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CASE REPORT

"Burnt out" dilated hypertrophic cardiomyopathy causing acute LVAD thrombosis

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Key Clinical Message

Case report illustrates obstruction encountered in a patient with end-stage dilated hypertrophic cardiomyopathy (HCM) who underwent LVAD implantation. The morphology reversed in early postoperative period to HCM. Pump replacement required coring of the ventricular muscle. Dilated end-stage hypertrophic cardiomyopathy can revert back to the original morphology on decompression.

Keywords

Dilated hypertrophic cardiomyopathy, hypertrophic cardiomyopathy, left ventricular assist device, pump obstruction, pump thtombosis.

Case History

A 47-year-old man with a diagnosis of dilated cardiomyopathy was referred for LVAD bridge to heart transplantation in August 2011. He had no history of hypertension and family history of heart disease. A transthoracic echocardiogram (TTE) revealed severely dilated left ventricle (LV) with left ventricular ejection fraction of 18%, severe restrictive diastolic function, tenting of mitral valve (MV) leaflets with mild to moderate mitral regurgitation, trivial aortic regurgitation (AR), LV internal diastolic diameter of (LVIDd) 9.41 cm, LV internal systolic diameter (LVIDs) 8.01 cm, interventricular septum in systole (IVSs) of 1.34 cm, and LV posterior wall in systole of (LVPWs) 1.69 cm. The right ventricle (RV) was normal in size and had normal systolic function. The tricuspid valve (TV) annulus excursion was 2.2 cm. The pulmonary artery (PA) systolic pressure was 56 mmHg. The pulmonary vascular resistance was 2.28 Wood units. In view of the LVIDd of 9.41 cm, he was managed for dilated cardiomyopathy. At the last admission, he required milrinone infusion. HeartMate II (Thoratec Corp, Pleasanton, CA) LVAD (HM) implantation was carried out on cardiopulmonary bypass (CPB) with a beating heart. At surgery, the left ventricle was found severely dilated and cavernous. On coring the LV apex, the wall was found to be unusually thick.

On completion, the patient was easily weaned off CPB to LVAD support, with RPM of 9400, pulsatility index (PI) of 4.7 and pump power of 6.4 watts. The PA pressure was 38/18 mmHg with 0.35 mcg/kg/min milrinone support. Transesophageal echocardiogram (TEE) revealed well-placed unobstructed inflow cannula pointing toward the MV. The RV function was adequate. The aortic valve opened partially and intermittently. He was electively ventilated overnight and extubated next morning. He remained stable with a mean arterial pressure of 76 mmHg, cardiac output (CO) of 6.7 L/min with adequate urine output and central venous pressure (CVP) of 15 mmHg. He was commenced on aspirin 100 mg and warfarin. Heparin bridging was not used. Milrinone infusion in low dose was maintained for pulmonary vasodilation.

However, on that evening he had episodes of hypotension. His mixed venous saturation dropped to 36%, pulmonary wedge pressure and CVP rose to 46 and 38 mm of mercury, respectively. The cardiac index dropped to 1.5 L/square meter body surface area. The HM showed increase in power consumption to 8.7 watts suggesting pump thrombosis. The previous nonpulsatile flow pattern in the arterial line tracing became pulsatile. Bedside TEE could not demonstrate pump flow, revealing full opening of aortic valve with every beat. A CT scan confirmed pump obstruction with no contrast in inflow and outflow conduits.

At emergency resternotomy, the HM with inflow and outflow conduits was found completely clotted. The heart had contracted and was severely hypertrophic. The LV was no more cavernous. The Inflow conduit was blocked by the prolapsed anterior hypertrophied papillary muscle confirming inflow obstruction. The clotted pump with the outflow graft was replaced. The LV inflow site was closed with 2 'O' pledgeted nonabsorbable polyester braided sutures. A new inflow site was created little laterally to point the inflow more medially to avoid the anterior papillary muscle. On completion, the new pump was found to be obstructed by the thickened anterolateral free wall by TEE. A third ventriculotomy with the coring knife was created through the diaphragmatic surface. The ventricular cavity could admit only a finger and was obstructed by the hypertrophied papillary muscles, trabeculae and the septum. The obstructing muscles were resected leaving free-floating mitral leaflets. On completion, good unobstructed pump flow was obtained. But soon RV dysfunction occurred. By then the CPB had exceeded 5 h. CentriMag (Thoratec Corp) RV support with an oxygenator was instituted, using PA and RA cannulation.

The Oxygenator was weaned off 4 days later. The RV support was withdrawn on day 14 . The HM support remained stable with pump flow of 5 L/min, PI of 4, 6.6 watts and 9200 revolutions per minute (RPM). He was discharged home on 41st postoperative day. The AV on TTE, remained closed throughout the cardiac cycle with mild central regurgitation. MV was flail with severe regurgitation.

He remained well for 4 months, when he presented with progressive AR and dyspnea. Transcatheter aortic valve implantation with CoreValve (Medtronic CoreValve LLC, Santa Ana, CA) failed due to CoreValve migrating into the left ventricular outflow tract. Emergency resternotomy with explantation of the CoreValve, aortic valve stitch closure and tricuspid valve annuloplasty was performed. He recovered well and underwent successful heart transplantation 6 months later.

Discussion

HCM is a relatively common genetic disease defined as myocardial hypertrophy of more than 1.5 cm, without an identifiable cause such as long-standing hypertension and aortic stenosis [1]. The diagnosis is made on TTE. Biagini et al. [2] have reported a prevalence of dilated hypokinetic evolution in 4.9%, amongst 222 consecutive HCM patients, prospectively evaluated with a mean follow-up of 11 to 9 years. The first signs of LV dilation and hypokinesis were noted from 5 to 33 years after the diagnosis of HCM.

Our patient did not have family history of HCM, nor history of hypertension and alcoholism. The possible clue for HCM was the TEE measurements of IVSs of 1.34 cm and LVPWs of 1.69 cm in the dilated LV. At the time of his initial presentation, our focus was on the dilated LVIDd of 9.41 cm and heart failure. The noticeable feature at the time of LVAD implant was the unusual thickness of the excised LV apical core muscle, unlike in dilated cardiomyopathy. The histological characteristics of HCM include disorganization of the muscle bundles with characteristic whorling and interstitial fibrosis, myocyte, and myofibrillar disarray [3]. Our patient's histology with the dilated heart revealed hypertrophied muscles with cytoplasmic vacuoles and fibrosis without any whorling.

Our diagnosis of HCM is based on the morphological features of the heart at the time of pump exchange where the dilated "burnt out HCM" had shrunken down on LV decompression by a continuous flow pump. This feature suggests that dilated end-stage HCM can revert back to the morphology of HCM on decompression. Regression of dilated-hypokinetic HCM by biventricular cardiac pacing has also been reported in the literature [4].

It may be argued that RV failure or hypovolemia could have resulted in inadequate LV filling that resulted in a suck down event, leading to pump obstruction and pump thrombosis. But this patient had adequate RV function preoperatively and was able to cope with a PA pressure of 56 mmHg. His RV stroke work index was 4.69 gm-m/beat. We had no difficulty in transferring the patient to LVAD support from CPB initially. His postoperative PA pressure was 38/18 mmHg with a CVP of 12 mmHg. He did not have excessive blood loss. His CVP and PA pressure began to rise with episodes of hypotension, after the onset of pulmonary edema with the pump thrombosis. At the time of pump exchange, the contracted LV cavity could admit only one finger requiring extensive resection of the septum, trabecular, and the papillary muscles, suggestive of HCM.

Conclusion

Dilated hypokinetic evolution is an uncommon progression of HCM. The diagnosis in a patient presenting with heart failure will be difficult unless HCM has been diagnosed before or if the patient has a family history of HCM. Echocardiographic finding of thick-walled LV or IVS in a dilated heart may offer a clue. One has to be aware of the possibility of reversal of LV morphology to that of HCM when a continuous flow LVAD is implanted.

Conflict of Interest

None declared.

References

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