Koşuyolu Heart Journal

Koşuyolu Heart J 2024;27(2):93–96
DOI: 10.51645/khj.2024.462

Case Report

Patient Who Had Total Situs Inversus with Dextrocardia and A Partial Anterior Papillary Muscle Rupture Who Underwent Successful Mitral Valve Repair with Coronary Revascularization

Rezan Aksoy,* **Muhammed Ali Oya,** **Cüneyt Arkan,** **Murat Bülent Rabuş**

Department of Cardiovascular Surgery, University of Health Sciences, Kosuyolu High Specialization Training and Research Hospital, İstanbul, Türkiye

Abstract

Total situs inversus (TSI) with dextrocardia is an infrequent anatomical characteristic and anterior papillary muscle rupture (PMR) is one of the rare mechanical complications following myocardial infarction. A 56-year-old man who had a history of mitral valve regurgitation (MR) presented with dyspnea to the emergency department. Upon transesophageal echocardiography, there were dextrocardia and chordal flail in the A1 scallop. Coronary angiography showed a substantial stenosis of the left anterior descending artery (LAD). Computed tomography of the thorax and abdomen showed TSI. During the operation, dextrocardia and chordal flail with partial anterior PMR at the A1 segment were observed. Following the mitral valve repair (MVr) with an artificial neo-chord and the ring annuloplasty implantation, coronary revascularization (LAD-right mammarian artery) was performed. On the control TOE, he had no MR. Conclusion: Patients who had TSI with dextrocardia and partial anterior PMR can undergo successful MVr.

Keywords: Dextrocardia; mitral valve repair; papillary muscle rupture; total situs inversus.

Dekstrokardi, Total Situs İnversus ve Kısmi Anterior Papiller Kas Rüptürü Olan Hastaya Koroner Revaskülarizasyon ile Yapılan Başarılı Mitral Kapak Onarımı

Özet

Dekstrokardi ile birlikte olan total situs inversus (TSİ) nadir görülen bir anatomik özelliktir. Anterior papiller kas rüptürü (PKR)'de miyokard enfarktüsü sonrası nadir görülen mekanik komplikasyonlardan biridir. Mitral kapak yetersizliği (MY) öyküsü olan 56 yaşında erkek hasta acil servise nefes darlığı ile başvurdu. Transözofageal ekokardiyografide (TEE) dekstrokardi ve A1 skallobunda kordal flail vardı. Koroner anjiyografide sol ön inen arterde (LAD) önemli bir darlık görüldü. Toraks ve batın bilgisayarlı tomografisinde TSİ saptandı. Ameliyat sırasında dekstrokardi ve A1 skallobunda kısmi anterior PKR ile birlikte kordal flail gözlendi. Yapay neo-kord ve ring anüloplasti implantasyonunu ile mitral kapak tamirini takiben koroner revaskülarizasyon (LAD-sağ mammarian arter) yapıldı. Kontrol TEE'sinde MY gözlenmedi. Dekstrokardi ve parsiyel anterior PKR ile birlikte TSİ olan hastaya başarılı mitral kapak tamiri uygulanabilir.

Anahtar sözcükler: Dekstrokardi; mitral kapak tamiri; papiller adele rüptürü; total situs inversus.

Introduction

Papillary muscle rupture (PMR), one of the infrequent as well as significant mechanical complications following myocardial infarction (MI), is a condition associated with increased mortality, which

*The current affiliation of the author: Department of Cardiovascular Surgery, Dr. Siyami Ersek Thoracic Heart and Vascular Surgery Training and Research Hospital, İstanbul, Türkiye.

Cite This Article: Aksoy R, Oya MA, Arkan C, Rabuş MB. Patient Who Had Total Situs Inversus with Dextrocardia and A Partial Anterior Papillary Muscle Rupture Who Underwent Successful Mitral Valve Repair with Coronary Revascularization. Koşuyolu Heart J 2024;27(2):93–96.

Address for Correspondence:
Rezan Aksoy

Department of Cardiovascular Surgery, University of Health Sciences, Kosuyolu High Specialization Training and Research Hospital, İstanbul, Türkiye

E-mail: rezanaksoy@gmail.com

Submitted: July 6, 2024
Accepted: July 17, 2024
Available Online: August 26, 2024

has an incidence of 1–5%. It results in severe mitral valve regurgitation (MR), which is often accompanied by cardiogenic shock and pulmonary edema, requiring both urgent medical treatment and surgical intervention.[1]

Its surgical treatment includes either mitral valve replacement (MVR) by sparing the subvalvular apparatus or mitral valve repair (MVr).[1] Mitral valve replacement was first described in 1965 in patients with PMR.[2] The surgical technique of MVR has been performed approximately at a rate of 70–80% and that of MVr at a rate of 20–30%.[3] The reason for the increased performance rate of MVR is that myocardial tissue becomes fragile and edematous, also hemodynamically unstable in high-risk patients after MI.[4] However, MVr is superior to MVR with respect to improving left ventricular function. Concomitant coronary revascularization enhances not only short-term but also long-term outcomes.[5]

Total situs inversus (TSI) with dextrocardia is an infrequent anatomical characteristic, in which the heart lies in the middle of the mediastinum, with its base-to-apex axis directed to the right and caudally. Anterior PMR is one of the rare as well as significant mechanical complications following MI.[6] We present a case with TSI with dextrocardia and anterior PMR, who underwent successful MVr with coronary artery bypass graft (CABG) and closure of patent foramen ovale (PFO).

Case Report

A 56-year-old man who had a history of MR presented with dyspnea to the emergency department. He had no history of coronary artery disease. Auscultation showed that the patient had bilateral basal crepitations and a systolic murmur in the anterior left lower chest. Transthoracic echocardiography revealed dextrocardia and severe MR, with no ventricular segmental dyskinesia. On transesophageal echocardiography (TOE), there was a chordal flail at the A1 segment, with an ejection fraction of 65%. Furthermore, color Doppler showed a posterior eccentric jet between the A1 and P1 segments with severe MR and a flow from the left atrium to the right atrium through the PFO tunnel. Normal sinus rhythm (NSR) was noted on the electrocardiography (ECG). Coronary angiography was performed for pre-operative evaluation and a substantial stenosis of the left anterior descending artery (LAD) was observed. Computed tomography (CT) of the thorax and abdomen showed that the patient had TSI, including the heart on the right side, the liver on the left side, and the spleen on the right side, also bilateral interstitial edema of the lungs, for which a diuretic therapy was administered. MVr, closure of PFO, and coronary artery bypass grafting were scheduled. After performing median sternotomy and opening the pericardium, dextrocardia was observed (Fig. 1). Following the preparation of the right internal mammary artery (IMA) and cannulation for cardiopulmonary bypass, the left atrium was explored, and chordal flail with partial anterior PMR was seen at the A1 segment. Subsequently, the PMR was resected (Fig. 2). The defect was primarily repaired with suture and one artificial neo-chord was implanted at the A1 segment following the water test. The ring annuloplasty was performed by the semirigid ring. After the closure of the PFO, the LAD-right IMA anastomosis was performed. On the intraoperative TOE, no MR was noted and the patient was transferred to the intensive care unit. On post-operative day 1, the patient was taken to the cardiovascular clinic. On the control echocardiography, he had no MR and an EF of 60%, and the mean mitral valve gradient was measured at 4 mmHg (Fig. 3). The patient had a NSR on ECG. Upon post-operative day 6, he was discharged home uneventfully on beta-blocker therapy. At a follow-up visit 22 months after the operation, the patient was doing well.

Discussion

TSI with dextrocardia is an infrequent anatomical characteristic, in which the heart lies in the middle of the mediastinum, with its base-to-apex axis directed to the right and caudally, with little known etiology.[7] Data are scarce on the coexistence of TSI and dextrocardia and MVr surgery for anterior PMR as well as CABG. The surgical approach to dextrocardia requiring a mitral valve together with CABG surgery and PFO closure is of prime importance.

Central cannulation is usually sufficient for surgical exposure while performing surgery for TSI. However, the inferior vena cava in the deeply located left atrium and abnormal rotation may be very hard to reach for cannulation. When performing MVR, cannulation of the femoral vein is more appropriate in complex anatomical situations,[6] because the decannulation process may cause a rupture of the right atrium and ventricule.[6]

The concept of situs is defined as the positioning of asymmetric organs, which has 3 anatomical types: the normal form of the placement of the organ is called situs solitus; a mirror image of the normal placement of the organs is called as TSI and situs ambiguous.[8]

All individuals with dextrocardia should have CT to detect abnormalities of vena cava veins. Persons in whom the right ventricle blocks access to the inferior vena cava may undergo the left atrial approach and a single dual-stage venous cannula.[9]

Coronary artery bypass grafting or mitral valve replacement with dextrocardia has appeared in recent literature.[6] Mitral valve surgery with dextrocardia in TSI may be associated with complicated surgical exposure owing to coexisting deviations of the caval connections and the localization of the atria, ventricles, and great vessels, so some surgeons preferred a left thoracotomy with peripheral cannulation to resolve this challenging issue.[6]

We performed a left classic atriotomy incision and reached the mitral valve easily; however, several publications reported that the performance of a classic left atriotomy incision to reach the mitral valve is likely to be impossible in patients with dextrocardia and TSI.

The mitral valve maintains its function by means of two papillary muscles (anterolateral and posteromedial). The anterolateral papillary muscle has a dual blood supply; however, the posterolateral papillary muscle has a single blood supply (the posterior descending artery); thus, the posterolateral PMR is 3–12 more frequently encountered, while the anterolateral PMR seems to be rare.[10] In addition, a complete PMR in the anterior papillary

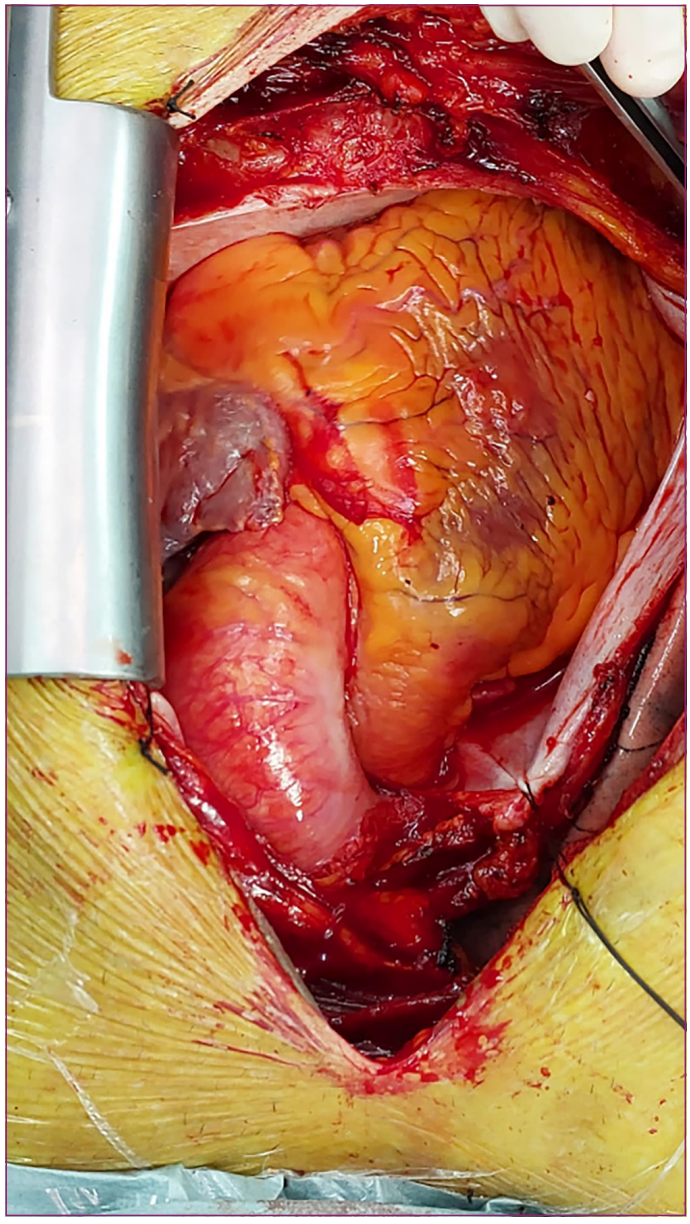

Figure 1. Surgical view of dextrocardia after median sternotomy.

Figure 2. Partial papillary muscle rupture.

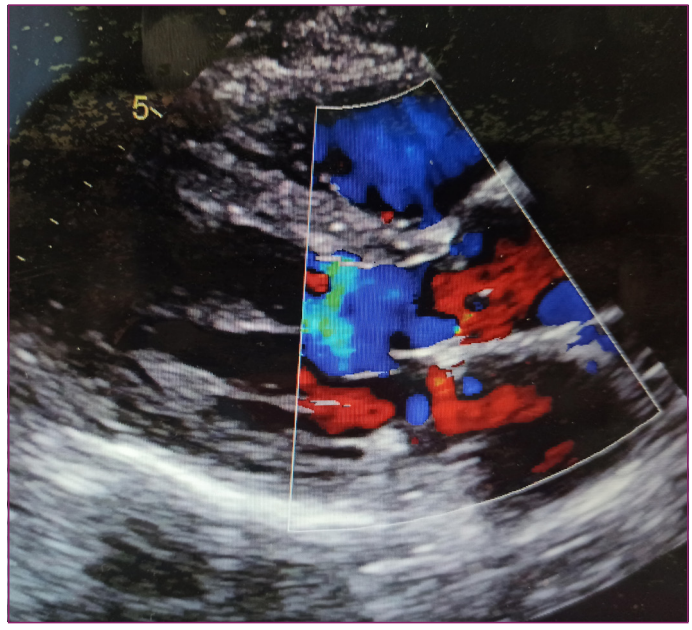

Figure 3. Post-operative echocardiography.

muscle is unfrequently.[6] Bouma et al.[5] reported that nine patients with PMR had undergone MVr, with a repair rate of 90% and a 5-year survival rate of 83%.

Conclusion

Patients who have TSI with dextrocardia and partial anterior PMR can undergo successful MVr, but when LAD revascularization is required, the use of RIMA is more suitable than that of LIMA during CABG. We performed cannulations and exploration of the left atrium as usual. The attending surgeon performed the surgery on the left side of the patient.

When performing surgery for severe MR with a flail segment concomitant with coronary artery stenosis, surgeons should explore mitral valve leaflets and sub-valvular apparatus in detail, because PMR is likely to be present, which should be kept in mind. In addition, the MVr techniques can be associated with successful outcomes in patients with PMR.

Disclosures

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Authorship Contributions: Concept – R.A.; Design – R.A.; Supervision – R.A.; Funding – R.A., M.A.O.; Materials – R.A.; Data collection and/or processing – C.A., M.A.O.; Data analysis and/or interpretation – R.A., M.B.R.; Literature search – R.A.; Writing – R.A.; Critical review – R.A., M.B.R.

Conflict of Interest: All authors declared no conflict of interest.

Use of AI for Writing Assistance: Not declared.

Financial Disclosure: The authors declared that this study received no financial support.

Peer-review: Externally peer-reviewed.

References

1. Burton LV, Beier K. Papillary muscle rupture. In: StatPearls. Treasure Island, FL: StatPearls Publishing; 2022.
2. Schroeter T, Lehmann S, Misfeld M, Borger M, Subramanian S, Mohr FW, et al. Clinical outcome after mitral valve surgery due to ischemic papillary muscle rupture. Ann Thorac Surg 2013;95(3):820–4. DOI: http://dx.doi.org/10.1016/j.athoracsur.2012.10.050
3. Kilic A, Sultan I, Chu D, Wang Y, Gleason TG. Mitral valve surgery for papillary muscle rupture: Outcomes in 1342 patients from The Society of Thoracic Surgeons database. Ann Thorac Surg 2020;110(6):1975–81.DOI: http://dx.doi.org/10.1016/j.athoracsur.2020.03.097
4. David TE. Papillary muscle-annular continuity: Is it important? J Card Surg 1994;9(2 Suppl):252–4. DOI: http://dx.doi.org/10.1111/j.1540-8191.1994.tb00937.x
5. Bouma W, Wijdh-den Hamer IJ, Klinkenberg TJ, Kuijpers M, Bijleveld A, van der Horst IC, et al. Mitral valve repair for post-myocardial infarction papillary muscle rupture. Eur J Cardiothorac Surg 2013;44:1063–9. DOI: https://doi.org/10.1093/ejcts/ezt150
6. St Rammos K, Bakas AJ, Panagopoulos FG. Mitral valve replacement in a Jehovah's Witness with dextrocardia and situs solitus. J Heart Valve Dis 1996;5:673–4.
7. Garg N, Agarwal BL, Modi N, Radhakrishnan S, Sinha N. Dextrocardia: An analysis of cardiac structures in 125 patients. Int J Cardiol 2003;88(2–3):143–55; discussion 155–6. DOI: http://dx.doi.org/10.1016/s0167-5273(02)00539-9
8. Lowenthal A, Tacy T, Punn R. Heterotaxy (Isomerism of the Atrial Appendages): Anatomy, Clinical Features, and Diagnosis. Waltham, MA: UpToDate Inc.; 2019. https://www.uptodate.com/contents/heterotaxy-isomerism-of-the-atrial-appendages-anatomy-clinical-features-and-diagnosis. Accessed Dec 08, 2019.
9. Koga S, Katayama Y, Koga K, Uchino M. Mitral valve and tricuspid valve regurgitation in a patient with dextrocardia and absence of the inferior Vena Cava. Kyobu Geka 2019;72(8):587–90.
10. Yamanishi H, Izumoto H, Kitahara H, Kamata J, Tasai K, Kawazoe K. Clinical experiences of surgical repair for mitral regurgitation secondary to papillary muscle rupture complicating acute myocardial infarction. Ann Thorac Cardiovasc Surg 1998;4(2):83–6.