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RARE CAUSE OF PULMONARY HYPERTENSION : UNILATERAL ABSENCE OF PULMONARY ARTERY. A CASE SERIES

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Abstract:
Unilateral absent pulmonary artery (UAPA) is one of the rare congenital anomaly seen in adulthood[1].Unilateral absence of pulmonary artery usually occurs with other congenital defects such as Tetrology of fallot or septal defects but it can also occur as a isolated anomaly which is very rare. Isolated absence of pulmonary artery is a rare cause of pulmonary hypertension.We report three cases of absent right pulmonary artery presenting with severe pulmonary hypertesion. Diagnosis needs high index of clinical suspicion and close scrutiny of radiological investigations for the diagnosis.

Key Words: Dyspnoea, Lungs, Edema, Right ventricular Hypertrophy.

Introduction :
Unilateral absent right pulmonary artery in isolation is one of the rare congenital anamoly with an estimated prevalance of 10 in 2 million adults[1]. Usually it occurs in association with congenital defects like tetrology of fallot, septal defects and coarctation of aorta but it can also occur in isolation[2]. Right pulmonary artey is most frequntly involved as compared to left[3]. It presents with varied manifestations. we report three cases of absent right pulmonary artery presenting with exertional breathlessness and on evaluation found to have severe pulmonary arterial hypertension.

Case 1 :
42 years old gentlemen presented with history of progressive symptoms of dyspnoea for 1 year, intermittant pedal edema for 2 months and atypical chest pain for 2 weeks. Examination revealed elevated Jugular venous pressure with Right ventricular apex and loud P2 suggesting pulmonary hypertension.Electrocardiogram(ECG) revealed sinus rhythm with right axis deviation and right ventricular hypertrophy with strain.

Chest X -ray showed cardiomegaly with right atrial enlargement,oligemic and reduced lung volume on right side as compared to left and inconspicious pulmonary vascular markings on right side with increased vacularity and lung volume on left side (Fig 1). At this stage possibilities considered were right pulmonary artery stenosis, absent right pulmonary artery and scimitar syndrome which can be associated with reduced lung volume on right side.

Echocardiogram showed dilated right atrium (RA), right ventricle (RV) with RV hypertrophy and mild Tricuspid regurgitation(TR) with intact septum and TR jet velocity is 109 mmHg (Fig 2). Inferior vena cava(IVC) was dilated and partially collapsing suggesting elevated RA pressures. Short axis view at great artery level revealed dilated RA, RV and main pulmonary artery(MPA) continues as left pulmonary artery and right pulmonary artery is not visualised and in suprastenal view right pulmonary artery is not seen (Fig 3) and at the usual pulmonary artery

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segment turbulance flow is seen and pulse wave doppler across the turbulant flow revealed low velocity and continuous flow suggesting the presence of collaterals. Computed Tomography (CT) Chest with contrast confirmed the above findings of absent right pulmonary artery (Fig 4).

To confirm the diagnosis and to delineate collaterals to right lung, invasive angiogram was done. Right ventricular and pulmonary artery pressures are suprasystemic. Invasive pulmonary angiogram showed main pulmonary artery continues as left pulmonary artery (LPA) and right pulmonary artery(RPA) is absent(Fig 5). Aortogram revealed small leash of blood vessels to right lung from the subclavian and vertebral arterial system and there is no evidence of right pulmonary artery arising from the aorta. Coronary angiogram showed huge collaterals from proximal right coronary artery to the right lung and hilum of right lung is supplied by left coronary artery (Fig 6).

Case 2:

26 years old gentlemen presented with progressive breathlessness of 6 months duration. Vital signs were normal and cardiovascular system examination was suggestive of pulmonary hypertension. Chest X ray revealed dextroposition of heart and rests of the X ray findings were similar to the previous patient (Fig 7). Echocardiography revealed dilated RA, RV and moderate TR with interatrial septum bulging towards left atrium suggesting elevated RA pressures and TR jet velocity is suprasystemic (Fig 8). IVC was dilated & non collapsing.RPA is not seen in suprasternal view and it was confirmed with CT pulmonary angiogram (Fig 9) and invasive angiography. Similar to the above case there is a huge collateral supply from the left coronary artery to right lung (Fig 10).

Case 3:

A 35 years old male previously asymptomatic presented with history of dyspnoea and fever since 1 week, Examination revealed Loud P2 and no other signs of pulmonary hypertension was present. His ECG shows sinus tachycardia, S1 Q3 T3 pattern with RV strain pattern. X ray shows absent RPA shadow, loss of right lung volume, homogenous opacities on right middle and lower zones (Fig 11). Echo shows Dilated RA, RV and TR Jet gradient of 50 mm of Hg, RPA not visible in Parasternal short axis view, MPA continuing as LPA. CT scan showed MPA continuing as LPA and absent RPA. Invasive pulmonary angiogram revealed absent right pulmonary artery (Fig 12).

Discussion:

The unilateral absence of pulmonary artery is one of the rare developmental anomaly seen at adulthood[1]. One third of adult patients with absent pulmonary artery will have an asymptomatic presentation. Often present in association with congenital defects like tetrology of fallot, septal defects and coarctation of aorta but can occur in isolation[2]. It occurs due to the altered development of 6th aortic arch[4]. Absent right pulmonary artery accounts for almost two third of cases[3]. Due to embryologic relationships, UAPA commonly occurs on the side of the chest opposite the aortic arch. Distal vessels in the affected lung are intact. These vessels are supplied by collateral vessels from bronchial, intercoastal, and internal mammary, subdiaphragmatic, subclavian, or even coronary arteries[5, 6].

These patients present with varied manifestations like dyspnoea on exertion, easy fatigability, chest pain, hemoptysis and recurrent respiratory tract infection[3]. In a review of 108 cases of unilateral absent pulmonary artery, 40% of patients were noted to have dyspnoea or effort intolerance. Pulmonary hypertension was seen in 44% of patients[3]. Two of our cases presented with dyspnoea on exertion and severe pulmonary hypertension.

Radiological investigations are key to the diagnosis. Chest X – ray shows oligemic and reduced lung volume on the affected side. Decreased or absent vascular markings in the affected lung. There can be displacement of heart towards the reduced lung volume. The lung with intact pulmonary artery will have increased vascularity and lung volume. When any of these findings are noted, CT pulmonary angiogram and transthoracic echocardiography can be used for definitive diagnosis as in our case. Transthoracic echo can also be used to diagnose UAPA (right pulmonary artery is not visualized in suprasternal and short axis view) and it has an advantage to look for coexisting cardiac anomalies. Invasive angiography is the gold standard for the diagnosis of UAPA. Other investigation which can be used is ventilation perfusion scan.

In UAPA the pulmonary hypertension is due to pulmonary arterial vascular remodeling due to the release of endothelin, which is a potent vasoconstrictor[7]. Endothelin stimulates smooth muscle proliferation, intimal hyperplasia, medial hypertrophy and collagenous replacement of intima in late stages.[7]

There is currently no established treatment of patients with UAPA. Patients with pulmonary hypertension can be managed medically with vasodilator therapy. [3, 7]. In pediatric population there are reports of revascularization of peripheral pulmonary branches of the affected pulmonary artery to the pulmonary hilum [2, 8, 9]. Massive Hemoptysis

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can be managed with embolization, lobectomy, or pneumonectomy[10, 11]. Severe infections may require lobectomy or pneumonectomy[5]

Conclusion:

Isolated absence of pulmonary artery is very rare. Clinicians should have high suspicion of UAPA in adults with unexplained hemoptysis and exertional dyspnoea. Diagnosis needs high index of clinical suspicion and close analysis of radiological investigations for the diagnosis.Early diagnosis and management of isolated UAPA in adult patients is crucial to avoid complications like massive hemoptysis and severe pulmonary artery hypertension in the long term.Patients with pulmonary hypertention needs vasodilator therapy. Massive Hemoptysis can be managed with embolization, lobectomy, or pneumonectomy. Severe infections may require lobectomy or pneumonectomy.

Aknowledgements: Nil

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Figure Legends

Figure 1: CXR PA view showing reduced vascularity and reduced lung volumes of right lung.

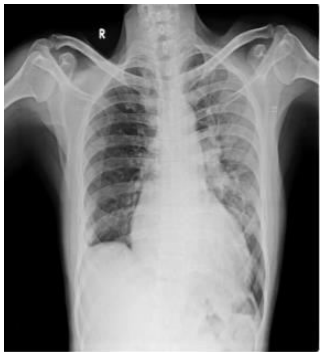

Figure 2: Echo Doppler showing dilatation of RA and RV and TR jet gradient of 109 mmHg.

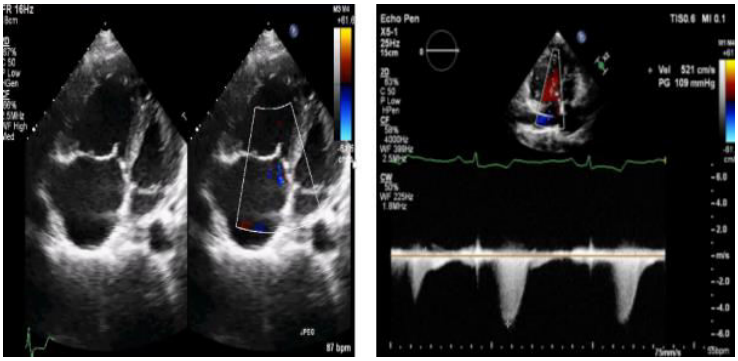

Figure 3: Echo in short axis and suprasternal view showing absence of right pulmonary artery.

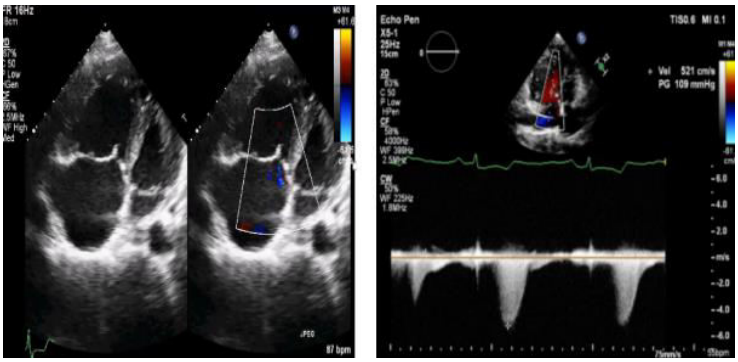

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Figure 4: CT scan showing absent right pulmonary artery.

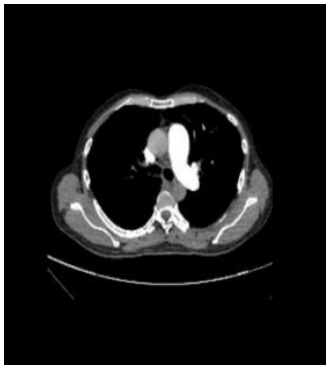

Figure 5: Angiogram showing absent right pulmonary artery.

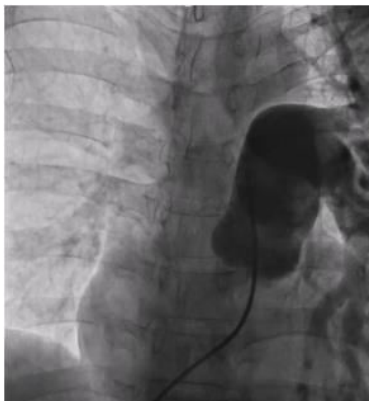

Figure 6: Coronary angiogram showing collaterals arising from right and left coronary arteries supplying right lung.

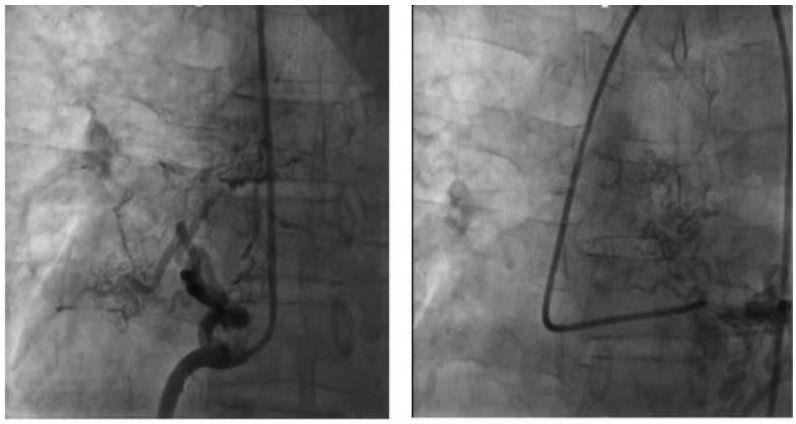

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Figure 7: CXR PA view showing reduced vascularity and lung volume of right lung.

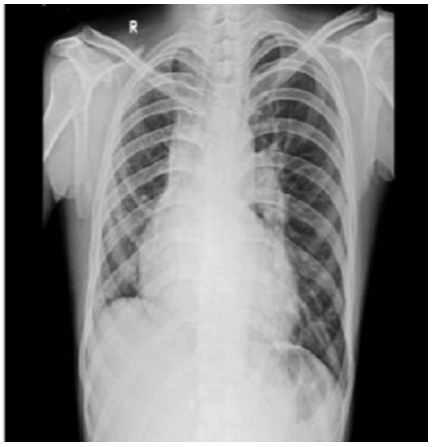

Figure 8: Echo Doppler showing dilated RA, RV and severe TR with severe PH.

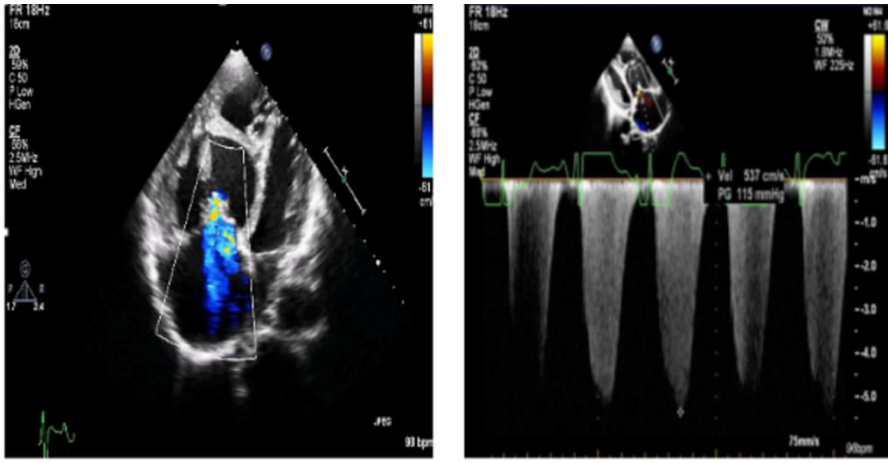

Figure 9: CT scan showing absent right pulmonary artery.

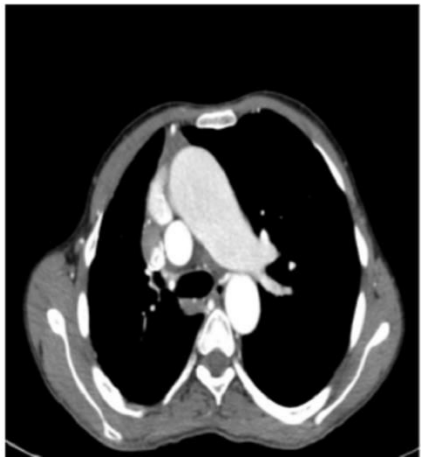

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Figure 10: Left coronary angiogram showing collaterals supplying right lung.

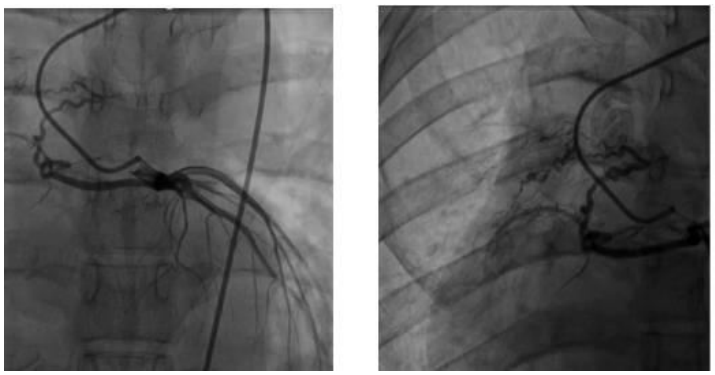

Figure 11: CXR PA view showing reduced vascularity and lung volume of right lung.

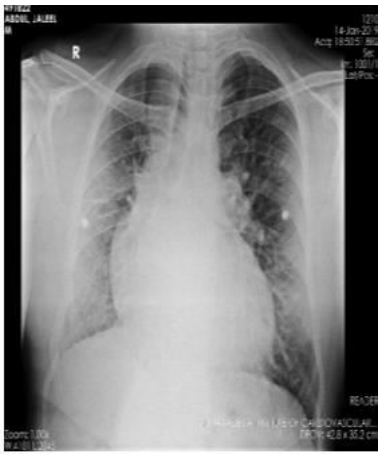

Figure 12: Angiogram showing absent right pulmonary artery.