstruction of computed tomography chest angiography shows an aneurysmal right internal mammary artery (arrow).

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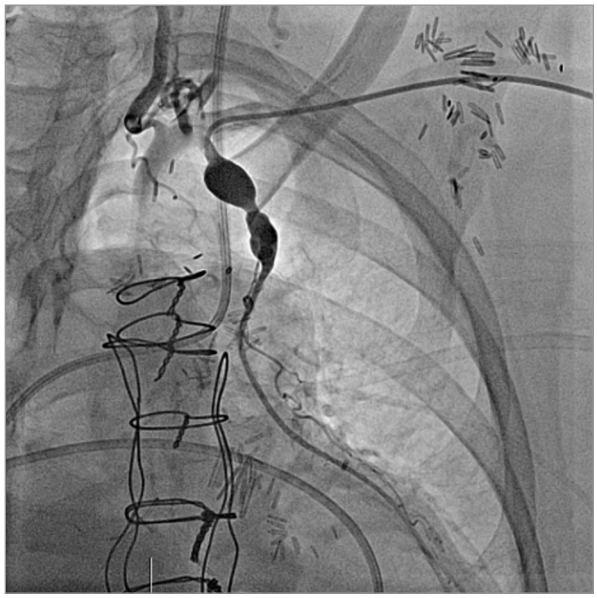

Fig. 3 Coronary angiography shows the left internal mammary artery graft to the left anterior descending artery.

Comment

Internal mammary artery aneurysms generally occur in the setting of trauma or are iatrogenic.[1] Bilateral IMA are extremely rare[2-4] and usually occur in the setting of genetically mediated aortopathies. These aortopathies often present in younger patients and can have a familial component, with up to 20% of patients with aortopathy having a family history of thoracic aortic dilation.[5] Notably, the mean patient age at presentation varies across aortopathy types, such as 56.8 years in familial nonsymdromic thoracic aortic dilation, 24.8 years in Marfan syndrome, and 64.3 years in sporadic cases.[5] These aortopathies can be categorized as either syndromic or nonsyndromic. Nonsyndromic conditions involve abnormalities that are limited to the cardiovascular system and do not manifest as external features of connective tissue disorders. Examples of nonsyndromic conditions include familial thoracic aortic aneurysm/dissection and bicuspid aortic valve with aneurysm. Syndromic conditions include Marfan syndrome, Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, Shprintzen-Goldberg syndrome, aneurysm-osteoarthritis syndrome, cutis laxa with aneurysm, and Turner syndrome.

The benefit of prophylactic surgery in the setting of hereditary thoracic aortic aneurysm disorders is uncertain and may depend upon the specific mutation of the aneurysm, the aneurysm's location, family history, absolute aortic size, and growth rate.[6]

There are currently no established guidelines for managing IMA aneurysms; however, previous cases have used surgical resection or, more frequently, coil embolization as treatment options.[2-4] No guidelines or recommendations on using aneurysmal IMA grafts in coronary artery bypass graft exist.

The patient in this case underwent extensive testing for both syndromic and nonsyndromic conditions, which yielded negative pathogenic alterations except for a variant of unknown significance in the *MYLK* gene. The *MYLK* gene, which encodes the myosin light chain kinase and plays a clinically significant role in aortic disease, particularly in the context of nonsyndromic heritable thoracic aortic disease.[7] Vascular smooth muscle cells, found predominantly in the medial layer of blood vessels, provide contractile tension and anchor the extracellular matrix's elastic fibers for stability. Mutations in these cells can disrupt their function, causing increased cell death, reduced aortic wall tone, and decreased extracellular matrix stability.[6]

Individuals harboring *MYLK* gene variations often manifest aortic dissection, even in the absence of clinically significant aortic enlargement. These variations predominantly affect the short form of myosin light chain kinase, which is the sole form expressed in the human aorta. Variations can either result in haploinsufficiency or be missense alterations that impair kinase activity, ultimately leading to reduced phosphorylation of the regulatory light chain and, consequently, diminished contraction of aortic smooth muscle cells.[8]

Abbreviations and Acronyms

IMA internal mammary artery

Article Information

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