ISOLATED UNILATERAL PULMONARY ARTERY ATRESIA WITH CONTRALATERAL PULMONARY ARTERY BRANCH STENOSIS - A 'WINDOW' FOR INTERVENTION

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History and physical:

37-year-old man with uneventful childhood presented with recent onset, progressively increasing exertional fatigue and pedal edema for past one month. Physical examination revealed elevated jugular venous pressures with prominent 'a' and 'v' waves, grade 3/3 left parasternal heave, RV S3, mid systolic murmur of grade 2/6 in left 3rd intercostal space parasternal and a soft continuous murmur in right lung fields.

Imaging:

Transthoracic echocardiogram revealed dilated right atrium and ventricle with RVH, severe tricuspid regurgitation with elevated RV pressures, dilated MPA, normal pulmonary valve morphology with moderate pulmonary regurgitation without any shunt lesion. Right pulmonary artery (RPA) had a proximal severe discrete stenosis (PG = 72 mm Hg). CT pulmonary angiography confirmed discrete membranous stenosis in proximal RPA with absent LPA (Figure 3). Non-arborizing collaterals from left subclavian artery supplied left lung.

Indication for intervention:

With essentially a single pulmonary artery arising from right ventricle which too, has a proximal severe stenosis, this pulmonary artery stenotic (PAS) lesion was hemodynamically equivalent to a significant RV outflow tract obstruction with no other vent for RV to eject its stroke volume which doesn't happens in a usual case of unilateral PAS or atresia where the contralateral PA accommodates and practically decompensates the right ventricle. Contralateral PAS protected the pulmonary bed from vascular hypertension effects at the cost of RV dilatation, pressure overload and dysfunction thereby forming the basis for intervention. Branch pulmonary angioplasty was planned to relieve obstruction to the sole outflow of RV.

Intervention:

Cardiac catheterization revealed elevated right atrial pressures, systemic RV pressures with mildly raised end diastolic pressures suggestive of RV dysfunction, ventricularization of MPA pressure with a pullback gradient of 85 mm Hg across RPA without any systemic desaturation. Right ventriculogram revealed a dilated chamber with mildly reduced contractility with severe TR, dilated MPA and RPA. PA angiogram revealed discrete membranous stenosis in proximal RPA with absent LPA. Lesion was dilated with 18x30 mm Tyshak II® (NuMed Inc, Hopkinton, NY, USA) balloon followed by deployment of balloon-expandable 8-zig, 39 mm Cheatham Platinum (CP) stent (NuMed Inc, Hopkinton, NY, USA) pre-mounted on a 22x40 mm balloon-in-balloon catheter (BIB®, NuMed Inc, Hopkinton, NY, USA).

Learning points of the procedure:

1. It was an extremely rare combination of unilateral pulmonary artery atresia (UPAA) and contralateral PAS in an adult patient without any other associated congenital cardiac malformation, like tetralogy of Fallot (TOF), transposition of great arteries or other cono-truncal 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 radiological and catheterization study.
3. 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. Post stenting, due to 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.