CASE REPORT

Dynamic left ventricular outflow obstruction in situs inversus with levocardia

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SUMMARY

A 69-year-old man with situs inversus, levocardia and inverted great arteries developed severe dynamic left ventricular outflow tract (LVOT) obstruction with systolic anterior motion of the anterior mitral leaflet. There was no asymmetric septal hypertrophy. A possible mechanism of the LVOT obstruction in the present case may have been related to an abnormally long and bent outflow tract resulting from overriding of the right ventricle over the LVOT due to a congenital malposition of the heart. Mitral valve replacement with septal myectomy was performed in order to eliminate systolic anterior motion. The postoperative course has been excellent.

BACKGROUND

Congenital heart disease in adults is an important entity as many children with congenital heart disease now live longer and cardiologists have increased opportunity to take care of these patients as adults. Left ventricular outflow tract (LVOT) obstruction associated with adult congenital heart disease may be caused mainly by fixed anatomical lesions such as discrete subaortic stenosis due to fibrous membrane, or tunnel-type obstruction with thick fibromuscular tubular narrowing of the LVOT, or abnormal accessory mitral tissues. We report a rare case of dynamic LVOT obstruction in a patient with situs inversus, levocardia and inverted great arteries. A possible pathophysiological mechanism is proposed.

CASE PRESENTATION

A 69-year-old man was successfully resuscitated from an episode of ventricular fibrillation and admitted to our hospital for evaluation and treatment of dynamic LVOT obstruction associated with congenital heart disease.

At 35 years of age, the patient had suffered from brain abscess complicated by posterior lobe infarction and bilateral blindness. A ventriculoperitoneal shunt was performed for obstructive hydrocephalus. The patient noticed shortness of breath during exercise at 46 years of age. At the time, detailed cardiac evaluation revealed situs inversus (inverted position of the visceral organs) and levocardia (the base-to-apex axis pointing to the left). A coincidental atrial septal defect (ASD) with pulmonary to systemic blood flow ratio (Qp/Qs) of 1.7 was found. At that time, there was no pressure gradient at the LVOT and coronary arteries were normal. The patient had been stable with medical follow-up, until he was 56 years of age, when he started to notice chest discomfort and worsening of

dyspnoea. Auscultation of the heart showed a grade 3 systolic ejection murmur at the lower sternal border. Transthoracic echocardiogram revealed systolic anterior motion (SAM) of the anterior mitral leaflet (AML) resulting in dynamic LVOT obstruction and moderate mitral regurgitation. Because his ordinary daily activity was so limited due to blindness (he walked with assistance and mainly used a wheel chair), surgery was deferred and he was started on a β -blocker.

At 69 years of age, the patient suddenly collapsed at home and was immediately resuscitated by his wife. An automatic external defibrillator operated by emergency personnel successfully converted ventricular fibrillation back to normal sinus rhythm about 15 min after the collapse. He was taken to the emergency room at a nearby hospital, where intravenous amiodarone was administered. After stabilisation, 2 weeks later, he was transferred to our hospital for further evaluation and treatment.

At our hospital, vital signs showed blood pressure of 130/80 mm Hg, heart rate of 70 bpm and respiratory rate of 18 breaths/min. Grade 3 systolic ejection murmur was noted in the precordium. Chest X-ray revealed cardiomegaly and right aortic arch (figure 1A). The apex of the heart was located to the left. Situs inversus was suspected as a gastric bubble was located in the right side. An ECG showed normal sinus rhythm, right axis deviation, tall R waves in V1 and V2, and T-wave inversion in V1–4 (figure 1B). A chest CT scan (figures 2A–D and 3) showed that the right ventricular inflow was located to the left posteriorly and the left ventricular inflow was located to the right anteriorly (L-loop). Also, the right ventricle over-rode on the LVOT, creating an over-and-under position (figures 2 and 3A–C). The spleen and the stomach were located on the right, while the inferior vena cava and superior vena cava were connected to the right atrium on the left (figure 2D). The connection of the atria, ventricles and great vessels were normal. The left ventricle was ‘banana-shaped’ and there was no asymmetric septal hypertrophy (ASH) (figure 2C). Transthoracic echocardiography revealed SAM of the AML and moderate mitral regurgitation (figure 3D). Angiographic examination confirmed the malposition of the heart suggested by CT. The left ventricle and aorta made a right angle and LVOT was long and narrow (figures 3A and 4A). There was a severe pressure gradient at the LVOT (figure 4C). An ASD was also noted and Qp/Qs was 1.6. A membranous structure suggestive of cor triatriatum (where the left atrium is subdivided by a thin membrane, resulting in 3 atrial

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Figure 1 Chest X-ray revealed that the heart was located on the left. (A) Cardiomegaly and right aortic arch (white arrow) were noted. A gastric bubble was located on the right (black arrows). (B) Twelve-lead ECG showing normal sinus rhythm, right axis deviation, tall R in V1 and diffuse T wave inversions.

chambers) was also noted (figure 3A). There was no significant coronary artery stenosis, and pressures of the heart chambers were normal. An electrophysiology study was not performed because the episode of ventricular fibrillation was already documented. Because of severe hemodynamic abnormality, the patient had surgical treatment and implantable cardioverter defibrillator (ICD) implantation.

TREATMENT

Because there was no significant ASH, septal myectomy alone was thought to be insufficient and difficult. Mitral valve replacement with bileaflet artificial valve prosthesis was performed in addition to septal myectomy. The ASD was closed and the coincidental cor triatriatum was resected. Because of the possible

difficulty and uncertainty of intravenous lead implantation, surgical lead placement was performed. ICD sensor leads were sutured on the surface of the left ventricle and patch electrodes were placed, wrapping around the heart. The entire surgical procedure was performed by cardiac surgeons, as our hospital does not have a specialised team for adult congenital heart disease.

OUTCOME AND FOLLOW-UP

After surgical treatment, the patient's symptoms and haemodynamic condition improved dramatically. On echocardiogram, the systolic ejection murmur and pressure gradient across the LVOT had disappeared. The size of the heart on chest X-ray was also improved. To date, the patient has been maintained on

Figure 2 (A–D) Contrast-enhanced CT scan of the chest and abdomen. The right side of each image corresponding to the left side of the body and vice versa. The liver is located on the left, and the stomach as well as spleen are located on the right (situs inversus). IVC is seen connected to the right atrium on the left side. The inflow of the anatomical right ventricle is located to the left relative to the left ventricle showing an L-loop. The left ventricle is located more anteriorly. There was no discordance of the atria, ventricles and great arteries (Ao, aorta; L, Liver; LA, left atrium; LV, left ventricle; PV, pulmonary vein; RA, right atrium; RV, right ventricle; RVOT, RV outflow tract; SP, spleen; ST, stomach).

Figure 3 (A and B) Reconstructed images of the long axis (A) and short axis (B) view of the LV. LV is located inferiorly and anteriorly relative to the RV. The RV is located superiorly, over-riding on the LVOT, making the relation an over-and-under position. The angle between the LV and Ao makes a right angle resulting in a long and narrow LVOT (A). A white arrow indicating calcification of the posterior mitral annulus. A black arrow indicating a membranous structure in the left atrium (cor triatriatum). (C) Volume rendering image of the LV and RV showing over-riding of the RV on the LVOT. (D) Apical three-chamber view of the transthoracic echocardiogram, showing systolic anterior motion (two small white arrows) resulting in AML-septal contact. (Ao, aorta; AML, anterior mitral leaflet; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; PA, pulmonary artery; RV, right ventricle).

Figure 4 (A) Left ventriculogram showing abnormal bending of the LVOT. The LV is 'banana' shaped. There is significant mitral regurgitation associated with LVOT obstruction. EF was 65% and was normal. Moderate mitral regurgitation was noted. (B) Right atrial angiography showing the RV located posteriorly and superiorly relative to the LV. (C) Significant pressure gradient across the LVOT. A pigtail catheter was pulled back from the apex to ascending Ao, and the pressure gradient was localised at the LVOT. There was no pressure gradient at the aortic valve and mid ventricle. The severity of the LVOT is seen (Ao, aorta; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; PA, pulmonary artery; RA, right atrium; RV, right ventricle; WP, pulmonary artery wedge pressure).

amiodarone and has been stable. Analysis of ICD memory has recalled no episode of ventricular tachyarrhythmias after surgery.

DISCUSSION

This case had a very rare congenital anomaly of situs inversus with levocardia.^{1 2} The incidence of situs inversus is estimated to be 1 in 10 000–25 000 births. The incidence of situs inversus with levocardia is much rarer. The position of the viscera and atrium was inverted, and the position of the anatomical right ventricular inflow tract was located in left side relative to the left ventricle (L-loop), while no transposition of the great arteries was noted. So there was no anatomical or functional discordance in relation to the atria, ventricles and great arteries (situs inversus of viscera and atria, L-loop of the ventricles, inverted great arteries: ILI in Van Praagh's symbolic convention).³ ASD and cor triatriatum were coincidental congenital anomalies. In patients with cardiac malposition, LVOT obstruction has been reported in transposition of the great arteries,⁴ situs inversus totalis^{5 6} and anatomically corrected malposition⁷ (situs solitus, D-loop, L-malposition). A case with LVOT obstruction in a patient with situs inversus with levocardia has never been reported. Usually, LVOT obstruction in adult patients with congenital heart disease results from fixed anatomical lesions such as discrete subaortic stenosis due to fibrous membrane, or tunnel-type obstruction with thick fibromuscular tubular narrowing of the LVOT, or abnormal accessory mitral tissues.⁸ In contrast, hypertrophic obstructive cardiomyopathy features a dynamic obstruction of the LVOT, usually associated with SAM of the AML. Structural and functional factors both contribute to LVOT obstruction.^{9 10} ASH, sigmoid septum, progressive narrowing of the LVOT during systole, and direction and velocity of the bloodstream, cause drag forces on the AML, resulting in SAM of the AML. This movement causes the AML to come in contact with the septum, obstructing the LVOT. The gradient in the LVOT may increase dramatically with decreased afterload or increased contractility. A possible mechanism of LVOT obstruction in the present case may be related to an abnormally long and bent outflow tract resulting from over-riding of the right ventricle over the LVOT (figure 3A–C). The right angle of the direction of the blood stream combined with these inborn anatomical factors may have caused drag forces predisposing to SAM of the AML. In addition, calcification of the posterior mitral annulus might have contributed to pushing the entire mitral apparatus anteriorly, narrowing the LVOT. Because there was no ASH, the surgery aimed to correct SAM by replacement of the mitral valve with a low-profile bileaflet mitral valve prosthesis.¹¹ After the surgery, the chest discomfort and dyspnoea disappeared dramatically. The heart murmur also disappeared and there was no LVOT pressure gradient in serial echocardiographic evaluation. The episode of sudden collapse appeared to have been more closely related to a severe haemodynamic

abnormality, as no further episode of serious ventricular arrhythmias have been detected by ICD recall for 6 years after the surgery.

Learning points

- Congenital anomalies are sometimes accompanied by left ventricular outflow tract (LVOT) obstruction. In this case, dynamic LVOT obstruction was caused by a long and bent LVOT due to over-riding of the right ventricle on the LVOT in a patient with situs inversus and levocardia.
- Dynamic LVOT obstruction can be seen without asymmetric septal hypertrophy.
- Correction of a haemodynamic abnormality was very important to prevent serious ventricular arrhythmias.

Contributors TT and HM performed the surgery. MW contributed to taking care of the patient.

Competing interests None declared.

Patient consent Obtained.

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