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Isolated Unilateral Absent Branch Pulmonary Artery with Peripheral Pulmonary Stenosis and Coronary Artery Disease

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ABSTRACT

Isolated Unilateral Absent Pulmonary Artery (UAPA) is a rare congenital anomaly. It is usually associated with congenital heart defects. A 45 year old male patient presented with complaints of fever with cough and expectoration for 15 days and retrosternal chest discomfort for the previous 2 days. ECG showed diffuse ST segment depression with T wave inversion in the inferior and lateral leads. Coronary Angiogram done through the right femoral approach revealed diffusely diseased Left Anterior Descending (LAD) artery that was totally cut off at the mid segment. The Left Circumflex (LCx) artery was providing blood supply to the right middle and lower lung areas. There was another collateral arising from the Left Subclavian Artery supplying the right middle and lower lung areas. The left pulmonary artery was normal, but branches supplying the middle and lower lobes of the right lung were absent and the upper lobe branch had pulmonary stenosis. UAPA is a rare clinical entity; collaterals from coronaries are extremely rare in this condition and till now there has not been any case report of unilateral absent branch pulmonary artery with peripheral stenosis of other branches, on the affected side and associated coronary artery disease.

1. Introduction

Isolated Unilateral Absent Pulmonary Artery is a very rare congenital anomaly, with an incidence of 1 in 200000 (1). It is usually associated with other congenital heart defects like Tetrology of Fallot (2), Septal defects (3), Subvalvular aortic stenosis (4), Transposition of Great Arteries (1), Congenitally Corrected Transposition of great arteries with pulmonary stenosis (5), and Scimitar Syndrome (6); it can also be an isolated defect. When it is isolated, it can be asymptomatic (7) or can present in adult life with recurrent chest infections, hemoptysis, symptoms and signs of pulmonary hypertension and left ventricular failure (8). Here, we present a case of an adult male who had Isolated Unilateral absence of branch pulmonary artery with peripheral pulmonary stenosis who had developed collaterals from the coronary artery.

2. Case Presentation

A 45 year old male patient presented with complaints of fever with cough and expectoration for 15 days and retrosternal chest discomfort suggestive of unstable angina over the previous 2 days. Chest pain was associated with breathlessness that was aggravated by supine position (NYHA Class IV). He had a history of repeated respiratory tract infections that were treated by local physician. The patient was a smoker (one pack-year is equal to smoking 20 cigarettes per day for a year) and a tobacco chewer. He was not a diabetic and normotensive. He didn't have a family history of premature Coronary Artery Disease.

On examination, his blood pressure was 110/70 mm of Hg, heart rate was 110/min and he wasn't maintaining adequate saturation on the room air; hence, he was put on supplemental O2 of 6L, following which his SpO2 was 92%. Cardiovascular system was normal on examination and he had bronchial breath sounds in the left inter scapular area along with inspiratory crepitations in the bilateral basal lung fields.

Chest X ray PA view showed haziness in the left upper zone. ECG showed diffuse ST segment depression with T wave inversion in the inferior and lateral leads. A provisional diagnosis of LRTI (Lower Respiratory Tract Infection) with

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ACS (Acute Coronary Syndrome) was made and IV antibiotics and Inj. UFH (Unfractionated Heparin) was started for him. The total leukocyte count and renal parameters were normal. Hemoglobin (Hb) and Packed Cell Volume (PCV) were elevated. Serial cardiac enzymes of CK-MB and Troponin I were negative. Hence, once the patient was hemodynamically stable and LRTI had resolved clinically, he was sent for a coronary angiogram (Figures 1, 2).

Coronary Angiogram was done through the right femoral approach using 6F JL 3.5 and AR 2 diagnostic catheters. The left coronary angiogram revealed a normal left main artery and a diffusely diseased Left Anterior Descending (LAD) artery that was totally cut off at the mid segment. The entire Left Circumflex (LCx) artery was providing blood supply to the right middle and lower lung fields. The Right Coronary Artery (RCA) was dominant, normal and it was providing collaterals to LAD. There was another collateral arising from the left subclavian artery supplying the right middle and lower lung fields. Then, we subjected the patient to the right heart catheterization for evaluation of the pulmonary anatomy. The left pulmonary artery was normal, but the branches supplying the middle and lower lobes of the right lung were absent and the upper lobe branch had pulmonary stenosis (Figures 3, 4, 5).

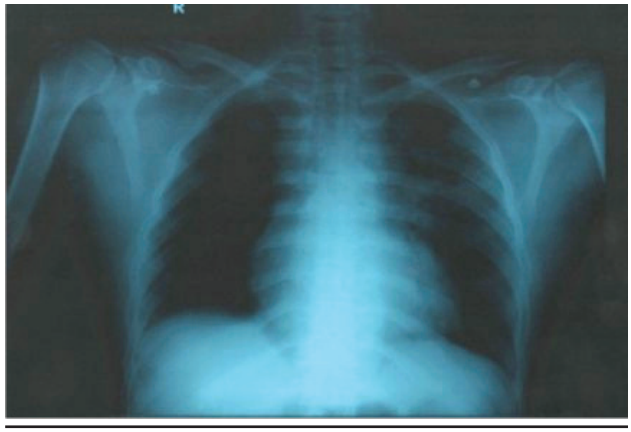

Figure 1. CXR Showing Left Upper Zone Haziness

3. Discussion

In 1868, Frantzel (9) first described a condition where unilateral pulmonary artery was absent. Since then, there have been several case reports about this very rare condition. The failure of the sixth aortic arch to connect with the pulmonary trunk results in this congenital anomaly (10).

A review by Harken et al. (11) of all cases published till 2002 found a total of 108 patients from 62 articles, in whom either the right or left pulmonary artery was involved. In this review, 62% or two thirds of the cases were those involving the right side of the lung and the median age of presentation was 14 years (although the range was very wide from less than a year to 58 years of age). About 15% to 30% of cases are usually reported to be asymptomatic (1). The most common presentations overall were repeated respiratory tract infections (37% of cases) and exercise intolerance due to pulmonary hypertension (40% of cases). The infections are usually mild, but they can progress in a

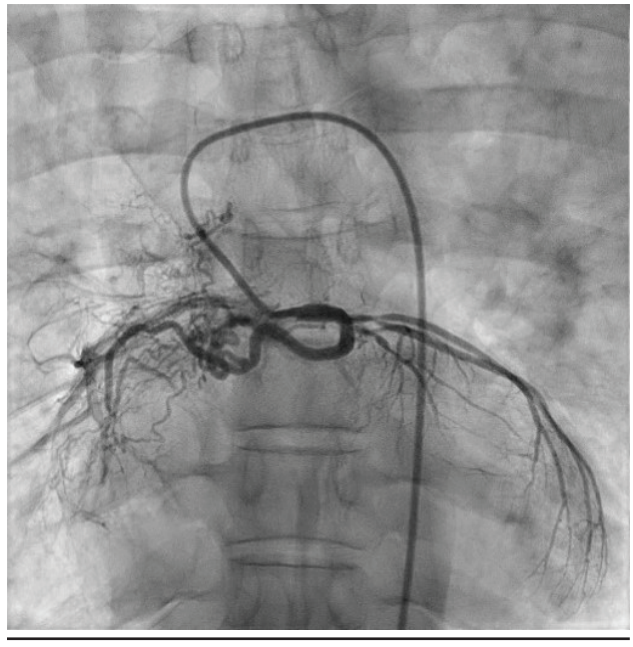

Figure 3. LCX Draining to Right Middle and Lower Lobes

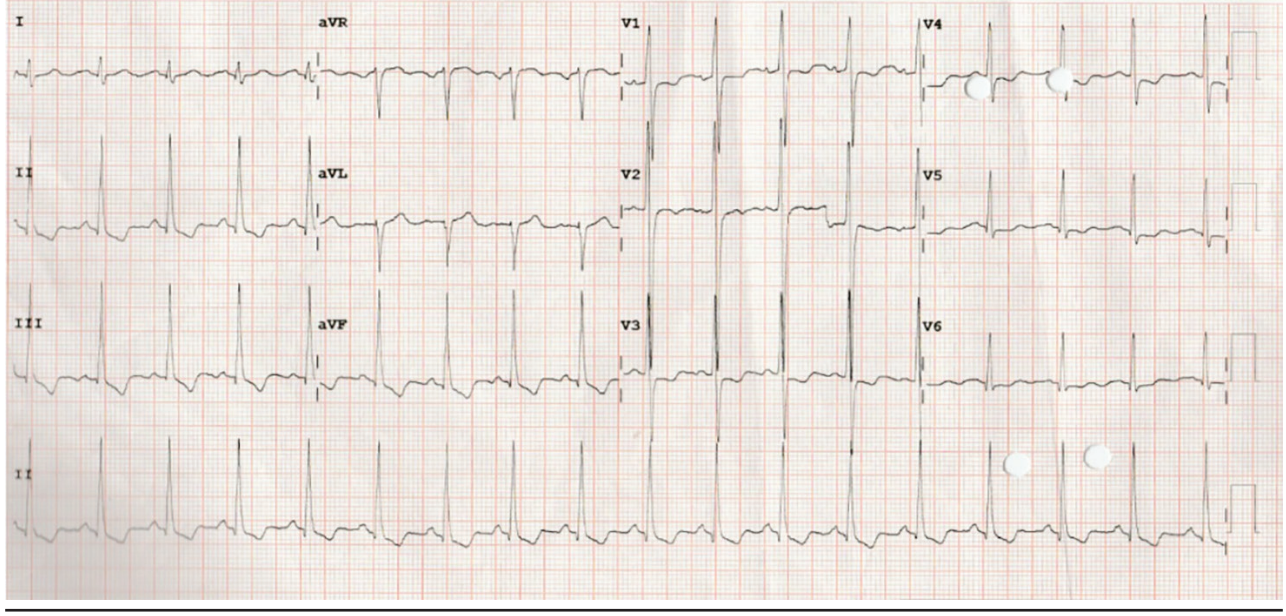

Figure 2. Electrocardiogram (ECG) Showing ST Depressions in V3-V6 and II, III, aVF

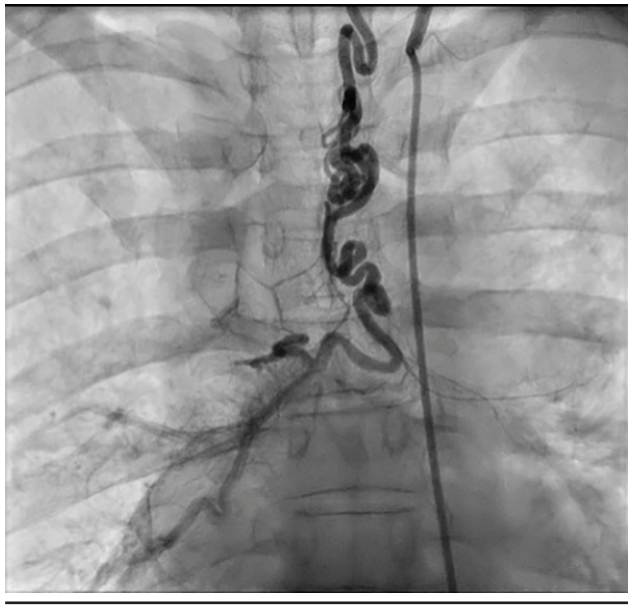

Figure 4. Collateral from Left Subclavian Draining to the Right Middle and Lower Lobes

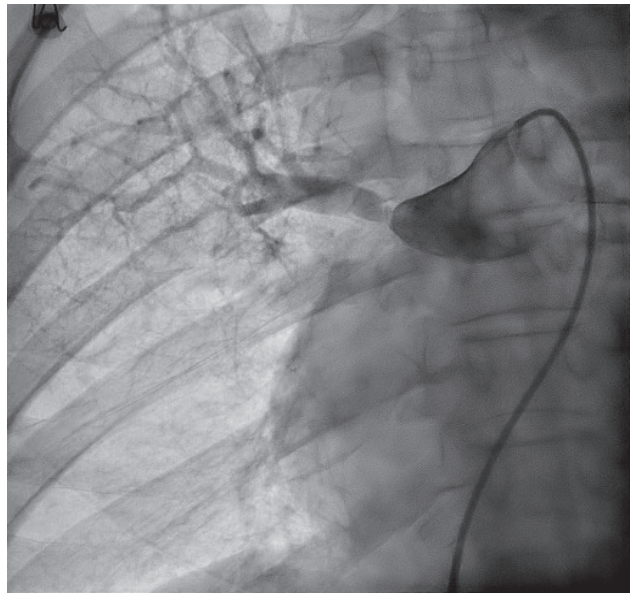

Figure 5. Absent Right Middle and Lower Lobe Branches of the Right Pulmonary Artery with Peripheral Pulmonary Stenosis of the Upper Lobe Branch

few cases to necrotizing bronchopneumonia (12). The main reasons for recurrent respiratory tract infections are alveolar hypocapnia causing bronchoconstriction and impaired mucocilliary clearance combined with decreased supply of inflammatory cells (1). Less common presentation includes hemoptysis, which is due to collaterals (13). Collaterals arise from Bronchial, Intercostal, Subclavian and Sub-diaphragmatic arteries. Collaterals from the coronary arteries are very rare; till now only six case reports have been published (14-19). Of these, three cases revealed the absence of the right sided pulmonary artery. Only two of the cases had evidence of myocardial ischemia.

In our cases, there was congenital absence of the branch pulmonary artery along with peripheral stenosis of the other branch pulmonary artery of the same side associated with coronary artery disease of the left anterior descending artery. Till now, there haven't been any case reports of such a unique presentation. There is a long delay between the onset of symptoms to the diagnosis of UAPA, due to mainly non-specific nature of symptoms. In the 2002 review by Harken et al, there was a median time delay of 30 years between the onset of symptoms and diagnosis of the disease.

Many diagnostic modalities are available for evaluation of UAPA starting with Chest X Ray. X ray chest findings that are suggestive of UAPA include absent hilar shadow, shrunk affected lung, shift of mediastinum towards the affected lung, and elevation of diaphragm on the affected side. A two D echocardiography contributes to exclusion of the cardiac abnormalities and also may confirm the diagnosis. High resolution CT scan can diagnose the condition and also contributes to identifying Bronchiectasis changes in the lung parenchyma. Cardiac catheterization helps to diagnose the condition and is usually performed when revascularization is planned. Pulmonary venous wedge angiography can demonstrate the hilar arteries.

Several patients of UAPA can be asymptomatic, but development of pulmonary artery hypertension leads to significant morbidity and mortality of about 7%. Patients with pulmonary hypertension can benefit from revascularization. There have been reports of successful revascularization being done in pediatric population that significantly improved the symptoms (20). If revascularization is not possible, then such patients can be treated as cases of primary pulmonary hypertension with long term vasodilator therapy. In our case, since the patient did not have pulmonary hypertension or hemoptysis, he underwent Bypass Grafting for the disease Left Anterior Descending Artery only.

3.1. Conclusion

UAPA is a rare clinical disorder that remains undiagnosed for long periods of time due to the non-specific symptoms and signs. In the setting of repeated respiratory tract infections, hemoptysis and exercise intolerance, the condition may be diagnosed using Chest X ray, Two D Echo cardiography and CT Chest. Collaterals from coronaries are extremely rare in this condition and till now there has not been any case report of unilateral absent branch pulmonary artery with peripheral stenosis of the other branch, on the affected side, and associated coronary artery disease. Appropriate diagnosis helps to devise treatment options that greatly improves the symptoms.

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Authors' Contribution

1. Dr. Sunil Abhishek B- Review of literature, collecting data, preparation of manuscript
2. Dr. Vijay Sai C – Review of Literature

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