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Original Research Paper

Respiratory Medicine

# CASE STUDY: 41-YEAR-OLD FEMALE WITH PROXIMAL INTERRUPTION OF RIGHT PULMONARY ARTERY (PIRPA) WITH PATENT DUCTUS ARTERIOSUS (PDA)


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**ABSTRACT** Proximal Interruption of the Right Pulmonary Artery is an un-commonest developmental anomaly and an estimated prevalence is around 1 in 200,000 young adults. Clinical manifestations can range from being asymptomatic to having massive hemoptysis. This is a case report of a 41-year-old female with right-sided Proximal Interruption of the Right Pulmonary Artery (PIRPA) confirmed on CT-Pulmonary Angiogram (CT-PA) and discharged with conservative management.

**KEYWORDS :** Proximal Interruption of the Right Pulmonary Artery, Patent Ductus Arteriosus (PDA), CT-PA, mMRC, ATT, ABG

## Case Report

This is a case report of a 41-year-old female, who presented with breathlessness for the past 6 months to the emergency unit of Respiratory Medicine on 1st of March, 2021. The patient was in progressing stage of the modified Medical Research Council dyspnoea scale (mMRC) grade IV in the last 1 week, not associated with orthopnoea or reduced urine output.

## Primary Evaluation

### Patient's History

The patient's history was recorded, and she has a history of pan digital clubbing of grade III since childhood, non-foul smelling, non-blood stained, productive cough with mucoid expectoration for past 3 months, no loss of weight, and intake of ATT for 6 months 6 years back. She has been amenorrheic for the past 4 months and had abortions twice in the first trimester. She has one daughter 19 years of age.

### Physical and Clinical Examination

On initial physical examination, she presented with pulse rate and tubular breath sounds of 80/min, Initial Arterial Blood Gas (ABG) showed severe type I respiratory failure with SpO2 of 48mmHg in room air, Blood Pressure 120/80mmHg, and a hyper-pigmented rash over the malar region. The patient's temperature was normal. On auscultation, left-sided cavernous breath sounds with tubular breath sound over the right side with inspiratory crepitations were heard. She had significant biomass exposure with a biomass index of 80.

### Biomarkers

Lab investigations showed polycythemia with Hb 17g/dl and hematocrit of 62.8%, elevated infective markers Total Count (TC)-15170, C-Reactive Protein (CRP)-121, elevated lactates-87 and dilutional hyponatremia. Pan cultures showed no growth and the COVID-19 swab was negative.

### Initial Findings

From the patient's history, physical, clinical, and biomarker investigations, the initial findings were a diagnosis of severe type I respiratory failure, an infective exacerbation of bronchiectasis, pan digital clubbing, polycythemia, elevated infective markers, dilutional hyponatremia, pulmonary tuberculosis, recurrent abortions,

### Symptoms that led us to hidden undiagnosed disease

Pointing out to the history of recurrent abortions, it was then we decided to send samples for ANA and Antiphospholipid antibodies tests. Both tests were positive, and our doubt about the presence of hidden undiagnosed disease was confirmed, so we further investigated by conducting a CT-Pulmonary Angiogram (CT-PA) and the findings were given in figure 1-4.

## Confirmative Evaluation

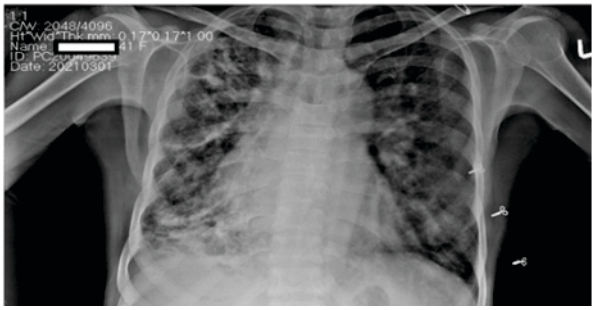

**Figure 1 shows the Chest X-ray (CXR)**

Figure 1 shows the presence of bilateral hyper-inflated lung fields with volume loss in the right lung of the patient.

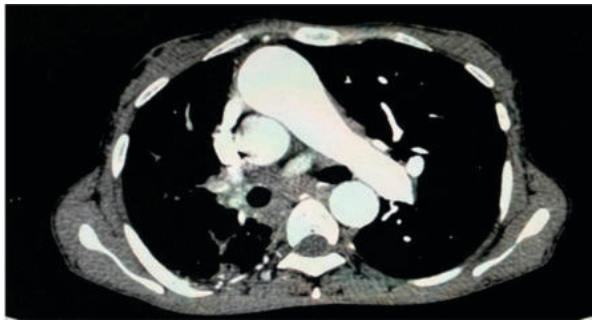

**Figure-2 (A): CT-Pulmonary Angiogram**

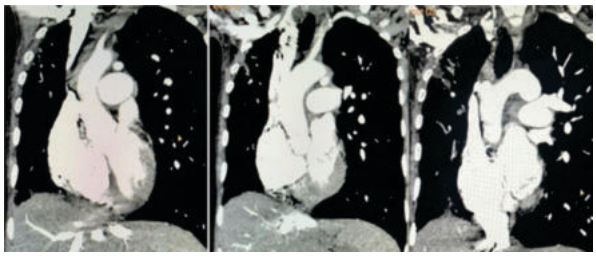

**Figure-2 (B): CT-Pulmonary Angiogram**

Figure 2 (A & B) shows the dilated main pulmonary and left pulmonary arteries with **non-visualization of the right pulmonary artery;** the right lung was perfused by multiple collaterals and a patent ductus arteriosus with an atrial septal defect. 2D Echocardiogram (2D-ECHO) shows RA/RV dilation with severe pulmonary hypertension (PASP of 95mmHg). Spirometry had shown a restrictive pattern with an FVC of 52%.

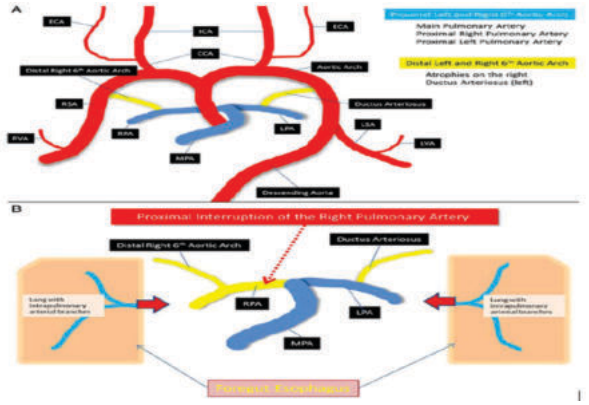


**Figure-3 (A & B): Diagrammatical Representation of Proximal Interruption of Right Pulmonary Artery (PIRPA)**

Figure 3 (A & B) depicts normal embryological development of the 6th aortic arches and Figure (B) shows abnormality resulting in right-sided PIPRA.

### Confirmatory Report

This 41-year-old female patient is with Proximal Interruption of the Right Pulmonary Artery (PIPRA) with Patent Ductus Arteriosus (PDA).

### On Discharge

As the patient was initially admitted for infective exacerbation of bronchiectasis this was treated and recovered with adequate antibiotics. Despite clubbing since her childhood and she was able to perform her daily routines with no difficulty, she was discharged as she was symptomatically improving with home Long-Term Oxygen Therapy (LTOT) to maintain a target SpO2 of 85%, T. Sildenafil TDS for Group I Pulmonary Hypertension and to continue on pulmonary vasodilators.

## DISCUSSION

The estimated prevalence of Proximal Interruption of the Pulmonary Artery across the globe is around 1 in 200,000 young adults, predominantly found on the right side [1]. PIRPA is an uncommon anomaly and is often an incidental finding in an asymptomatic patient, but in symptomatic patients, dyspnea with reduced exercise intolerance, recurrent infections, and pulmonary hypertension are the critical prognosticator [2].

During embryogenesis, PIRPA is formed by the developmental arrest in the proximal 6th aortic arch. Prenatally, blood flow is maintained through a persistent connection of the right intra-pulmonary PA to the distal 6th aortic arch via the right-sided ductus arteriosus, due to this phenomenon, the restricted or abrogated growth of the corresponding intrapulmonary PA occurs [3].

In patients with inadequate bronchial collateral supply, the collateral vessels from inter-costal arteries may develop pleural adhesions and directly anastomose with branches of the pulmonary artery that will limit the flow of the blood to the affected lung. The distal portion of the left pulmonary arch develops normally and the left ductus remains patent due to peripheral blood flow condition [4].

Our patient showed severe type I respiratory failure with SpO2 of 48mmHg in room air at the time of admission, but Saladi L et al study showed 92% oxygen saturation was found in the patient with mildly dilated pulmonary artery, indicating the difference in dilation and interruption [5]

As per Weldetsadik, A. Y et al study, no consensus was formed for asymptomatic proximal interruption of the pulmonary artery, the treatment of asymptomatic patients was to treat infection, and vasodilators, for our patient we recovered from the infection, recommended long-term oxygen therapy, and T. Sildenafil TDS, hence from this study, we wanted to suggest that global consensus has to be formed and followed for patients with asymptomatic proximal interruption of pulmonary artery [6].

Diagnosis of asymptomatic proximal interruption of the pulmonary artery is difficult says the Khalilian, M., et al study, and we also with excellent difficulty ruled out this case report with the help of the patient's history, symptoms, physical, and clinical examination, biomarkers, initial findings, and confirmative evaluation such as chest X-ray, spirometry, and CT-Pulmonary Angiography [7].

## CONCLUSION

PIRPA is an uncommon developmental anomaly often overlooked, undiagnosed, and absences of consensus for asymptomatic patients, hence the clinicians must rule out exactly any diseases in the patients with the help of the patient's history, symptoms, physical, clinical examination, biomarkers, X-ray, radio-imaging, CT and MRI.

We suggest that for patients with Proximal Interruption of the Pulmonary Artery which is also known as Unilateral Pulmonary Artery Atresia (UPAA), the confirmative investigation for correct diagnosis must be CT or MRI. A consensus for asymptomatic patients will also improve the Quality of life and reduce the morbidity and mortality in patients with Proximal Interruption of the Pulmonary Artery.

## REFERENCES

1. Williams, E. A., Cox, C., Chung, J. H., Grage, R. A., & Rojas, C. A. (2019). Proximal interruption of the pulmonary artery. Journal of Thoracic Imaging, 34(1), 56-64.
2. Rojas, C. A. (2019). Proximal Interruption of the Pulmonary Artery. Journal of Thoracic Imaging, 34(5), W129-W130.
3. Cambronero-Cortinas, E., Rivero-Jiménez, N., Bret-Zurita, M., Moratalla-Haro, P., & García-Guereta Silva, L. (2019). Unusual distal right pulmonary artery origin from right ductus arteriosus with uncommon left-sided aortic arch. Revista Colombiana de Cardiología, 26(5), 300-303.
4. Mukherjee, A., Pandey, N. N., Ramakrishnan, S., & Jagia, P. (2022). Proximal interruption of the right pulmonary artery: A rare anomaly. Journal of Cardiac Surgery, 37(7), 2094-2095.
5. Saladi, L., Roy, S., & Diaz-Fuentes, G. (2018). Unilateral pulmonary artery agenesis: An unusual cause of unilateral ARDS. Respiratory medicine case reports, 23, 148-151.
6. Weldetsadik, A. Y., Asfaw, Y. M., & Tekleab, A. M. (2018). Isolated absence of right pulmonary artery in a 4-year old child: a case report. International Medical Case Reports Journal, 11, 297.
7. Khalilian, M., Langroudi, T. F., Dabbagh, A., Tehrani, R. B., & Tahouri, T. (2022). Interruption of the Right Pulmonary Artery in a Neonate. Case Reports in Cardiology, 2022.