Case Report

Unilateral Absence of Pulmonary Artery in an Adult Patient Presenting with Haemoptysis : A Case Report with Brief Review of Literature

Tony Ete[1], Swapan Saha[2], Vanlalmalsawmdawngliana Fanai[3], Arun Kumar[3], Habung Mobing[4], Shakeel Ahamad Khan[3], Utpal Kumar[3], Narang Naku[5], Animesh Mishra[6]

Unilateral absence of pulmonary artery should be considered in patients presenting with haemoptysis and recurrent respiratory infections. Usually it is a diagnosis of exclusion. Patient may present with non specific symptoms. A high index of suspicion with proper investigations, non invasive as well as invasive, are required for diagnosis and management. The diagnosis is usually confirmed by CT and MRI. Angiography is done only for patients who require embolisation or revascularisation surgery.

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Unilateral absence of a pulmonary artery (UAPA), a rare condition, usually occurs in combination with other cardiovascular conditions like tetralogy of Fallot (TOF) or septal defects. Patients with isolated absence of one pulmonary artery often present with dyspnoea, chest pain, hemoptysis or recurrent chest infections but may be asymptomatic till late adulthood. About 20% of the patients develop inconsequential hemoptysis, although massive hemoptysis is very rare[1]. Therefore, diagnosis may be difficult due to these nonspecific presentation[2], sometimes diagnosed incidentally on chest radiographs. Here we are presenting a case with isolated absent right pulmonary artery with haemoptysis, as clinical presentation.

Case Report

A 37-year gentle man, non hypertensive, non diabetic and non smoker, presented with recurrent episodes of hemoptysis for the last six months, scanty in amount. There was no history of fever, breathing difficulty or recurrent chest infection. No history of pulmonary tuberculosis in the past. Physical examination revealed body temperature of 36.6°C, pulse rate of 90 beats per minute, respiration rate of 16 per minute and blood pressure of 100/70 mmHg. Clinically there were no signs of cyanosis, edema, or clubbing of the fingers. Cardiac and chest auscultation was normal. The electrocardiogram revealed normal sinus rhythm.

Department of Cardiology, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences, Shillong 793018
[1]MBBS, MD, DM (Cardiology), Assistant Professor and Corresponding Author
[2]MBBS, MD, DM (Cardiology), Consultant, Department of Cardiology, Desun Hospital Siliguri
[3]MBBS, MD, Senior Resident
[4]MBBS, MD, Senior Resident, Department of Medicine
[5]MBBS, MS, Assistant Professor, Department of General Surgery
[6]MBBS, MD, DM (Cardiology), Professor and Head
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Editor's Comment :

- **Unilateral absence of pulmonary artery (UAPA) should be considered in patients presenting with hemoptysis and recurrent respiratory tract infections.**
- **A high index of suspicion is required for diagnosis. It is usually a diagnosis of exclusion. Chest radiograph may suggest the diagnosis whereas echocardiography can be used for the evaluation of possible associated cardiac anomalies and assessment of pulmonary hypertension.**
- **The diagnosis is usually confirmed by CT scan and MRI. Angiography is done for patients who require embolisation or revascularization surgery.**

Echocardiogram was normal with normal pulmonary artery pressure except for absent right pulmonary artery. A chest radiograph (postero-anterior view) revealed dilated artery (?Main pulmonary) in the right hilum with alveolar infiltrates in right lung field. The right hemi diaphragm was elevated without any cardiac and mediastinal displacement (Fig 1).

Computed tomography (CT) angiogram revealed the absence of the right main pulmonary artery. A focal vascular dilation was detected in right lung possibly representing an aneurysmatic formation or an arteriovenuous fistula (Fig 2 A). Blood was supplied to the right lung by tortuous, dilated arterial branches of indeterminate origin (Fig 2 B).

In view of patient's symptom and to look for source of blood supply to the right lung, patient was taken for catheterization and pulmonary angio and aortogram was done with an intention to proper decision making for the management. Pulmonary angiogram showed absent right pulmonary artery (Fig 3 A). Selective angiogram of right subclavian artery showed upper zone of right lung is supplied by right vertebral artery and lower zone is supplied by right internal mammary artery (Fig 3 B).

As the patient's symptom improved thereafter, he was kept under close follow-up on an outpatient basis, with symptomatic

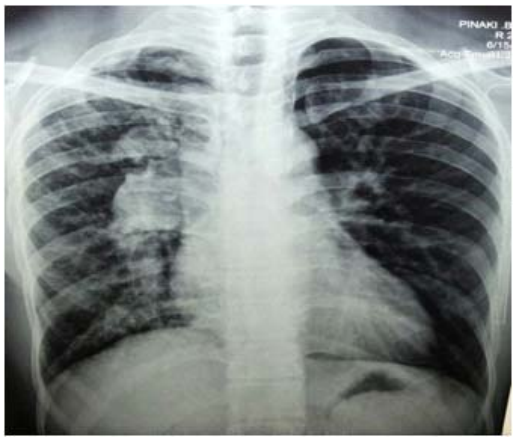

Fig 1 — Chest radiograph (postero-anterior view) shows dilated artery in the right hilum with alveolar infiltrates in right lung field with elevated right hemi diaphragm

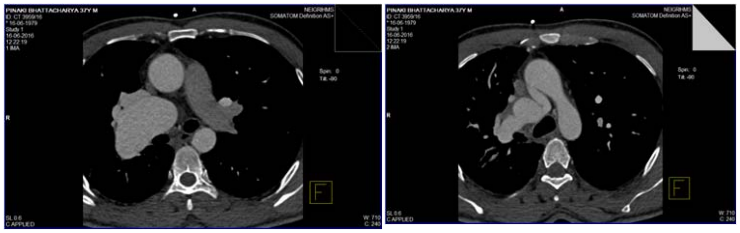

Fig 2 — Transaxial CT angiogram with intravenous contrast in a soft tissue window (A) shows an absent left pulmonary artery (B) tortuous, dilated arterial branches supplying right lung

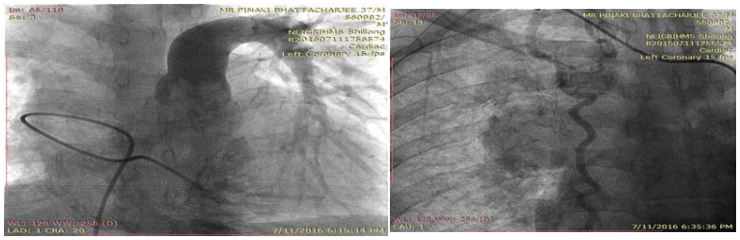

Fig 3 — (A) Pulmonary angiogram showed absent right pulmonary artery Selective angiogram of right subclavian artery showed upper zone of right lung is supplied by right vertebral artery and lower zone is supplied by right internal mammary artery

and supportive treatment for recurrent haemoptysis.

Discussion

The prevalence of UAPA is 1 in 200,000 young adults[2] and it can occur in an isolated manner although most cases are associated with other cardiovascular anomalies like tetralogy of fallot[2,3]. In about two third of cases, isolated UAPA involves the right lung[1]. Developmental alteration of ventral bud of the ipsilateral 6th aortic arch is thought to be the embryological basis of UAPA[4]. In the affected artery, generally the distal intrapulmonary branches remain intact which can be supplied by collaterals from bronchial, intercostal, internal mammary, subdiaphragmatic, subclavian or even coronary arteries[5]. Clinical course of many patients with isolated UAPA is benign and a diagnosis is not made until adulthood[2]. In symptomatic patients, one study showed, chest pain, pleural effusion and recurrent infections to be present in 37% of patients, while dyspnea or exercise intolerance in 40% of patients. Pulmonary hypertension was present in 44% of patients. Hemoptysis occurred in about 20% of patients whereas high-altitude pulmonary edema was seen in approximately 10% of patients[1]. Hemoptysis is caused by collateral circulations that create high pressures in venous system[5]. The systemic collaterals usually arise from the bronchial, intercostals, subclavian or subdiaphragmatic arteries[1]. The diagnosis of UAPA is, in generally, based on history, physical examination and findings on chest radiographs. Pulmonary function test in patients with UAPA is usually unremarkable[6]. CT thorax with contrast enhancement confirms the absence of the affected pulmonary artery. Echocardiography is helpful for exclusion of other cardiac anomalies and pulmonary hypertension. Pulmonary angiography is the gold standard and is usually reserved for patients requiring embolisation or revascularisation surgery[7].

At present there is no consensus regarding management of patients with UAPA. Some recommends serial echocardiography of asymptomatic patients for the development of pulmonary hypertension[8]. On the other hand, revascularization of peripheral branches to the pulmonary hilum can be attempted[9]. Hemoptysis can be managed with embolization, lobectomy or pneumonectomy[10].

In conclusion, UAPA should be considered in patients presenting with haemoptysis and recurrent respiratory infections. Chest radiograph may suggest the diagnosis whereas echocardiography can be used for the evaluation of possible associated cardiac anomalies and assessment of pulmonary hypertension. The diagnosis is usually confirmed by CT and MRI. Angiography is done only for patients who require embolisation or revascularisation surgery.

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