Internal mammary arteriovenous fistula found late after aortic root replacement

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A 37-year-old male patient with Marfan syndrome was referred for a routine check-up at our department. At the age of 17, he was operated due to aortic root dilatation and severe aortic valve regurgitation. A composite graft was placed in the aortic position and the coronary arteries were re-implanted.

He had been seen elsewhere on a regular basis since the operation. Despite successful outcome and treatment with heart failure medication the ejection fraction remained <30%.

Upon examination, the patient was uncongested; however, in NYHA III. Surprisingly, there was a continuous right-sided subcostal murmurs and frémissence in the right intercostals spaces. The echocardiogram revealed an ejection fraction of 25% and dilatation of the proximal part of the coronary arteries. It was possible to visualize a continuous flow in a large-vessel-like structure in the right subcostal region (Panels 1A and B). A subsequent CT-scan revealed a large-distal arteriovenous fistula between the right internal mammary artery and vein (Panels 1C and D). A catheterization (Panels 1E and F) revealed a moderate arteriovenous shunt and the fistula was closed with an occluder (Supplementary data online, Figures S1 and S2).

An iatrogenous arteriovenous fistula between the mammary vessels is a rare complication to sternal closure, caused by perforation of the vein and the artery from the wire cerclage. It is not associated with Marfan syndrome in particular.

Most fistulas are discovered immediately. However, in this rare case it was not discovered until 20 years after surgery, in which the fistula had developed to considerable size. The fistula may have had a significant impact on the heart failure condition over the years.

Panel A: Colour Doppler of the distal part of the fistula, done with a 5 MHz echocardiography probe (Acuson, SC 2000; Siemens, USA). Panel B: CW Doppler demonstrated the continuous flow in the vessel shown in Panel A. Panel C CT scan with 3D volume rendering reconstruction of the large arteriovenous fistula between the right internal mammary vessels (arrow). Projection from the neck towards the diaphragm. Panel D: Same CT technique now looking from the back towards the front. The arrow indicates the fistula. Please notice the normal sized left-sided mammary vessels on the other side of the sternum. Panel 1E: Injection of contrast in the proximal part of the tortuous right mammary artery. Panel 1F: Contrast in the right mammary vein after a few cardiac cycles. Notice the filling of contrast in the superior caval vein (arrow).

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

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