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Original Research Paper

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A CASE REPORT OF RIGHT UNILATERAL PULMONARY ARTERY ATRESIA (UPAA) MISDIAGNOSED AS RIGHT CHRONIC PULMONARY THROMBO-EMBOLISM

Dr. Ronak Raval	M.ch. CVTS 3rd Year Resident
Dr. Amit Choudhary	M.ch. CVTS 3rd Year Resident
Dr. Aneesh Lawande	M.Ch. CVTS 3rd Year Resident
Dr. Chirag Doshi*	M.Ch. CVTS 3rd Year Resident *Corresponding Author

ABSTRACT Unilateral pulmonary artery atresia is a rare condition with clinical presentation as exercise intolerance, recurrent infections, hemoptysis to asymptomatic state with incidental diagnosis. Common modalities of management include medical management for pulmonary hypertension , angioembolisation of collaterals followed by pneumonectomy. However surgical reconstruction of atretic pulmonary artery segment is an uncommon surgical management strategy employed in our case with satisfactory outcome.

KEYWORDS : Unilateral pulmonary artery atresia, rare, surgical reconstruction,misdiagnosis, pulmonary thromboembolism

INTRODUCTION

Isolated unilateral pulmonary artery atresia UPAA is quite a rare disorder with prevalence of 1 in 200000 or less. UPAA is a defect believed to be caused by failure of the embryonic sixth aortic arch to fuse with pulmonary trunk during development. The clinical spectrum may vary from asymptomatic to exercise intolerance, recurrent infections, hemoptysis to severe pulmonary hypertension with possible fatal outcome [4].

Management is based on symptom management be it pulmonary hypertension, severe hemoptysis or recurrent infections. Management includes surgery in adults in form of lobectomy or pneumonectomy, whereas paediatric population can sometimes benefit from reconstruction if anatomically possible. Patients with no symptoms but incidental diagnosis are best left untreated, but followed.

Case Study

A 38 year old male patient presented with chief complaints of exertional dyspnoea, hemoptysis and palpitation for last 02 years with progressive increase in severity of symptoms. Laboratory investigations were within normal limits. Chest X-ray was suggestive of left sided reduced pulmonary markings and right lower zone consolidation changes[Figure 1].

2D Echo showed moderate RV dysfunction with severe TR and severe PAH with RPA complete occlusion by thrombus. CTPA showed RPA complete chronic occlusion just after origin with occlusion in right upper, middle and few lower lobe segmental branches [Figure 2 & 3] . There was diffuse narrow caliber with collateral refilling of right descending pulmonary artery and few right lobe segmental branches; left PA system was normal; few aorta-pulmonary collaterals were present.

Based on above investigations and clinical findings, decision was taken to perform surgery of pulmonary endarterectomy for RPA thrombotic occlusion. Intraoperatively, there were dense pericardial adhesions and RPA was found to be atresia and calcified; after going on CPB with cardioplegic arrest, on opening RPA, no thrombus was found.

After dividing aorta and SVC for adequate exposure, RPA opened up to hilum and reconstruction done with autologous pericardial patch.

Post operatively, patient made a steady progress and was discharged on POD 20. Post operative CECT Thorax, showed significant improved right pulmonary arterial flow. [Figure 4]

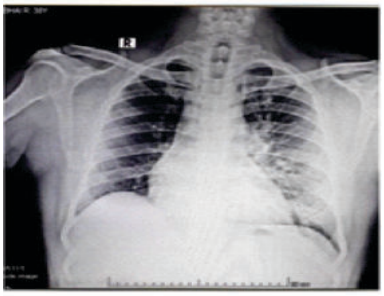

Figure 1

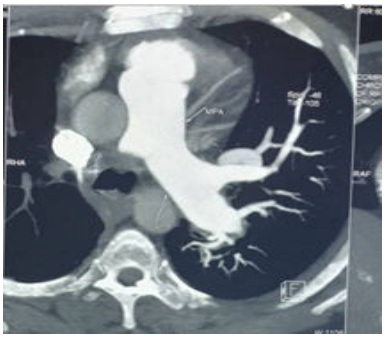

Figure 2

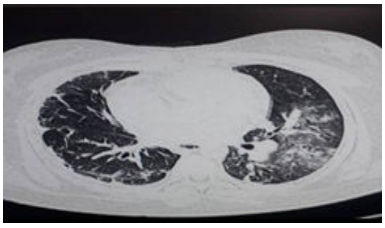

Figure 3

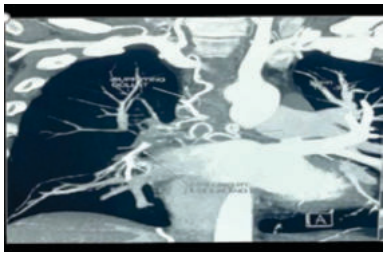

Figure 4

DISCUSSION

UPAA is a rare entity with approx 400 cases reported in literature with 65 reported since 1990 [3]. Adult patients present with hemoptysis or exertional dyspnoea typically but also chest pain, repeated LRTI or even high susceptibility for pulmonary oedema. The lung parenchyma on affected side can have bronchiectasis (particularly in patients with recurrent infections), interstitial changes or multiple bullae.

In keeping with an embryonic developmental abnormality as root cause, just under half of patients with absent left pulmonary artery will have right sided aortic arch and vice versa. Collateral pulmonary flow develops from many sources in thorax, including bronchial arteries (47%), the internal thoracic artery (44%), intercostal arteries (44%),the subclavian or axillary artery (34%),directly from aorta (13%), and oesophageal branches (9%) [2].

Therapies are directed first towards stabilizing the patient, with securing of the airway in unstable patients, and immediate management of any severe bleeding that could compromise ventilation. For the few patients that present in extremis, prompt intubation with bronchial blocking of the bleeding lung can be life saving. Angioembolization of active bleeding is then undertaken. Symptoms of bleeding and exertional dyspnea can be managed by selective angioembolization of large collaterals and/or medical management of pulmonary hypertension with beta-blockers, calcium channel blockers, or phosphodiesterase inhibitors in patients unable to tolerate surgical therapy. Pneumonectomy offers the best long term therapy in patients able to tolerate the procedure. While this may seem somewhat extreme, patients with a unilaterally absent pulmonary artery are functioning with little gas exchange occurring in the affected lung, so removal of the abnormal lung usually does not significantly affect their functional status. Consequently, pulmonary function tests and oxygen exchange evaluations should be interpreted cautiously, recognizing that the usual drop by 50% in gas exchange and FEV1 is not expected post-pneumonectomy [1]. In asymptomatic patients, close surveillance is also an appropriate approach, provided the patient is reasonably able to understand the potential for future problems and has ready access to adequate medical care.

CONCLUSIONS

UPAA in adults is an extremely rare disorder which presents typically with hemoptysis but also with recurrent infections, exertional dyspnoea or incidentally. Management includes close follow up with no intervention in otherwise symptomatic patients with incidental diagnosis to collateral embolization, pneumonectomy and medical management of pulmonary hypertension for those surgically unfit. However surgical reconstruction is uncommon form of surgical management attempted in our case with good outcome[5].

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