Endocardial linear infarct exclusion technique for non-ischaemic functional mitral regurgitation caused by cardiac sarcoidosis: a case report

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Introduction
Damage to the posterior wall of the left ventricle (LV) can cause tethering mitral regurgitation (MR). We present a patient with non-ischaemic tethering MR and congestive heart failure due to cardiac sarcoidosis who was treated using an endocardial linear infarct exclusion technique.

Case presentation
A 63-year-old woman with a history of uveitis presented to our hospital complaining of dyspnoea. Echocardiography revealed dyskinesis of the posterolateral wall of the LV and severe tethering MR (regurgitation volume: 92 mL). The LV ejection fraction was reduced to 45%. Cardiac catheterization revealed no stenosis. Magnetic resonance imaging with late gadolinium enhancement revealed a contrast effect and thinning of the posterolateral wall. The abnormal accumulation was also observed with fluorodeoxyglucose-positron emission tomography. Together, these findings indicated cardiac sarcoidosis, and we determined that cardiac sarcoidosis had resulted in aneurysm development in the posterior wall of the LV, subsequent advanced tethering at the posterior mitral valve cusp, and severe functional MR. The patient underwent an endocardial linear infarct exclusion technique (ELIET), mitral annuloplasty, tricuspid annuloplasty, and the full MAZE procedure. Histopathological analysis of the posterior wall myocardium revealed marked thinning of the endocardium, replacement fibrosis, lymphocyte infiltration, and epithelialization. These findings were consistent with sarcoidosis. The patient’s condition improved to New York Heart Association (NYHA) Class I, and cardiac events were rare at 6 months after surgery.

Discussion
Endocardial linear infarct exclusion technique is useful for treating tethering MR. To our knowledge, this is the first reported case of successful treatment using ELIET for non-ischaemic tethering MR caused by cardiac sarcoidosis.

Keywords
Mitral regurgitation • Non-ischaemic functional mitral regurgitation • Cardiac sarcoidosis • Case report

Learning Points
• Cardiac Sarcoidosis caused non-ischaemic tethering mitral regurgitation (MR).
• Endocardial linear infarct exclusion technique (ELIET) is useful for treating tethering MR.
• Endocardial linear infarct exclusion technique shortens the papillary muscle length, which improved tethering of the mitral leaflet. Beyond resolving left ventricle enlargement between the papillary muscles that was caused by the aneurysm of the posterior wall.
Introduction

Cardiac sarcoidosis involves focal myocardial injury. When it develops in the posterolateral wall of the heart, the patient often presents with severe tethering mitral regurgitation (MR). Herein, we report the case of a patient with cardiac sarcoidosis with severe functional MR and dyskinesis of the posterior wall that was treated successfully using the endocardial linear infarct exclusion technique (ELIET).

Timeline

| Date         | Event                                      |
|--------------|--------------------------------------------|
| 12/10/2016   | Patient became aware of dyspnoea            |
| 1/30/2017    | Patient became aware of pretibial pitting oedema |
| 2/1/2017     | Patient was admitted to our hospital and diagnosed with congestive heart failure (B-type natriuretic peptide (BNP): 1141 pg/mL) and patient was hospitalized. |
| 3/10/2017    | Patient’s condition worsened (BNP: 2601 pg/mL) and patient was hospitalized. |
| 4/18/2017    | We performed precise examinations and diagnosed with cardiac sarcoidosis |
| 4/24/2017    | Operation was performed                     |
| 5/11/2017    | Discharge                                   |
| 12/8/2017    | Latest follow-up echocardiography (BNP: 291 pg/mL) |

Case presentation

A 63-year-old woman with a history of hypertension, hyperuricemia, and uveitis presented to our hospital complaining of dyspnoea upon exertion (NYHA Class II). We heard coarse crackling in both lungs, and systolic murmur of the Levine IV/VI within the apex. Mild pitting oedema was detected on both legs. Laboratory tests indicated an elevated B-type natriuretic peptide (BNP) value of 2601 pg/mL (normal < 18.4 pg/mL). A chest X-ray showed a 62% increase in the cardiothoracic ratio (normal < 50%).

Transhthoracic echocardiography showed dyskinesis of the posterior wall of the left ventricle (LV) accompanied by thinning. The left chamber diameter was increased [LV diastolic diameter (normal 30–50 mm), 66 mm; LV systolic diameter (normal 20–30 mm), 51 mm] and the LV ejection fraction (LVEF) (normal 55–80%) was reduced to 33.5%.

A transoesophageal echocardiogram (TOE) revealed that contraction of the posterior wall caused significant tethering of P2, thereby resulting in severe functional MR. (Figure 1D). The regurgitate volume was 92 mL (severe: >60 mL) (Figure 1A–C).

Computed tomography (CT) and magnetic resonance imaging with late gadolinium enhancement revealed contrast effect and thinning of the posterior wall (Figure 2), thereby suggesting that it was not viable. The abnormal accumulation was also observed on fluorodeoxyglucose-positron emission tomography (Figure 2B). We suspected cardiac sarcoidosis due to the presence of wall motion abnormalities that were confined to portions of the LV. The patient’s uveitis was confirmed to be iris adhesions, which is the characteristic ocular lesion of sarcoidosis. Coronary angiography was performed to exclude ischaemic heart disease, and no significant stenosis was detected. The patient experienced a period of heart failure, during which her BNP increased to 2601 pg/mL (normal < 18.4 pg/mL). This patient had moderate TR (TR pressure gradient 49 mmHg) associated with mild right heart failure [TAPSE (normal > 16 mm): 14 mm; FAC (normal > 35%): 32% with accompanying tricuspid valve annulus expansion (47 mm)], such that intervention was necessary. Pre-operative cardiac catheter data after heart failure treatment using a diuretic was as follows: Pulmonary artery (PA) (normal 15–30/3–12 mmHg): 33/12 (20) mmHg; Right ventricle (RV) (normal 15–30/2–8 mmHg): 34/1 (3) mmHg; Right Atrium (RA) (normal 2–8 mmHg): 5/2 (2) mmHg; pulmonary capillary wedge pressure (normal 2–15 mmHg): 6/6 (5) mmHg.

The surgery was performed using ELIET, mitral annuloplasty, tricuspid annuloplasty, and the MAZE procedure. Initially, an incision was made in the LV at the centre of the scar area along the coronary artery. The first layer was linearly closed with 3-0 polypropylene continuous sutures to approximate the border between the normal and infarcted myocardium. The second layer was closed with a combination of interrupted and continuous sutures (Figure 3). The cardiopulmonary bypass time was 273 min, and the aortic cross clamp time was 132 min.

Histopathological analysis of the resected posterior wall myocardium revealed thinning of the endocardium, replacement fibrosis, lymphocyte infiltration, and epithelialization (Figure 4A). CD68 staining was positive (Figure 4B). Together, these findings are consistent with sarcoidosis. An echocardiogram taken 6 months after surgery revealed reversal of remodelling and a disappearance of the tethering. There was no recurrence of MR and no signs of heart failure.

In the TOE, the pre-tenting height/PL angle was 13 mm/56°, which was reduced to 10.2 mm/23° post-operatively. These data showed improvements in tethering. Additionally, MR had not recurred during the 6 months of follow-up. Additionally, post-operative CT scans can indicate the efficacy of linear aneurysm resection compared to Dor-plasty, since the sphericity index was decreased from 0.75 to 0.61 (normal 0.45–0.62) (Figure 2C). There were no post-operative complications or symptoms of heart failure. Since this patient did not have a diagnosis of sarcoyiosis before being admitted to our hospital, there was no history of relevant medical treatment before surgery. Corticosteroid treatment has been used as a therapeutic option for patients with cardiac sarcoidosis, and the initiation of corticosteroid therapy before systolic dysfunction develops produces excellent clinical outcomes. Thus, prednisolone therapy was performed post-operatively in this case.

Discussion

In functional MR, there is no conclusive evidence that mitral valve intervention was associated with a survival benefit. When mitral annuloplasty with downsizing was performed, it was reported that tethering of the posterior leaflet was reinforced and MR recurred. Therefore, in the American Heart Association guidelines, chordal-sparing mitral valve replacement (MVR) is recommended for patients with severe disease (NYHA Class III or above). However, valve replacement carries the risk of rupturing the LV when its myocardium...
Figure 1 Echocardiographic data: (A, B) Transthoracic echocardiography showed dyskinesis of the posterior wall of the left ventricle accompanied by thinning. (C) The expansion of the annulus caused by the enlargement of the left ventricle, also merged the effective regurgitant orifice of 0.71 cm². (D) Transoesophageal echocardiogram revealed that contraction of the posterior wall caused significant tethering of P2, resulting in severe functional mitral regurgitation. (D) The regurgitant volume was 92 mL.

Figure 2 Computed tomography and magnetic resonance imaging. (A) Revealed contrast effect and thinning of the posterior wall. (B) The abnormal accumulation was also observed on fluorodeoxyglucose-positron emission tomography. (C) Computed tomography after endocardial linear infarct exclusion technique; post-operative computed tomography scans can indicate the efficacy of linear aneurysm resection compared to Dor-plasty, since the sphericity index was decreased from 0.75 to 0.61. (D) Magnetic resonance imaging with late gadolinium enhancement revealed contrast effect and thinning of the posterior wall suggesting it was not viable.
is thin. Moreover, this does not indicate that MVR is more suitable than the subvalvular procedure for repairing mitral valve prolapse. Several studies describing the use of the subvalvular procedure with good results have been described. Myocardial infarction of the posterior wall of the LV causes the myocardium between the mitral valve annulus and papillary muscle attachment on the posterior wall of the LV to become thin and expand, thereby resulting in ischaemic MR, for which left surgical ventricular reconstruction (SVR) of the LV posterior wall is the most effective treatment. Yaku et al. developed a novel method of surgical ventricular restoration for an infarcted posterolateral wall. The left ventricular lumen morphology affects wall stress (i.e. wall stress decreases as the lumen elongates and becomes larger, thereby closely approximating a spherical shape). A study of dilated cardiomyopathy, which is essentially an enlarged lumen, demonstrated that the left ventricular cavity also becomes spherical in shape. It has been reported that contractility decreases with such changes to lumen morphology, as lumen morphology is related to wall stress and cardiac function.

During Dor-plasty, which is as commonly performed as left ventriculoplasty, the infarct part is incised, and a tobacco suture is

**Figure 3** Surgical procedure of endocardial linear infarct exclusion technique; (A, B) Initially, an incision was made in the left ventricle at the centre of the scar area along the coronary artery. (C) The first layer was linearly closed with 3-0 polypropylene continuous sutures to approximate the border between the normal and infarcted myocardium. (D) The second layer was closed with a combination of interrupted and continuous sutures. (E) Schematic diagram of endocardial linear infarct exclusion technique seen in left ventricular short axis cross section.

**Figure 4** Histopathological analysis of the resected posterior wall myocardium revealed thinning of the endocardium, replacement fibrosis, lymphocyte infiltration, and epithelialization (haematoxylin and eosin staining) × 40 (A). CD68 staining was positive × 400 (B).
constructed on the side of the left ventricular lumen. Once this area is stitched, the left ventricular long axis is shortened after the operation, which decreases the tendency of the ventricle to develop into a spheroid.

During the ELIET, which we used in this case, the LV was shortened along the short axis rather than along the long axis by cutting the infarcted part longitudinally and suturing circumferentially. In the original report describing this procedure, the sphericity index increased in both groups, but the rate of increase tended to be larger in the Dor-plasty group than in the ELIET group.14

In this patient, we speculated that cardiac sarcoidosis caused the LV posterior wall to form an aneurysm, thereby resulting in advanced tethering at the posterior mitral valve cusp and severe functional MR.1 One advantage of ELIET for the posterior all is that it shortens tethering at the posterior mitral valve cusp and severe functional LV posterior wall to form an aneurysm, thereby resulting in advanced in the Dor-plasty group than in the ELIET group.14

In this patient, we speculated that cardiac sarcoidosis caused the LV posterior wall to form an aneurysm, thereby resulting in advanced tethering at the posterior mitral valve cusp and severe functional MR.1 One advantage of ELIET for the posterior all is that it shortens the papillary muscle length, which leads to improved tethering of the mitral leaflet. Beyond resolving LV enlargement between the papillary muscles that was caused by the aneurysm of the posterior wall, the greatest benefit of the ELIET is that the SVR can restore the left ventricular volume and improve tethering. Additionally, by using the ELIET, the LV is incised, but not excised. Therefore, the coronary arteries are less likely to be sacrificed. The risk of bleeding is low because ventriculotomy is closed with two layers. Consequently, we can control severe MR with ELIET and mitral annuloplasty with no severe complications. As described herein, the procedure can promote good LV reverse remodelling without the recurrence of heart failure.

We successfully treated a case of cardiac sarcoidosis combined with severe functional MR. There are several valuable take-away from this case, including: (i) two key procedures in this operation (i.e. surgical ventricular restoration and the repair of tethering of posterior leaflet); (ii) applying the ELIET to the posterior wall shortens the papillary muscle length, thereby improving tethering of the mitral leaflet; and (iii) SVR and mitral valve plasty are still feasibly and the preferred treatment method, even in challenging and complex cases.

### Patient perspective

The patient was satisfied with her treatment and her only concern was regarding the recurrence of sarcoidosis, for which we prescribed prednisolone.

### Consent

The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

### Conflict of interest

none declared.

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