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CASE REPORT

Contrast-enhanced computed tomography with myocardial three-dimensional printing can guide treatment in symptomatic hypertrophic obstructive cardiomyopathy

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Abstract

Both surgical myectomy and percutaneous transluminal septal myocardial ablation are effective treatments for drug-refractory symptomatic hypertrophic obstructive cardiomyopathy (HOCM). However, in some cases, it is not easy to elucidate the abnormal structure of left ventricular outflow obstruction to adopt these treatments. Here, we presented a young female patient with drug-refractory symptomatic HOCM. In this case, contrast-enhanced computed tomography enabled us to assess the suitability of percutaneous transluminal septal myocardial ablation. By creating three-dimensional printed models using computed tomography data, we could also visualize intracardiac structure and simulate the surgical procedure. A multimodality assessment strategy is useful for evaluating patients complicated with drug-refractory symptomatic HOCM.

Keywords Hypertrophic obstructive cardiomyopathy; 3D printing; Surgical myectomy

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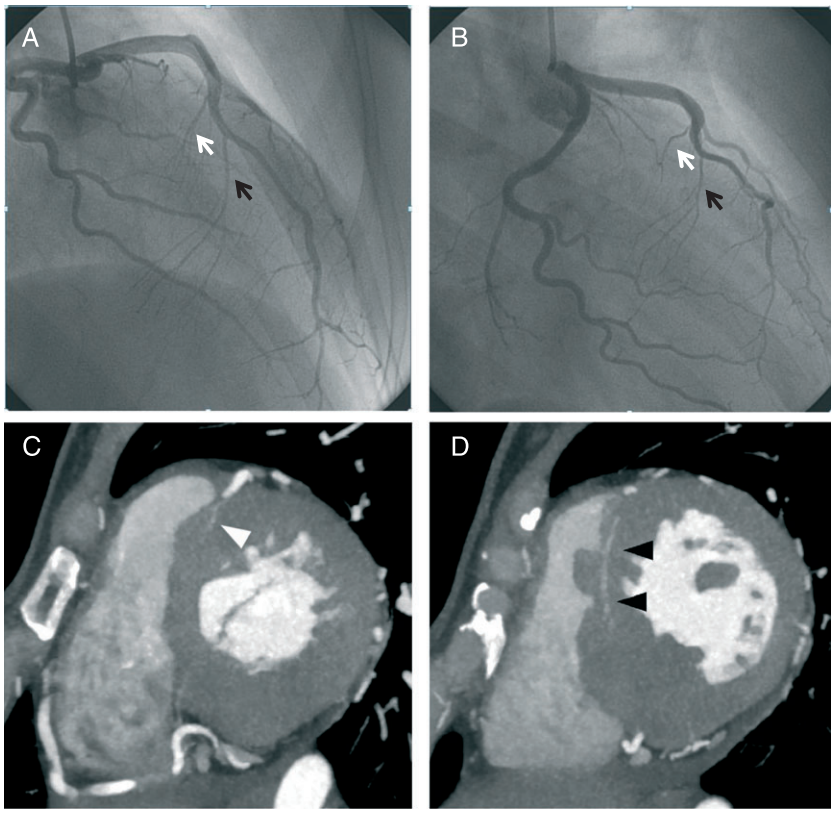
Introduction

Cardiac imaging is of value in the management of hypertrophic obstructive cardiomyopathy (HOCM). Contrast-enhanced computed tomography (CT) has enabled us to visualize the entire coronary tree with diagnostic image quality and assess the suitability of percutaneous transluminal septal myocardial ablation (PTSMA). Besides, three-dimensional (3D) printing model created by CT data is able to reproduce complex cardiac anatomy and is useful for surgical planning.

Case Report

A 41-year-old woman previously diagnosed with HOCM was referred to our hospital with dyspnoea on exertion and presyncope. Echocardiographic findings showed an asymmetrically hypertrophied left ventricle with maximal thickness of 23 mm in the posterior portion. Systolic anterior motion of the mitral valve was observed, causing severe mitral regurgitation. The peak pressure gradient at the left ventricular outflow tract (LVOT) was 148 mmHg at rest. The level of brain natriuretic peptide (BNP) was elevated to 1200 pg/mL. Left heart catheterization revealed that there was a high LVOT pressure gradient of 131 mmHg (left ventricle 216/24 mmHg; aorta 85/45 mmHg) at rest. Coronary angiography demonstrated no significant coronary artery stenosis, and two septal branches (small first septal branch and large second septal branch) of the left anterior descending coronary artery (*Figure 1A,B*). Because of documentation of non-sustained ventricular tachycardia and the history of presyncope, an implantable cardioverter defibrillator (ICD) was planned to

Figure 1 Coronary angiography and computed tomographic short axis view. (A) Right anterior oblique cranial view. (B) Right anterior oblique caudal view. White arrow indicates the first septal branch, and black arrow indicates the second septal branch. (C) The first septal branch (white arrow) supplies the myocardium related to the left ventricular outflow tract obstruction. (D) The second septal branch (black arrow) supplies the myocardium at the inferior-posterior mid portion.



implant. Before implantation of an ICD, electrocardiogram-gated contrast-enhanced CT was performed using a Siemens SOMATOM Definition Flash 2*128-slice computerized tomography scanner (Siemens Medical Solutions, Forchheim, Germany) to identify the septal branch and to assess whether the area supplied was equivalent to the area of maximum flow acceleration. Contrast-enhanced CT clearly revealed that the septal branch supplying the myocardium that was responsible for the LVOT obstruction was a small first septal branch and was not suitable for PTSMA (*Figure 1C,D*). After implantation of an ICD, on top of cibenzonline, bisoprolol, and verapamil were added at maximum tolerated dose. Repeated echocardiography demonstrated that LVOT obstruction and severe mitral regurgitation did not improve, and LVOT peak pressure gradient was 130 mmHg. Right ventricular pacing did not improve the LVOT pressure gradient. Despite a persistent high pressure gradient, her symptoms slightly improved, and she was discharged.

Three months later, the BNP level increased to 2300 pg/mL, with recurrent episodes of presyncope. The patient was readmitted to our hospital. Despite maximum tolerated medical therapy, a high pressure gradient still remained with a peak pressure gradient of 186 mmHg. Transesophageal echocardiography showed systolic anterior motion of the mitral valve and mitral valve leaflet-septal contact, relatively thin septal wall thickness, severe mitral regurgitation, and abnormal attachment of the chordae (*Figure 2*). We discussed the treatment strategy and concluded that surgical procedure was a better treatment strategy than PTSMA for the patient.

For better understanding of the left ventricular anatomy and surgical planning, 3D printing of the heart was performed using previously collected data of contrast-enhanced CT performed to assess the suitability of PTSMA. A stereolithography file of myocardial model was generated using the software Mimics (Materialize, Leuven, Belgium) and exported to a 3D printer system (RM-6000 II, CMET; Yokohama, Japan). 3D printing enabled better visualization of the geometric relationship among the hypertrophied myocardium, papillary muscle, and abnormal chordae (*Figure 3A–C*). 3D printing clearly showed abnormally thickened chordae and indicated that we could resect this abnormal structure. Furthermore, we simulated the surgical procedure using a 3D printed model (*Figure 3D*). We

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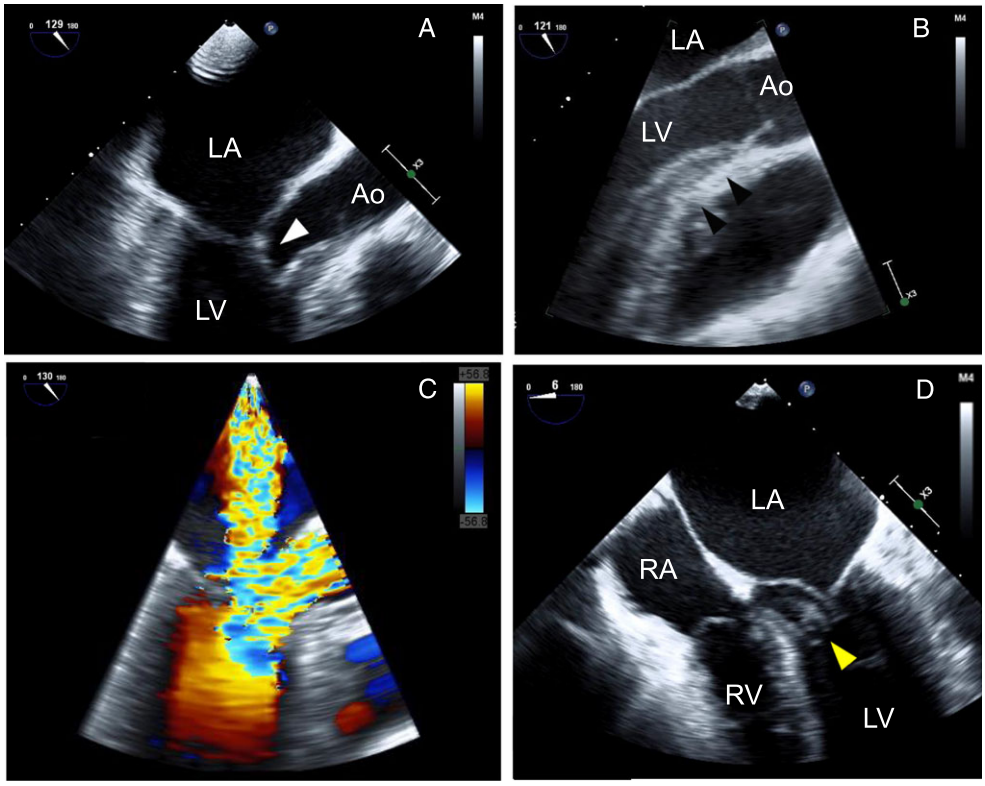
Figure 2 Transesophageal echocardiography before surgery. (A) Systolic anterior motion of the mitral valve, and mitral valve leaflet-septal contact were observed (white arrow). (B) Relatively thin septal wall (maximum wall thickness 17 mm) was observed (black arrow). (C) Severe mitral regurgitation and acceleration flow at the left ventricular outflow tract were observed. (D) Abnormal chordae were observed (yellow arrow). Ao, aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.



examined the extent to which we could resect the myocardium, because the patient had relatively thin septal wall thickness. By simulation, we could determine the amount of myocardium that should be excised.

Extended septal myectomy was performed via a trans-aortic approach (Morrow procedure), and the hypertrophied septum was excised. Despite poor visualization of the left ventricular cavity, hypertrophied abnormal chordae, which were visualized by 3D printing, could be also excised as planned preoperatively. By simulating the surgical procedure, we could excise a sufficient amount of myocardium at the actual operation without any complications. Mitral valve plication of the posterior leaflet was performed to achieve a height reduction of the coaptation point of the mitral leaflet (*Figure 4*).

The postoperative course was uneventful. Transesophageal echocardiography before discharge showed that systolic anterior motion of the mitral leaflet had improved from the preoperative state. Mitral regurgitation and acceleration flow at LVOT were also improved. The pressure gradient across the LVOT was markedly improved, from 186 to 24 mmHg. BNP level decreased from 2300 to 1000 pg/mL. The patient was discharged uneventfully at 14 days after operation without any symptoms. She was asymptomatic on exertion, without an episode of presyncope, on review at our outpatient clinic.

Discussion

Septal reduction therapies, including surgical myectomy and PTSMA, are effective treatments for drug-refractory symptomatic HOCM patients.[1,2] A debate on the relative merits of these two strategies has been going on over this period of time.[3] We sometimes have difficulty deciding which treatment strategy to perform, and preoperative evaluation is very important.

Recently, some reports have documented the usefulness of contrast-enhanced CT for noninvasive evaluation of HOCM patients prior to PTSMA.[4–6] Contract-enhanced CT enables us to identify the proximal septal perforator and to assess whether the area supplied is equivalent to the area of maximum flow acceleration. As in previous reports, we

Figure 3 Three-dimensional (3D) printed model of hypertrophic obstructive cardiomyopathy. (A) through (B) 3D printing showed left ventricle geometry. (C) Intracardiac geometry. Black arrow shows abnormally thickened chordae. (D) Simulation of myectomy using 3D printed model.

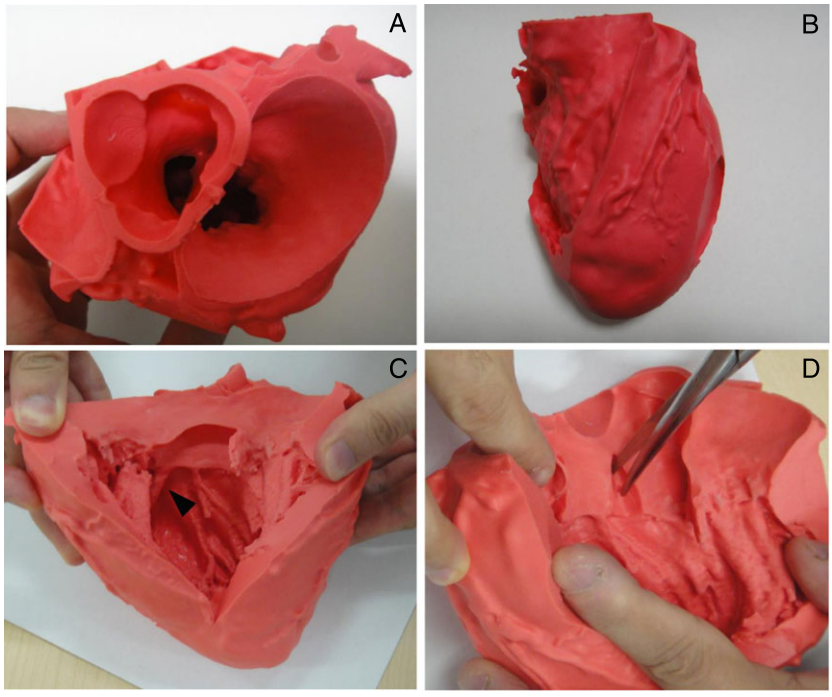
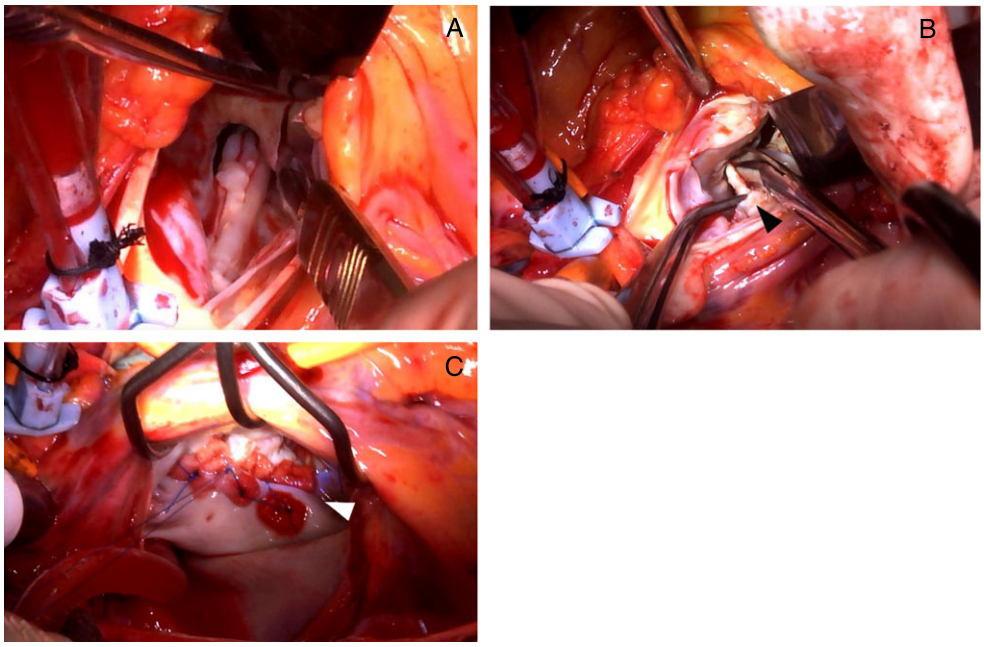


Figure 4 Intraoperative photography. (A) The view is from the aortic side looking into the left ventricle. The surgeon is cutting the septal myocardium using a surgical scalpel. Hypertrophied septal wall was resected. (B) Abnormal chordae were also resected (black arrow). (C) Plication of posterior mitral valve was performed (white arrow).



performed contrast-enhanced CT to assess the feasibility of PTSMA in this case. We could conclude that PTSMA might not be effective in the patient based on the results of contrast-enhanced CT.

Surgical myectomy is the gold standard and safest septal reduction therapy for most severely symptomatic patients with HOCM refractory to maximum medical management. Surgical myectomy can be performed with minimal operative mortality and morbidity in experienced centers; however, the number of operation is so limited that it is hard to arrange a concrete plan to alleviate the LVOT obstruction for less-experienced surgeons. In addition, surgical procedure is difficult because of poor visualization of the left ventricular cavity in the surgical field and heterogenous LVOT anatomy. 3D printing is an emerging technology in various fields and is able to reproduce complex cardiac anatomy,[7,8] so this tool could be of value in the planning of surgical treatment for selected cases. To date, there are only two reports describing the usefulness of 3D printing in HOCM patients.[9,10] Both reports showed that a good postoperative course could be achieved using 3D printing.

In our case, by performing only one contrast-enhanced CT scan, we could assess the suitability of PTSMA and also could create 3D printed models of the heart. By using 3D printed models, we could achieve better visualization of complex intracardiac morphology and mitral apparatus. We could confirm the relatively thin septal wall and abnormal chordae and thereafter simulated the surgical procedure, resulting in a good postoperative course in this young female patient. In conclusion, performing contrast-enhanced CT to assess the suitability of PTSMA and to create a 3D printed model of the heart in the same sequence can visualize intracardiac structure and simulate the surgical procedure. A multimodality assessment strategy using contrast-enhanced CT and 3D printing is useful for treating patients complicated with drug-refractory symptomatic HOCM.

Conflict of interest

None declared.

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