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Hypertrophic cardiomyopathy with mid-cavitary obstruction and apical aneurysm thrombus after transapical myectomy and aneurysmectomy

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DESCRIPTION

A 58-year-old woman with known hypertrophic cardiomyopathy (HCM) was hospitalised for recurrent, intractable heart failure despite guideline-directed medical therapy. Her HCM phenotype was concentric with mid-cavitary obstruction (MCO) and an associated large apical aneurysm. Her cardiac history included resuscitated sudden cardiac arrest with subsequent implantable cardioverter-defibrillator (ICD), and ventricular tachycardia (VT) endocardial and epicardial ablation for appropriate ICD shocks during VT storm.

Given her aggressive clinical course, surgical reduction of her intracavitary gradient—invasively calculated as 90 mm Hg during a prior endocardial VT ablation—was entertained. To that end, she underwent preoperative CT angiography for coronary assessment, which identified myocardial bridging segments of the left anterior descending and first obtuse marginal arteries but no significant coronary stenosis. Dual source CT (Somatom Force, Siemens, Erlangen, Germany) cine images at peak systole (figure 1) demonstrated MCO with apposition of left ventricular walls and a typical 'hourglass' appearance (maximal wall thickness 2.6 cm). Moreover, a cardiac MR (CMR) study was ordered as part of surgical planning to better characterise ventricular morphology, function and scar pattern. CMR was performed using a 1.5 T scanner (Avanto, Siemens, Erlangen, Germany). Electrocardiography-gated breath-hold steady-state free precession cine images were obtained in short axis slices at 10 mm intervals (slice thickness 6 mm, 4 mm gap) and two-chamber, three-chamber and four-chamber views. This revealed severe left ventricular systolic dysfunction and a quantitative ejection fraction of 27%, consistent with 'burnt out' HCM. A gadolinium-based contrast agent (Dotarem, Guerbet, Villepinte, France) was injected in a 0.2 mmol/kg bolus and late gadolinium-enhancement images were acquired 10 minutes later using an inversion recovery gradient echo sequence. This revealed transmural scar in the thin-walled apical aneurysm and patchy, midmyocardial scar in the hypertrophied segments (figure 2A).

She subsequently underwent transapical myectomy of her midventricle, resection of her apical aneurysm with endocardial cryoablation of its orifice and Dacron patch aneurysmorrhaphy. A routine postoperative transthoracic echocardiogram with an ultrasound-enhancing agent (UEA) raised suspicion for a layered apical thrombus. A large mural thrombus with a typical jet-black

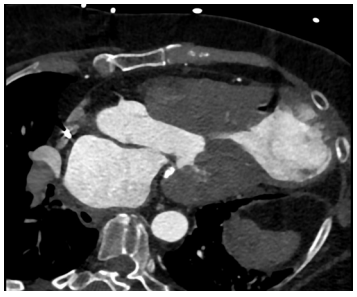

Figure 1 CT cine image of the left ventricle in a three-chamber long axis view demonstrating systolic obliteration of the midventricle and a large apical aneurysm.

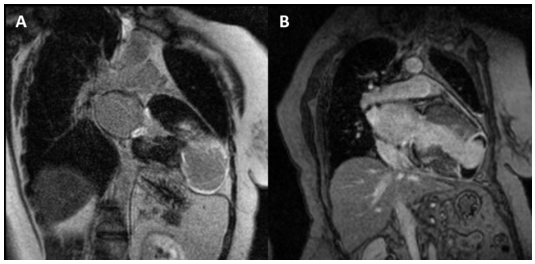

Figure 2 Late gadolinium-enhancement imaging of the left ventricle in a two-chamber long axis view reveals transmural scar in the apical aneurysm and patchy, midmyocardial scar in the hypertrophied segments preoperatively (A). Long inversion recovery sequence after contrast administration shows a large mural apical thrombus postoperatively (B). Considerable reduction in wall thickness is also apparent.

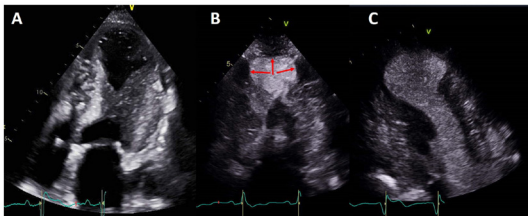

Figure 3 Transthoracic echocardiographic images showing systolic midventricular obstruction in a four-chamber apical view preoperatively (A) and left ventricular opacification with ultrasound contrast and a suspected layered apical thrombus producing filling defects (red arrows) in a two-chamber apical view (B). The thrombus is later confirmed with cardiac MRI and is no longer visible after 5 months of anticoagulation (C).

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appearance was subsequently confirmed with CMR using a long inversion time (TI) sequence (TI=600 ms, figure 2B) after contrast administration. She was started on systemic anticoagulation with intravenous heparin, bridged to warfarin with a target international normalized ratio (INR) of 2.0–3.0, and the remainder of her postoperative course was uneventful. An outpatient echocardiogram with a UEA approximately 5 months after warfarin initiation confirmed thrombus resolution (figure 3). She is currently being evaluated for cardiac transplantation by our institution's advanced heart failure service.

Patient's perspective

As a heart patient I have become very aware of how important it is to be checked as young as possible for any heart defects if there is a history of heart disease of any kind in the family tree. If I had been diagnosed at a younger age then maybe my disease would have not gotten to the point of surgeries. This is preventable with the help of medicines and great doctors! This is a genetic disease!

Learning points

- Hypertrophic cardiomyopathy (HCM) with mid-cavitary obstruction (MCO) is a well-established HCM phenotype that occurs in approximately 10% of HCM patients.[1 2] The presence of MCO has been associated with adverse outcomes in large HCM cohorts, including progression to end-stage heart failure, sudden cardiac death and ventricular arrhythmia as an initial manifestation of the condition.[3]
- Apical aneurysms are a characteristic feature of HCM with MCO, occurring in an estimated 25%–30% of such patients and thought to contribute to their poorer prognosis.[1 2] Thrombus formation is a known complication of apical aneurysms.
- Current data support transapical myectomy as an effective surgical therapy to relieve MCO and intracavitary gradients, with intermediate-term survival comparable to age-matched and sex-matched controls.[4]

Contributors BA-S is the primary and corresponding author responsible for the overall content and drafting of the manuscript. DD, EYY and FN made substantial contributions to the editing and review of the final manuscript. FN is the senior author and was also involved in the care of the patient described.

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