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CASE REPORT
Congenital heart disease

MitraClip repair of right-sided atrioventricular valve in a patient with congenitally corrected transposition of the great arteries: a case report

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Background Patients with congenitally corrected transposition of the great arteries (ccTGA) often develop tricuspid valve (systemic atrioventricular valve) dysfunction due to right ventricular overload and dilatation, but isolated mitral valve (MV) disease is rarely found. Isolated mitral (subpulmonic atrioventricular valve) interventions, specifically catheter-directed, have not been reported up to date.

Case summary A man with ccTGA is evaluated for dyspnoea. Multimodality imaging assessment confirmed severe right-sided MV regurgitation due to prolapse. In light of high surgical risk, a minimally invasive transcatheter MitraClip procedure was pursued.

Discussion To our knowledge, this is the first case of successful MV repair via percutaneous approach using MitraClip in a patient with ccTGA and biventricular failure. Our case illustrates the safety and feasibility of the edge-to-edge procedure in such a rare instance, but also the importance of multimodality imaging (both invasive and non-invasive) and the Heart Team approach when caring for these complex patients.

Keywords Transposition of the great arteries • Mitral valve • Congenital heart defect • Valve repair • Case report

ESC Curriculum 1 Imaging modalities • 4.3 Mitral regurgitation

Learning points

- To show the rarity of isolated subpulmonic atrioventricular valve [morphologic mitral valve (MV)] disease in patients with congenitally corrected transposition of the great arteries.
- To describe the novel use of the transcatheter MitraClip system for right-sided MV prolapse as well as demonstrating its safety and efficacy.
- To highlight the importance of multimodality imaging, specifically intracardiac and transoesophageal echo in the percutaneous approach of right-sided MV prolapse repair.

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Introduction

Congenitally corrected transposition of the great arteries (ccTGA) is a rare complex congenital heart disease characterized by atrioventricular and ventriculo-arterial discordance. The systemic atrioventricular valve [morphologic tricuspid valve (TV)] is prone to becoming progressively incompetent during the 2nd to 5th decades of life due to dilatation and dysfunction of the morphologically right systemic ventricle,[1] but isolated cases of subpulmonic atrioventricular valve [morphologic mitral valve (MV)] dysfunction are rare and therefore surgical interventions for MV alone are not performed unless in combination with TV repair/replacements. We present the first documented case of a percutaneous MV repair using MitraClip® in a patient with ccTGA.

Timeline

Day 1	A 60-year-old man with a medical history of congenitally corrected transposition of the great arteries is referred for dyspnoea on exertion.
Day 3	Patient sent for transoesophageal echocardiography (TOE) assessment which showed the presence of right-sided mitral valve (MV) prolapse with severe regurgitation.
Day 4	Patient presented at Heart Team discussion due to high surgical risk, decision was made to proceed with transcatheter MV repair using MitraClip.
Day 10	Procedure day. Patient electively admitted for placement of MitraClip. Operation took place in a hybrid cath lab suite under TOE and intracardiac echocardiography guidance. • Sedation initiated, patient intubated by Cardiovascular Anaesthesiologist. TOE probe inserted. Femoral access obtained. Intracardiac echocardiography and MitraClip catheters inserted. • First clip deployed without complications. • Second clip placement was associated with single leaflet detachment. • Attempt at inserting third clip was abandoned due to difficulty in visualization and manoeuvring. • Final haemodynamic assessment revealed 53% reduction in right atrial pressures.
Day 40	Post-operative assessment: patient with significant symptomatic improvement.
Day 100	Transthoracic echocardiography at 3 months revealed overall 2+ mitral regurgitation reduction in severity with stable clip positioning

Case presentation

A 60-year-old man with a history of atrial fibrillation and ccTGA presented to the cardiology clinic for exertional dyspnoea. The patient complained of 6 months of deterioration of his functional class from New York Heart Association (NYHA) I to NYHA III. Other associated symptoms included orthopnoea and mild leg swelling. The patient medication regimen included lisinopril 5 mg daily, carvedilol 3.125 mg twice daily, apixaban 5 mg twice daily, and furosemide 40 mg daily. Vital signs were within normal limits. Physical exam demonstrated laterally displaced point of maximal impulse. The heart rhythm was regular with a holosystolic murmur at the base. Fine bibasilar crackles were heard and +1 bilateral lower extremity oedema was found. Blood work revealed an N-terminal pro B-type natriuretic peptide (NT pro-BNP) of 1450 pg/mL (reference range 0–125 pg/mL) with a negative troponin. An electrocardiogram demonstrated sinus rhythm, non-specific intraventricular conduction delay with non-specific T-wave changes, and premature ventricular contractions. Transthoracic echocardiography showed a moderately reduced ejection fraction (EF) of 35% involving the systemic morphologic right ventricle (RV) with mild left-sided TV regurgitation. The right-sided morphologic left ventricle (LV) exhibited both systolic dysfunction and enlargement with severe mitral regurgitation (MR). Transoesophageal echocardiography (TOE) showed the presence of right-sided MV prolapse involving the anterior leaflet with severe regurgitation (*Figure 1*). Quantitative assessment with cardiac magnetic resonance demonstrated the following measurements: subaortic ventricle: end-diastolic volume (EDV) 260 mL, end-systolic volume (ESV) 175 mL, EF 32%; subaortic TV: regurgitant volume 15 mL/beat and regurgitant fraction of 18%; subpulmonic ventricle: EDV 225 mL, ESV 89 mL, EF 59%; subpulmonic MV: regurgitant volume 48.4 mL/beat, and regurgitant fraction of 37% (*Figure 2*). There was no evidence of late gadolinium enhancement.

Isolated surgical MV repair in the setting of ccTGA with a dysfunctional systemic RV and right-sided LV, was felt to be high risk [Society of Thoracic Surgeons (STS) score 9%], with failed repair presenting difficult choices regarding the optimal valve replacement strategy, so Heart Team decision was made to proceed with transcatheter edge-to-edge repair of the right-sided morphologic MV using a MitraClip system (Abbott Vascular, IL, USA).

Intervention

The procedure was performed in a hybrid operating suite under general anaesthesia and 3D TOE and intracardiac echocardiography (ICE) imaging. Intracardiac echocardiography re-demonstrated severe prolapse of MV (*Video 1*). The 24-Fr MitraClip steerable guide catheter was advanced into the confluence of the inferior vena cava and right atrium (RA) through the right femoral vein adding negative knob to straighten the distal shaft. Next, the MitraClip XRT and catheter delivery system (CDS) were introduced through the steerable guide catheter using a non-conventional 'mis-keyed' technique 180° opposite to the blue tracking line. This was done based on current practice for TriClip™ placement, in which due to the more anterior anatomic location of the TV (and in this case morphologic right-sided MV), the CDS is intentionally mis-keyed to adjust for orientation difference. This manoeuvre optimizes the position of the device relative to the valvular annulus, allowing a clockwise rotation of the guide and enabling the 'A-knob' to bend the CDS into a direction perpendicular to the TV plane.[2]

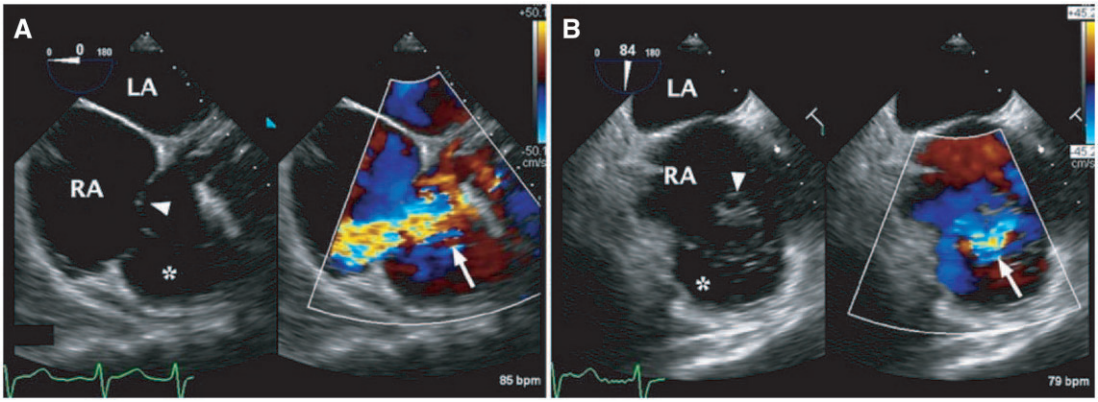


Figure 1 Transoesophageal echocardiography examination at mid-oesophageal (*A*) and trans-gastric (*B*) showing right-sided mitral valve prolapse of the anterior leaflet (arrowhead) with severe eccentric regurgitation jet (arrow). LA, left atrium; RA, right atrium; *morphologic left ventricle.

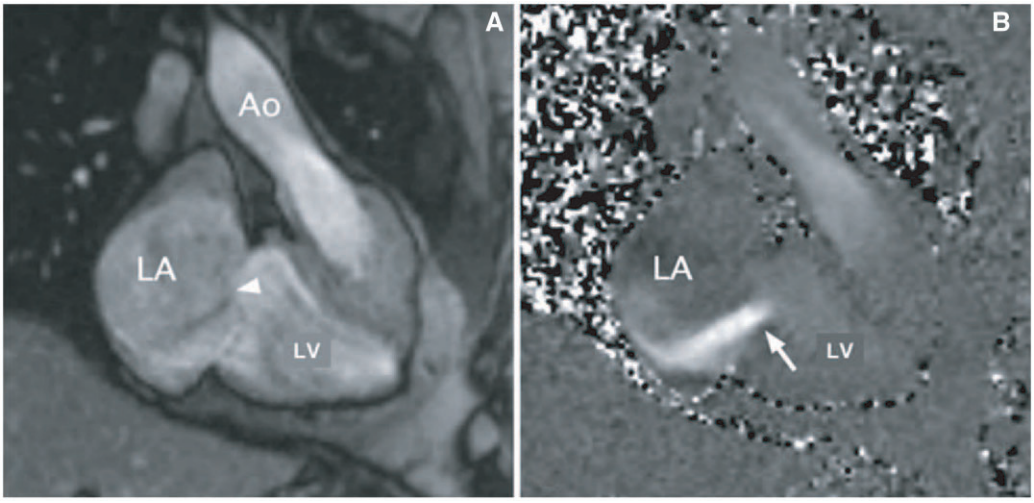


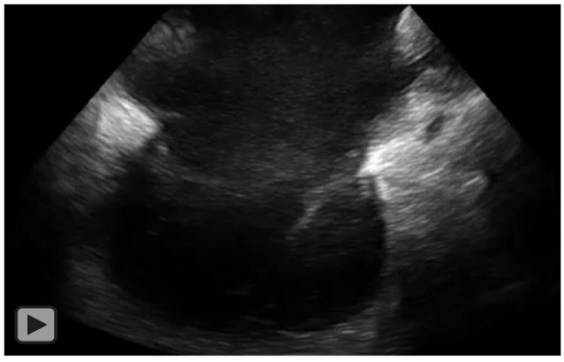
Figure 2 Cardiac magnetic resonance in long-axis cine gradient recalled echo (*A*) and phase-contrast (*B*) during mid-systole showing right-sided mitral valve prolapse of the anterior leaflet (arrowhead) with severe eccentric regurgitation jet (arrow). Ao, ascending aorta; LA, left atrium; LV, morphologic left ventricle.

The MitraClip was advanced carefully beyond the steerable guide catheter tip within the RA and into straddling position under fluoroscopic and TOE guidance (*Figure 3A*). The 'A' knob was used to orient the MitraClip towards the morphologic MV. Transoesophageal echocardiography examination using multi-plane 2D and 3D imaging allowed for clip arm orientation and alignment (*Figure 3B*). The clip was subsequently advanced into the sub-valvular space in closed position and opened to grasp position at 160° (*Video 2*).

Transoesophageal echocardiography did not allow for adequate leaflet visualization during grasp. A 9-Fr Viewmate ICE catheter (Abbott Vascular, IL, USA) was required and introduced through a 10-Fr sheath into the RA from the left femoral vein (*Figure 3A*). Intracardiac echocardiography provided the image clarity necessary to safely grasp and confirm leaflet insertion (*Figure 4A–C*, *Video 3*). The clip arm was locked and closed fully to create a tissue bridge and double orifice MV (Supplementary material online, *Video S1*). The clip was successfully released following a noticeable 2+ reduction in regurgitation and mean residual transvalvular gradient of 1 mmHg (Supplementary material online, *Video S2*). Attempts were made to deliver a second XTR clip with the expectation of further minimizing the degree of residual MR (Supplementary material online, *Video S3*). Visual interaction of the first clip made placement of the second clip challenging and resulted in posterior single leaflet detachment despite the use of both TOE and ICE (*Figure 4D–F*, Supplementary material online, *Videos S4–S6*). Attempts to deploy a third clip were abandoned for the same reasons. Final

haemodynamic assessment revealed a 45% reduction in mean right atrial pressure from 15 to 8 mmHg with significant improvement in peak V waves (*Figure 5*).

The patient was extubated in the procedure room and discharged home the next day. At 1-month follow-up, the patient reported remarkable clinical improvement of functional class from NYHA III back to NYHA I. Transthoracic echocardiography at 3 months revealed overall 2+ reduction in MR severity, with stable A2/P2 clip in juxtaposition to the previously detached second clip (Supplementary material online, *Video S7*) along with sustained symptomatic benefit. Physical exam at the time demonstrated improved signs of hypervolaemia and NT pro-BNP levels decreased to 997 pg/mL.

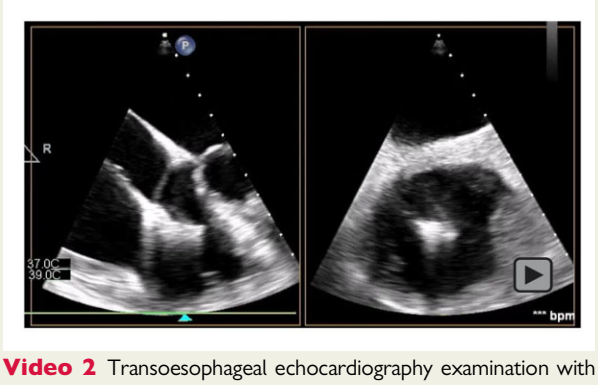


Video 1 Intracardiac echocardiography showing severe prolapse of anterior mitral valve leaflet with wide regurgitant gap and thickened anterior leaflet tips.

Discussion

To our knowledge, this report is the first describing percutaneous valve repair of right-sided MR in a patient with ccTGA. The case demonstrates the safety and feasibility of the MitraClip procedure for patients with ccTGA that are considered high risk for surgical repair. It also illustrates the limitations of TOE imaging during intraoperative assessment of right-sided heart structures, and the value of ICE imaging in selected cases.

Although ccTGA has been associated with Ebstein's anomaly of the left atrioventricular valve (TV) and other forms of congenital heart defects,[3,4] we found no association between ccTGA and MV prolapse. Neither did we find any cases in literature of isolated right-sided MV repair in patients with ccTGA. Mitral valve surgery, when necessary, has been reported in combination with left-sided TV



Video 2 Transoesophageal echocardiography examination with bi-plane imaging during placement of first clip, with visually detectable double orifice mitral valve (short-axis transoesophageal echocardiography, right panel).

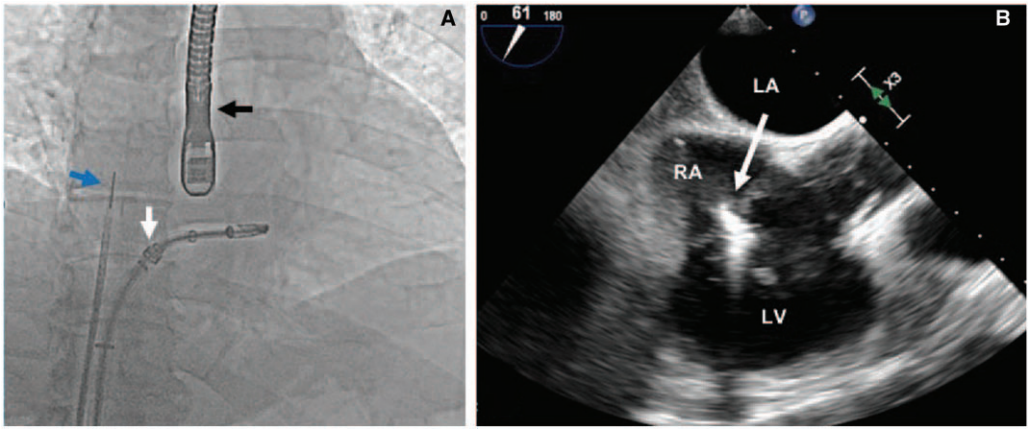


Figure 3 (*A*) Fluoroscopic image demonstrating MitraClip in straddling position (white arrow), directed towards the right-sided mitral valve, along with transoesophageal echocardiography probe (black arrow) and intracardiac echocardiography catheter (blue arrow) at the level of the inferior vena cava to right atrial junction. (*B*) Mid-oesophageal transoesophageal echocardiography examination showing MitraClip arms (white arrow) perpendicular to the mitral valve plane in a patient with congenitally corrected transposition of the great arteries. LA, left atrium; LV, morphologic left ventricle; RA, right atrium.

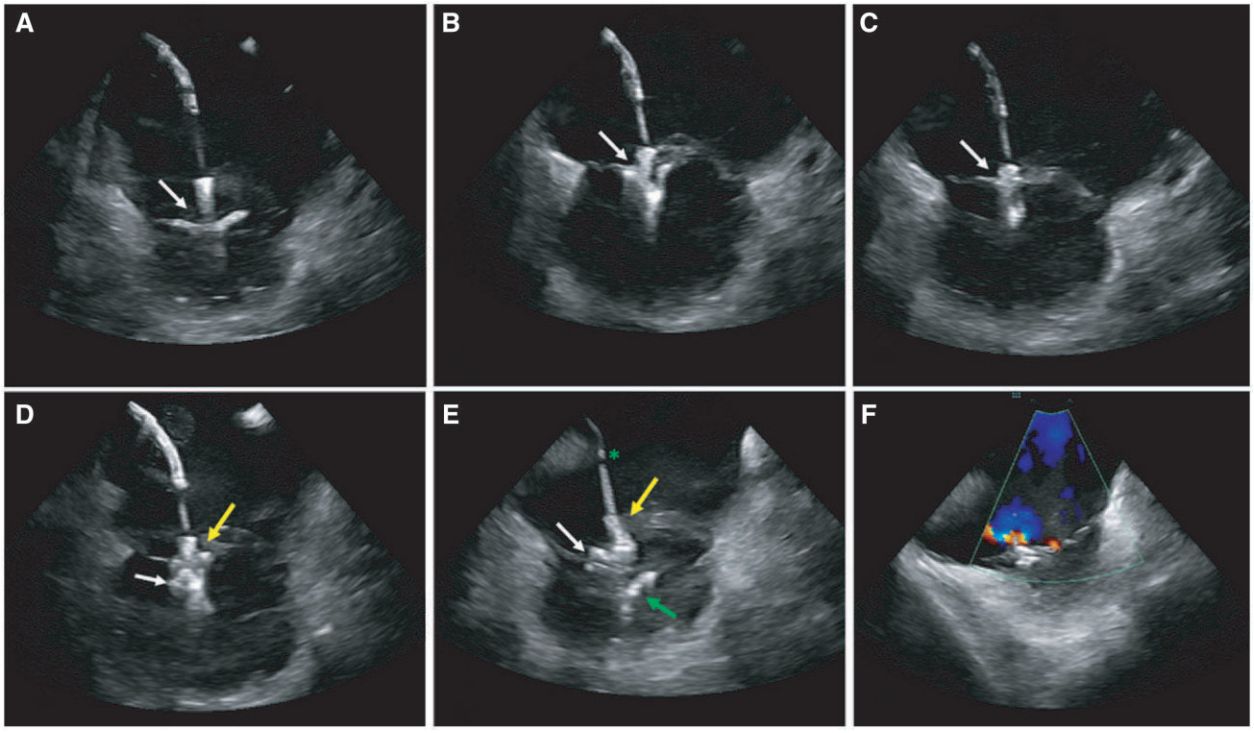
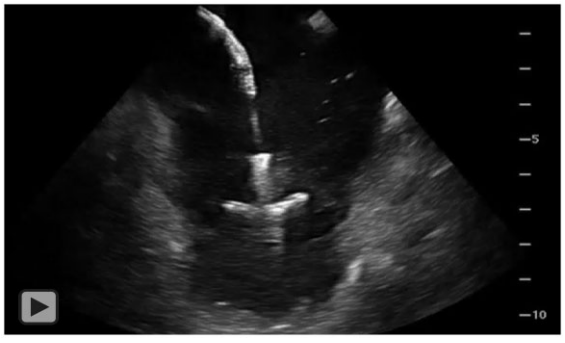


Figure 4 Intracardiac echocardiography showing right-sided mitral valve with MitraClip arms (white arrow) in open position (*A*), followed by successful leaflet insertion (*B*) and final grasp upon complete closure (*C*). (*D* and *E*) Challenging deployment of second clip with subsequent detachment (yellow arrow) and failed deployment of third (green arrow) due to visual interference from first clip are shown. Green asterisk = MitraClip delivery system. (*F*) Final mitral regurgitation at the end of the procedure is shown.



Video 3 Intracardiac echocardiography showing clip arms and mitral valve leaflets during grasp.

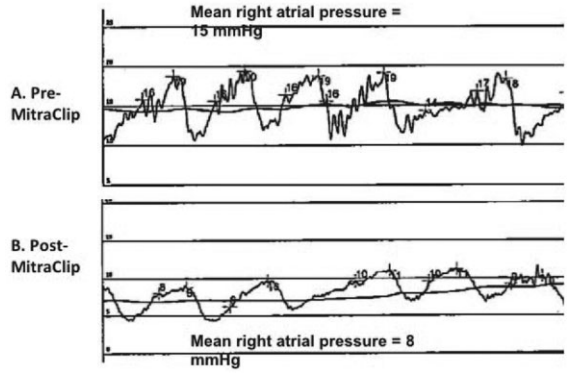


Figure 5 Haemodynamic tracing pre- and post-MitraClip deployment showing 45% reduction in mean right atrial pressures and prominent V waves.

surgery to preserve right ventricular function; the most important determinant of morbidity and mortality in patients with ccTGA.[3] However, MR is usually functional in this setting and not degenerative as in our case.[3,4]

While our case represents the first description of MitraClip in a right-sided MV, the edge-to-edge technique has been examined as a therapeutic option in the more commonly encountered left-sided tricuspid regurgitation of patients with ccTGA.[1,5,6] In contrast with the morphologic MV, the aim of MitraClip in anatomical TVs is to clip two of the three leaflets (more commonly the septal leaflet) to create a double orifice TV.[7]

In our patient with ccTGA, isolated A2 prolapse of the morphological anterior MV leaflet resulted in severe regurgitation and heart failure. While the option of an isolated surgical MV repair was

considered, the minimally invasive transcatheter approach was preferred due to concerns of concomitant biventricular dysfunction. Following the technical principles of a right-sided TriClip procedure,[7] successful A2/P2 grasp of the morphologic MV resulted in a double orifice valve with significant reduction in MR severity and sustained clinical improvement at 7 months, demonstrating safety and feasibility.

In our experience, the combined use of TOE and ICE may be complimentary. Transoesophageal echocardiography allowed for anatomic orientation of the MitraClip, while ICE provided the image resolution necessary to assess leaflet insertion during grasp. Our observations are similar to those of Pozzoli *et al.*,[6] describing the benefits of ICE during right-sided TriClip interventions. While ICE imaging is limited by its single plane acquisition, its proximity to near-field objects in right-sided procedures provides the added benefits of superior visualization. With the advent of the newer generation 3D ICE catheters it is likely that we will further appreciate its value during such interventions.

Conclusion

To our knowledge, this is the first case of successful MV repair via percutaneous approach using MitraClip in a patient with ccTGA and biventricular failure. Our case illustrates the safety and feasibility of the edge-to-edge procedure in such a rare instance, but also the importance of multimodality imaging (both invasive and non-invasive) and the Heart Team approach when caring for these complex patients.

Lead author biography

Dr Antonio Lewis is an International Medical Graduate from Colombia currently undergoing Cardiology training at Cleveland Clinic Florida (Weston, FL) as a PGY-4. He has a long-standing passion for Cardiology and his main interests are Preventive Cardiology, Heart Failure & Heart Transplant and Electrophysiology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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