Images in Cardiovascular Disease

Never Too Old for a Congenital Heart Disease: A Case of a Unileaflet Mitral Valve in a 76-year-old Man

Ahmed Salem , MBBS¹ Georgia Zachou , MBBS¹ Kanchani Makuloluwa , MBBS¹, and Attila Kardos , PhD^{1,2}

¹Department of Cardiology, Milton Keynes University Hospital, Milton Keynes, UK

²School of Sciences and Medicine, University of Buckingham, Buckingham, UK

OPEN ACCESS

Received: Aug 12, 2019

Accepted: Aug 27, 2019

Address for Correspondence:

Ahmed Salem, MBBS

Department of Cardiology, Milton Keynes
University Hospital NHS Foundation Trust,
Standing Way Milton Keynes MK6 5LD, UK.
E-mail: dr.ahmedsalem11@hotmail.com

Attila Kardos, PhD

Department of Cardiology, Milton Keynes
University Hospital NHS Foundation Trust,
Standing Way Milton Keynes MK6 5LD, UK.
E-mail: attila.kardos@cardiov.ox.ac.uk

Copyright © 2020 Korean Society of

Echocardiography

This is an Open Access article distributed
under the terms of the Creative Commons
Attribution Non-Commercial License (<https://creativecommons.org/licenses/by-nc/4.0/>)
which permits unrestricted non-commercial
use, distribution, and reproduction in any
medium, provided the original work is properly
cited.

ORCID iDs

Ahmed Salem

<https://orcid.org/0000-0003-2207-170X>

Georgia Zachou

<https://orcid.org/0000-0001-5792-4747>

Kanchani Makuloluwa

<https://orcid.org/0000-0002-5402-3502>

Attila Kardos

<https://orcid.org/0000-0002-0231-7605>

Conflict of Interest

The authors have no financial conflicts of
interest.

Congenital malformation of the mitral valve is considered to be a rare and complex entity.^{1,2)}

Singular leaflet mitral valve is the least prevalent form and has been mostly observed in infants, associated with symptomatic mitral regurgitation (MR) and tending to be incompatible with life beyond the neonatal period.^{1,2)} We report herein an unusual cause of unileaflet mitral valve in a 76-year-old man who presented with 5-day history of dyspnea. His cardiac background comprises repair of atrial septal defect (ASD) and percutaneous coronary intervention. He had clinical signs of fluid overload and a pan-systolic murmur in keeping with MR.

Transthoracic echocardiography revealed moderate to severe MR due to coaptation deficit of unidentified etiology (**Figure 1**). Two and three-dimensional transesophageal echocardiography demonstrated elongated anterior mitral valve leaflet and hypoplastic posterior leaflet with shortened chordae (**Figure 2, 3; Movie 1, Movie 2, Movie 3**) causing prominent MR (**Figure 4; Movie 4, Movie 5**). The left and right ventricles were dilated with systolic impairment with elevated pulmonary systolic pressure.

Figure 1. Transthoracic echocardiogram in para-sternal long axis showing coaptation deficit of the mitral valve. LA: left atrium, LV: left ventricle.

Figure 2. Transesophageal echocardiography mid-esophageal view, 0°, showing the mitral valve with a rudimentary posterior mitral leaflet adherent to the LV posterior wall (blue arrowhead) and elongated anterior mitral leaflet (red arrowhead). LA: left atrium, LV: left ventricle.

Figure 3. Three-dimensional short-axis view of the mitral valve showing hypoplastic posterior mitral valve leaflet (red arrowhead) and elongated myxomatous anterior mitral valve leaflet (AMVL).

The patient was stabilised on heart failure therapy and considered for mitral valve replacement.

Unileaflet mitral valves among adults are extremely rare and described mainly in case reports. In our case we propose that the congenital agenesis of posterior mitral leaflet and its functional importance has become apparent as a result of structural changes of the mitral annulus due to left ventricular dilatation secondary to ischemic heart disease.

Although, different forms of ASD could have genetic basis and are associated with anomalous pulmonary venous drainage or mitral valve cleft³; the link between ASD and unileaflet mitral valve has not been reported thus far.

Figure 4. Two-dimensional transesophageal echocardiography mid esophageal view showing showing the unileaflet mitral leaflet and color Doppler image of the mitral regurgitation.

SUPPLEMENTARY MATERIALS

Movie 1

2D TOE - mid-oesophageal 4 chambers view of the unileaflet mitral valve

[Click here to view](#)

Movie 2

2D TOE - X-plane mid-oesophageal view of the unileaflet mitral valve

[Click here to view](#)

Movie 3

3D rendering TOE image of the mitral valve showing the posterior leaflet agenesis

[Click here to view](#)

Movie 4

2D TOE mid oesophageal view showing the unileaflet mitral leaflet and colour Doppler image of the mitral regurgitation

[Click here to view](#)

Movie 5

2D TOE - X-plane mid-oesophageal view of the unileaflet mitral valve and mitral regurgitation

[Click here to view](#)

REFERENCES

1. Shah J, Jain T, Shah S, Mawri S, Anathasubramaniam k. Rare case of Unileaflet mitral valve. *J Cardiovasc Ultrasound* 2016;24:168-9.
[PUBMED](#) | [CROSSREF](#)
2. Bezgin T, Elveran A, Karagöz A, Çanga Y, Yılmaz F. Mitral valve with a single leaflet. *Türk Kardiyol Dern Ars* 2014;42:80-2.
[PUBMED](#) | [CROSSREF](#)
3. Pourafkari L, Baghbani-Oskouei A, Toufan M, Ghaffari S, Nader ND. Hypoplastic posterior mitral valve leaflet: A case report and review of the literature. *Echocardiography* 2018;35:1052-5.
[PUBMED](#) | [CROSSREF](#)