Isolated unilateral pulmonary artery atresia with contralateral pulmonary artery branch stenosis: A "window" for intervention

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ABSTRACT

Adult presentation of unilateral pulmonary artery atresia in association with contralateral branch pulmonary stenosis is rare. We present the case of a quadragenarian, who manifested with right ventricular failure and hemoptysis. This report discusses the diagnostic workup and therapeutic options along with a brief overview of the concerned literature.

Keywords: Adult-onset congenital heart disease, multimodality imaging, peripheral pulmonary stenosis, pulmonary angioplasty, unilateral pulmonary artery atresia

INTRODUCTION

Isolated unilateral pulmonary artery atresia (UPAA) results from the failure of embryonic sixth aortic arch to fuse with the pulmonary trunk during embryonic development.[1] Significant narrowing of the pulmonary artery (PA) may lead to an overall reduction in pulmonary blood flow with disproportionate distribution of right ventricular (RV) cardiac output to the lungs. In addition, an increase in RV pressure portends right ventricular hypertrophy (RVH) and possible failure. We, herein, present a rare case of isolated UPAA and contralateral branch PA stenosis presenting in adulthood, with subsequent diagnostic evaluation and management strategies.

CASE SUMMARY

A 37-year-old man with uneventful childhood presented with recent-onset, progressively increasing exertional fatigue and pedal edema for the past 1 month. Physical examination revealed elevated jugular venous pressure with prominent "a" and "v" waves, grade 3/3 left parasternal heave, RV S3, mid-systolic murmur of grade 2/6 in the left 3rd intercostal space in the parasternal area, and a soft continuous murmur in the right lung fields. ECG showed right axis deviation, clockwise loop, and RVH with strain pattern with the absence of early transition. The chest X-ray showed cardiomegaly, RV and right atrial enlargement, prominent main pulmonary artery (MPA) with differential lung vascularity (right > left), and mild right-sided pleural effusion. Transthoracic echocardiogram revealed dilated right atrium and ventricle with RVH, severe tricuspid regurgitation (TR) with elevated RV pressures, dilated MPA, left aortic arch, and normal pulmonary valve morphology with moderate pulmonary regurgitation without any shunt lesion. The left pulmonary artery (LPA) could not be visualized, and the right pulmonary artery (RPA) had a proximal severe discrete stenosis (peak gradient = 72 mmHg) [Figure 1]. Computed tomography pulmonary angiography confirmed discrete membranous stenosis in the proximal

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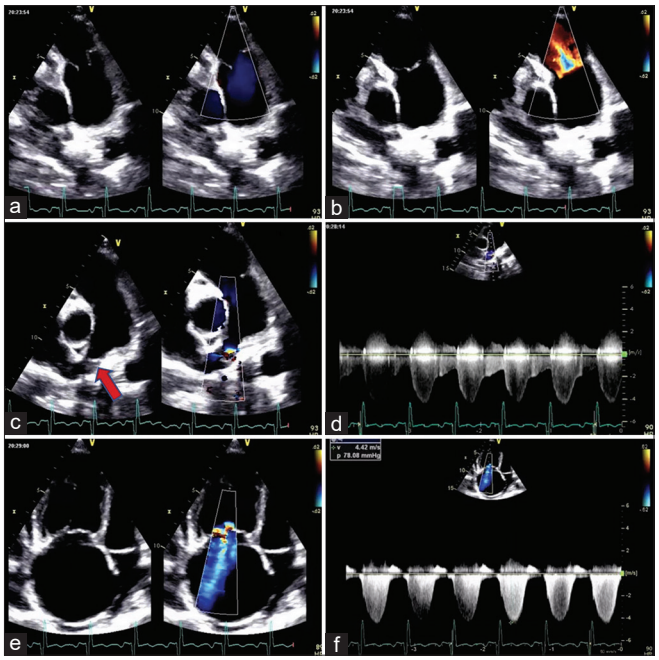

Figure 1: Preoperative two-dimensional echocardiographic assessment. (a) Dilated MPA with laminar flow across nonthickened pulmonary valve. (b) Moderate pulmonary regurgitation (PAEDP = 15 mmHg). (c) Turbulent Doppler flow in the proximal RPA caused by an obstructive thick membrane with continuous wave Doppler showing peak instantaneous velocity of 4 m/s (PG = 72 mmHg) (d). (e) Dilated RA and RV with severe TR (RVSP = 88 mmHg) (f). MPA: Main pulmonary artery, RPA: Right pulmonary artery, RA: Right atrium, RV: Right ventricle, PG: Peak gradient, PAEDP: Pulmonary artery end-diastolic pressure, RVSP: Right ventricular systolic pressure, TR: Tricuspid regurgitation

RPA with absent LPA [Figure 2]. RPA diameter at the site, proximal and distal to stenosis, measured 20.5 mm, 23 mm, and 24 mm, respectively. Collateral from the left subclavian artery (LSCA) provided vascular supply to the left lung.

Cardiac catheterization [Table 1] revealed elevated right atrial pressure, systemic RV pressure with raised end-diastolic pressures (RVEDP = 14 mmHg) suggestive of moderate RV dysfunction, and ventricularization of MPA pressure with a pullback gradient of 85 mmHg across RPA without any systemic desaturation [Figure 3]. PA angiogram revealed dilated MPA and discrete membranous stenosis in the proximal RPA with absent LPA [Videos 1-3]. Selective LSCA injection revealed a nonarborizing collateral supplying the left lung [Video 5]. Branch pulmonary angioplasty was planned to relieve obstruction to the sole outflow of RV. Pulmonary valve and proximal RPA stenosis was crossed with a 7F Swan-Ganz catheter over an 0.035" J-tip exchange-length hydrophilic-coated guidewire (Terumo Glidewire®, Terumo Medical Corp., NJ, USA) which was changed to 260-cm Amplatz® ultra-stiff guidewire (St. Jude Medical, Plymouth, MN, USA). The lesion was dilated with an 18 × 30 mm Tyshak II® (NuMed Inc., Hopkinton, NY, USA) balloon following

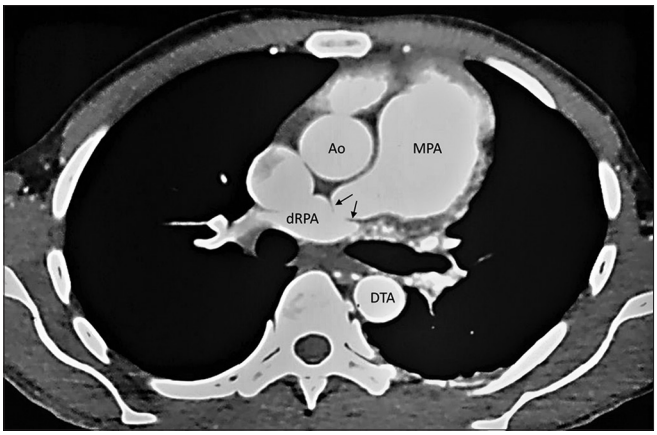

Figure 2: Preoperative axial cross-section of 128-slice computed tomography pulmonary angiogram showing dilated MPA with prominent membranous obstruction (black arrows) in the proximal RPA. LPA was conspicuously absent. MPA: Main pulmonary artery, RPA: Right pulmonary artery, LPA: Left pulmonary artery, CT: Computed tomography

which a 90-cm 12F Flexor® sheath (Cook Medical Inc., Bloomington, IN, USA) was advanced up to MPA [Video 4]. A balloon-expandable 8-zig, 39 mm cheatham-platinum stent (NuMed Inc., Hopkinton, NY, USA) premounted on a 22 × 40 mm balloon-in-balloon catheter (BIB®, NuMed Inc., Hopkinton, NY, USA) was subsequently deployed at the lesion site [Figure 4]. Poststent deployment, there was no angiographic contrast leak or residual gradient across the lesion [Videos 6 and 7]. However, around 15 min postprocedure, the patient went into frank pulmonary edema due to reactionary overflooding of the right lung post-RPA stenting that was managed conservatively with noninvasive ventilation, intravenous vasodilators, diuretics, and low-dose phosphodiesterase-5 inhibitors. At 1-year follow-up, the patient is asymptomatic with RV systolic pressure being reduced to 50 mmHg along with marked improvement in RV function.

DISCUSSION

UPAA is a rare entity to encounter in an adult patient in the absence of any underlying congenital cyanotic heart diseases such as tetralogy of Fallot (TOF), transposition of great arteries, or other conotruncal anomalies.[2-5] Our case demonstrated an extremely rare combination of UPAA and contralateral PA stenosis without any other associated congenital cardiac malformation, leading to RV pressure overload dysfunction. Isolated pulmonary artery of ductal origin is a close mimic of the above condition despite being an entirely different embryological entity in which the isolated agenesis of the branch PA is almost always opposite to the sidedness of the aortic arch which was not observed in our case.[6]

With essentially a single PA arising from the right ventricle, which too has a proximal severe stenosis, this pulmonary artery systolic (PAS) lesion was

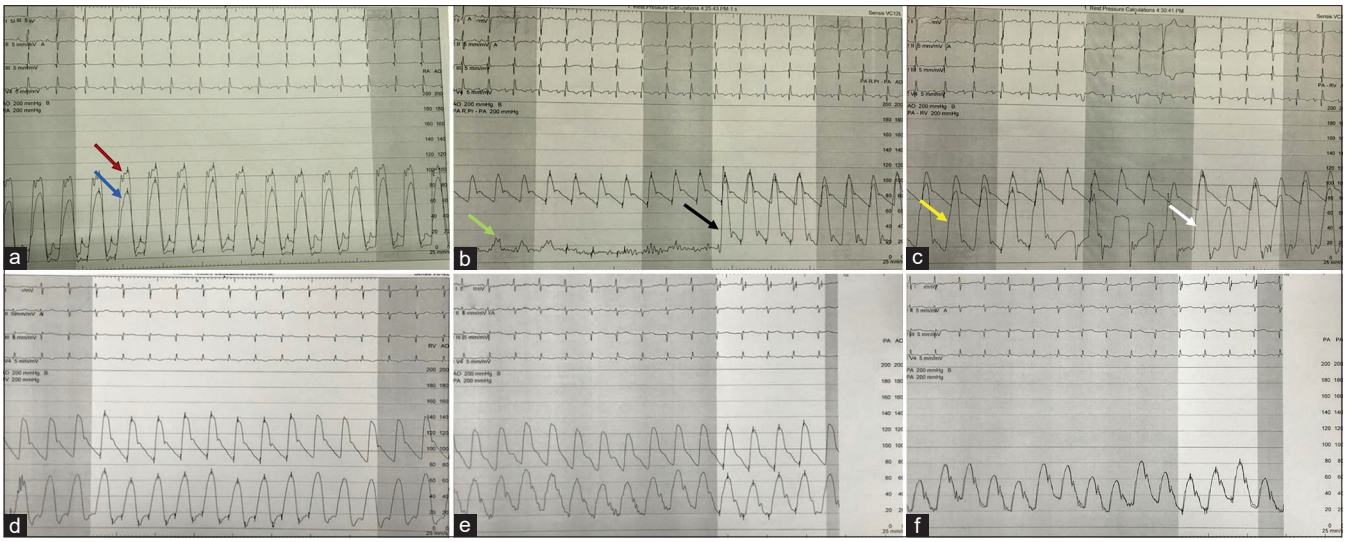

Figure 3: Pressure tracing during cardiac catheterization. (a) Simultaneous LV (red arrow) and RV pressures (blue arrow). (b) Pullback pressure tracing from distal RPA (green arrow) to MPA (black arrow) with a peak-to-peak gradient of 80 mmHg across RPA. (c) No pressure gradient during pullback tracing from MPA (yellow arrow) to RV (white arrow). (d) Reduction in RV pressure postintervention. (e) Pullback pressure tracing from distal RPA to MPA with abolition of gradient. (f) MPA pressure tracing. MPA: Main pulmonary artery, RPA: Right pulmonary artery, LV: Left ventricle, RV: Right ventricle

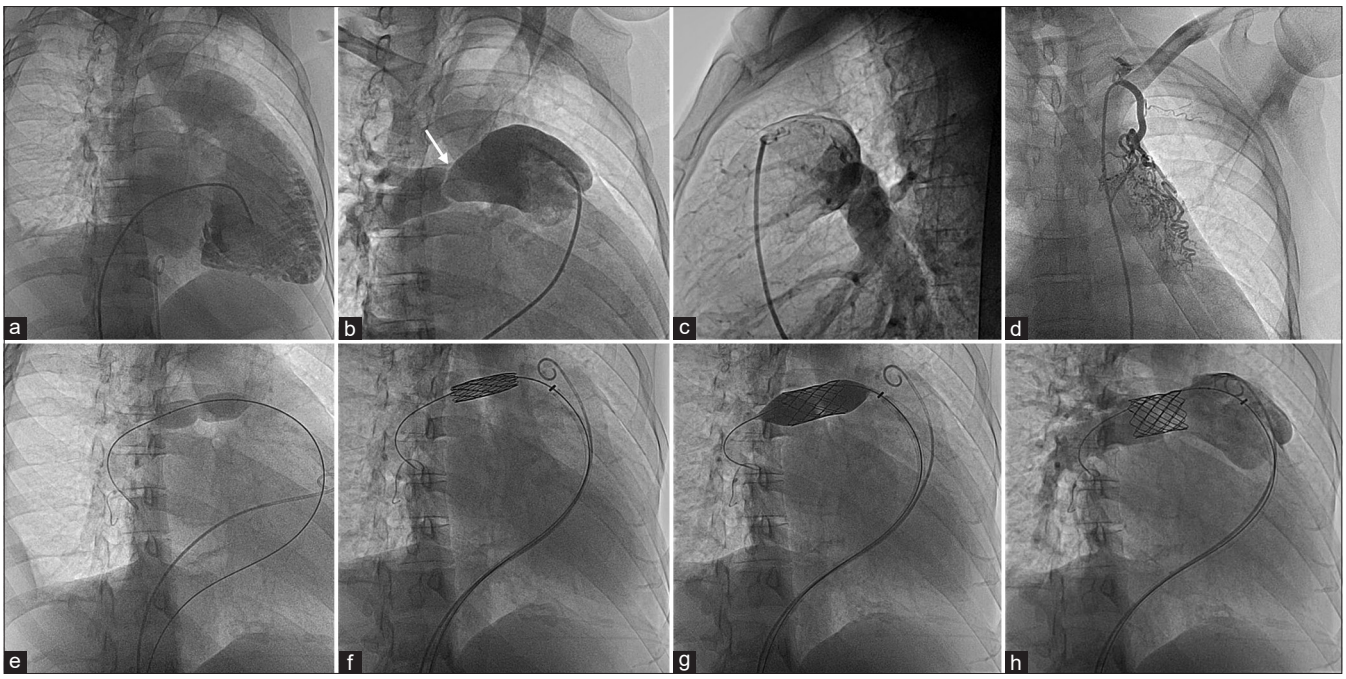

Figure 4: Basal right ventriculogram in RAO view done using 7F NIH catheter showing dilated RV, RA, and MPA with severe TR (a). MPA angiogram in RAO view showing grossly dilated MPA with a thick membranous obstruction (white arrow) in the proximal RPA (b). MPA angiogram in the left lateral view shows blunt cutoff MPA not giving origin to LPA (c). (d) An indirect nonarborizing MAPCA arising from the LSCA supplying the left lung. (e-h) Percutaneous balloon dilatation with stenting of proximal RPA stenosis. MPA: Main pulmonary artery, RPA: Right pulmonary artery, RA: Right atrium, RV: Right ventricle, LPA: Left pulmonary artery, MAPCA: Major aortopulmonary collateral, RAO: Right anterior oblique, TR: Tricuspid regurgitation, LSCA: Left subclavian artery

hemodynamically equivalent to a significant RV outflow tract obstruction with no other vent for RV to eject its stroke volume, which does not usually happen in unilateral PAS or UPAA where the contralateral PA accommodates and practically decompresses the right ventricle. Contralateral PAS also protected the pulmonary bed from vascular hypertension effects at the cost of RV dilatation, pressure overload, and dysfunction. Since the left lung vascular supply was contributed by a non-unifocalizable collateral from LSCA, it was decided to target the RPA stenosis through percutaneous balloon angioplasty after demonstrating the right lung being free

Table 1: Cardiac catheterization oximetric and pressure data

	Saturation (%)	Pressure (mmHg)
HSVC	50.7	-
LSVC	49.6	
IVC	53.6	-
HRA	61.8	
MRA	59.6	20, 22 (19)
LRA	63.6	
LA		
RV	61.6	100, 0, 14
MPA	58.5	100, 12, 51
Proximal RPA		100, 12, 51
Peripheral RPA	59.6	15, 8, 11
Wedge	94.5	4, 5, 3
LV	95.7	110, 0, 4
AO	96.0	112/82 (97)

HSVC: High superior vena cava, LSVC: Low superiuor vena cava, IVC: Inferior vena cava, HRA: High right atrium, MRA: Mid right atrium, LRA: Low right atrium, LA: Left atrium, RV: Right ventricle, MPA: Main pulmonary artery, RPA: Right pulmonary artery, LV: Left ventricle, AO: Aorta

of irreversible pulmonary hypertension. Post-stenting, an increase in distal RPA pressure was seen secondary to a sudden redistribution of blood flow to the single-lung circulation thereby leading to hyperperfusion lung injury in the previously flow-deprived right lung for which medical management generally suffices. Stepwise gradual dilatation of PA, initial expansion with a smaller stent, and a staged re-dilatation could be employed to prevent such complication.

Management strategies for UPAA associated with TOF include single-lung repair, double-lung repair, and palliative procedures (including systemic-to-pulmonary shunts and balloon dilatation).[7] Pneumonectomy and surgical revascularization including unifocalization with conduit repair are considered only after recurrent hemoptysis, pulmonary infections, and pulmonary hypertension develop.[8] We, in our case, decided not to consider any intervention beyond branch PA stenting and continue with medical management as pathophysiology in our case was predominantly governed by RV outflow tract obstruction secondary to RPA stenosis without any evidence of pulmonary vascular obstructive disease on radiological and catheterization study.

CONCLUSION

We report a rare presentation of UPAA in adults. Multimodality imaging is helpful in anatomical assessment before planning of appropriate interventions. Cardiac magnetic resonance can prove useful in understanding the nature and morphology of branch PA obstruction, functional assessment of flows, and pulmonary regurgitation on subsequent follow-up visits besides cutting down on multiple radiation exposures required in computed tomography angiography. Catheter-based interventions are a safe and effective option in relieving pressure overload on the right cardiac chambers, especially in the setting of underlying ventricular dysfunction, where surgical risks are invariably high.

Learning points

1. It was an extremely rare combination of UPAA and contralateral PAS in an adult patient without any other associated congenital cardiac malformation, like TOF, transposition of great arteries, or other conotruncal anomalies
2. Pathophysiology in our case was predominantly governed by RV outflow tract obstruction secondary to RPA stenosis, with UPAA being probably clinically silent without any evidence of pulmonary vascular obstructive disease on a radiological and catheterization study
3. The use of long-braided sheath and/or super-stiff buddy guidewire support is recommended for crossing peripheral PA stenosis and tracking stents reinforcing balloon dilatation
4. Poststenting, due to a sudden return of perfusion to the flow-deprived lung, acute pulmonary edema is a common complication for which medical management (including vasodilators and diuretics) is generally sufficient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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