UNUSUAL CAUSE OF RECURRENT HEMOPTYSIS

Dr.K.R.Senthamizh
Department of respiratory medicine
Dr.D.Y.Patil Medical College Hospital and Research Institute

CHIEF COMPLAINTS

34 years old female, No comorbidities

Presented to us on 5.2.2022 with

- Multiple episodes of hemoptysis (each 50-100ml) for 6 days
- Dyspnea on exertion mMRC grade1 for 6 days
- No cough, chest pain, fever, loss of weight or loss of appetite
- No history of any past respiratory illness

CLINICAL EXAMINATION

VITALS

General Examination was WNL

Temp : 98.1F

PR : 112 BPM , regular, good volume , all peripheral pulses well felt.

RR : 19 breaths /min

BP : 120/70 mmHg , right arm , supine position

Spo2 : 98% RA

SYSTEMIC EXAMINATION

RS : *Mild tracheal shift to right*
B/L NVBS
Right infra-scapular fine crepitations

CVS : S1, S2 heard
No murmurs

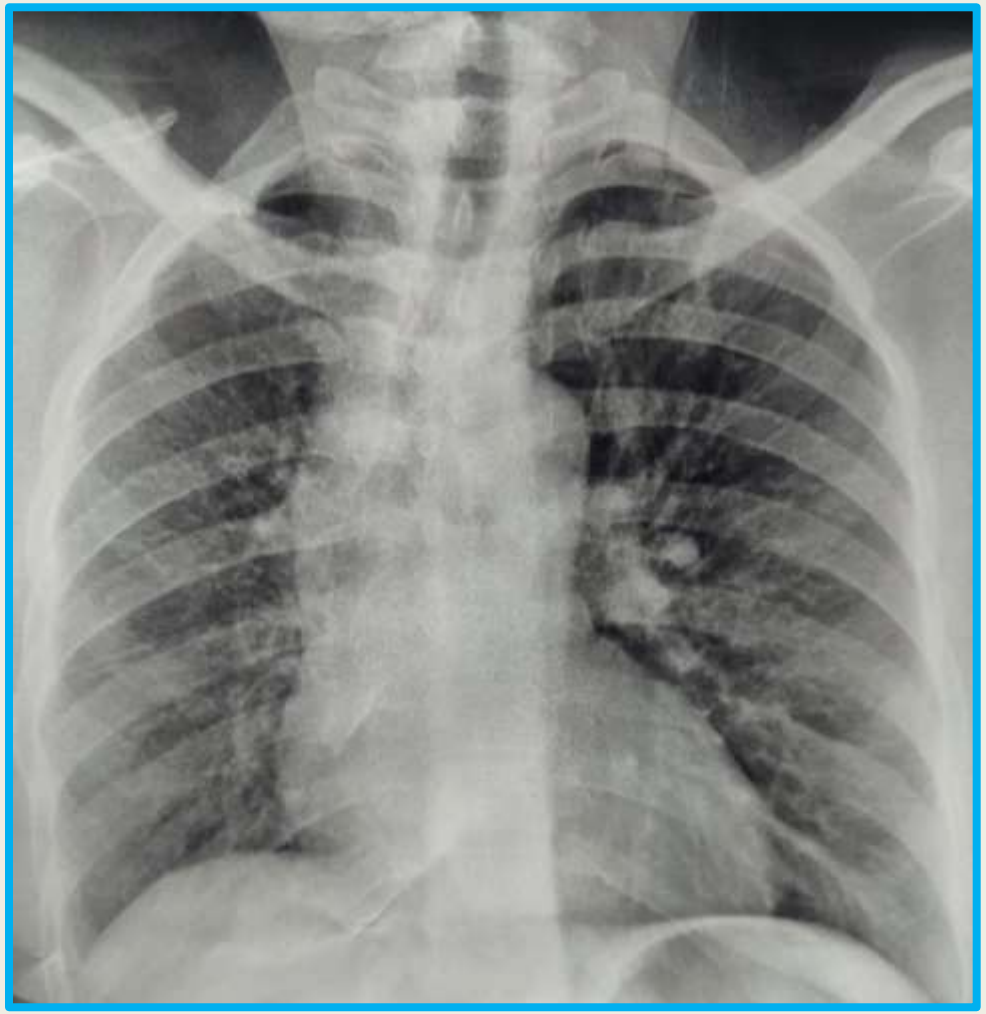
P/A : Soft
No organomegaly

CNS : No focal neurological deficit

INVESTIGATIONS

Chest X-ray

- Right lung volume loss
- Compensatory left lung hyperinflation
- Mild mediastinal shift to right



INVESTIGATIONS

ECG showed sinus rhythm

CBC showed normocytic normochromic anemia with Hb of 7.8

2D ECHO - Mild pulmonary artery hypertension with no septal defects

All hematological and biochemical parameters were normal

PT/INR, apTT and BT/CT was normal

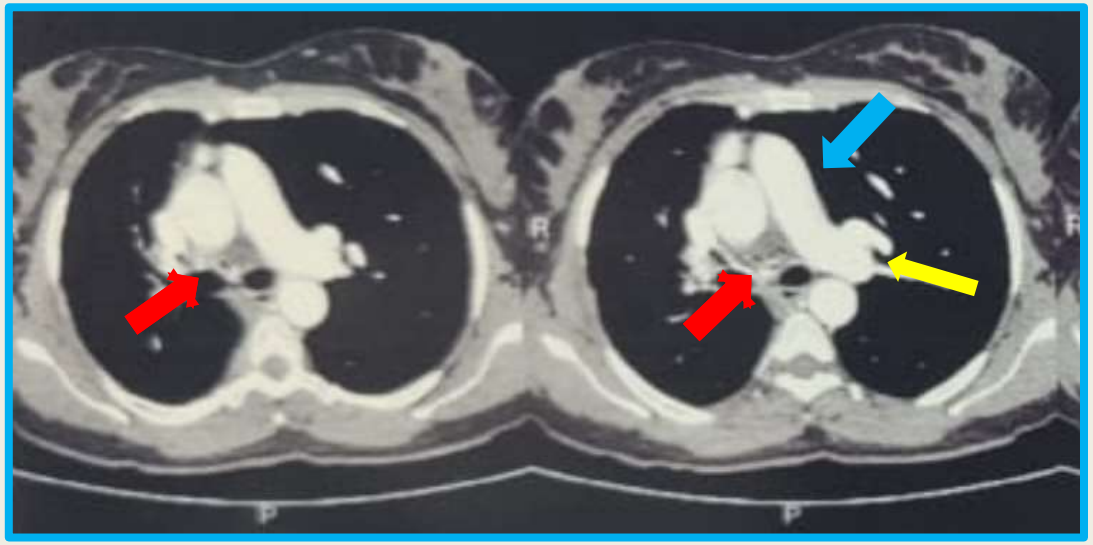
INVESTIGATIONS

CT-PULMONARY ANGIOGRAPHY

No evidence of pulmonary thromboembolism.

Main pulmonary artery measures 2.3cm and left pulmonary artery 2.1cm.

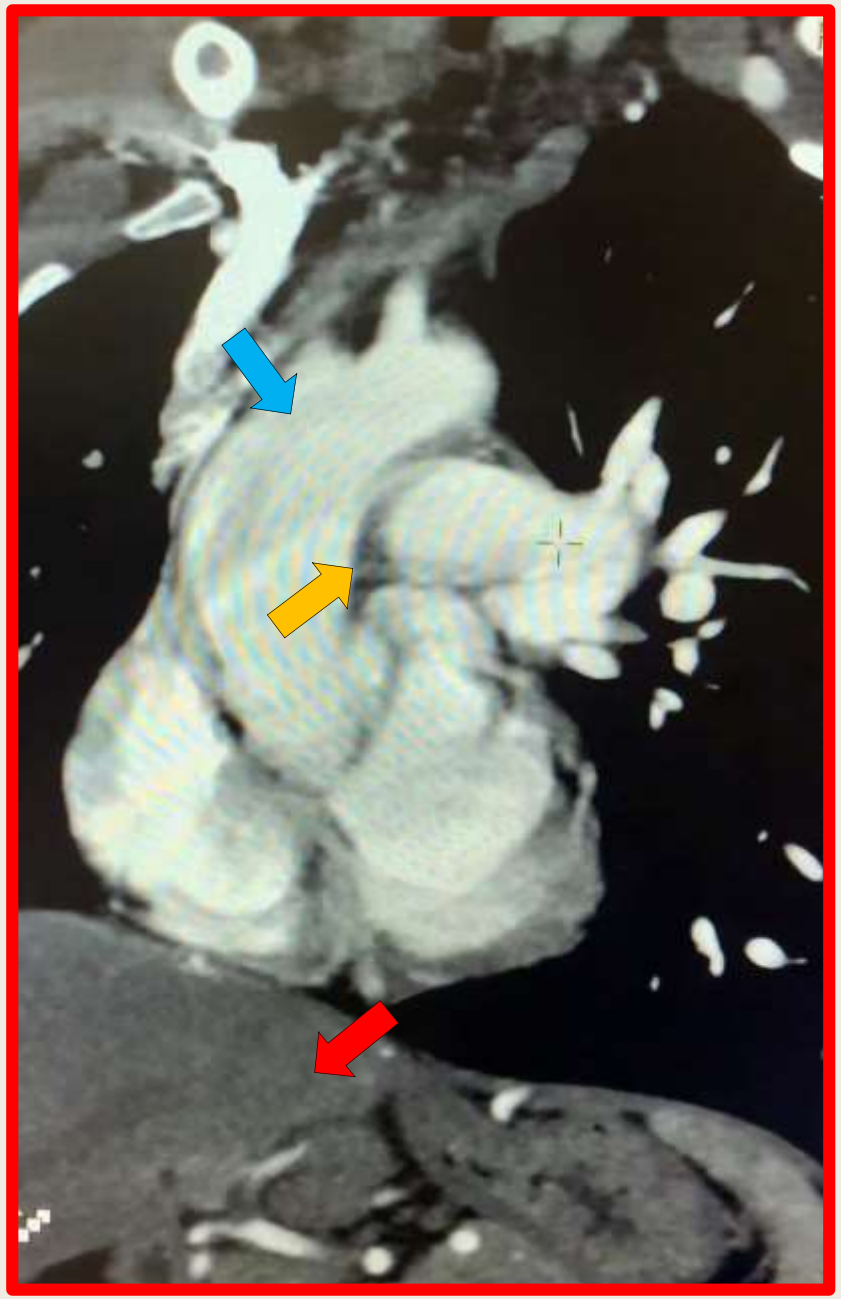
Absence of right pulmonary artery



Red – Absence of Right pulmonary artery

Blue – Main pulmonary artery

Yellow – Left pulmonary artery



Blue arrow – Hypertrophied bronchial arteries with increase in diameter of >3mm

Yellow arrow – Absence of right pulmonary artery

Red arrow – Hypertrophied inferior phrenic artery

INVESTIGATIONS

CT Aortogram

Right absence of pulmonary artery

Hypertrophic bronchial arteries, Right intercostal arteries, internal mammary arteries and right subclavian artery

ISOLATED UNILATERAL AGENESIS OF PULMONARY ARTERY

MANAGEMENT

Case was discussed with interventional radiologist and Cardio thoracic vascular surgeon

Patient was planned for ***Radical Embolization of right bronchial*** arteries, intercostal arteries and right internal mammary.

Pneumonectomy

(Recurrence of hemoptysis is common after SECA due to high pressure vascular system)

DISCUSSION

- Unilateral absence of pulmonary artery is a rare condition, with prevalence of **1:200000** young adults.
- It usually occurs with cardiac anomalies in 60% of the cases like Septal defects, Coarctation of aorta, Right aortic arch, Truncus arteriosus, Tetralogy of fallot.
- 40% of cases, they occur in isolation, hence called as isolated UAPA

DISCUSSION

Chest
Volume 122, Issue 4, October 2002, Pages 1471-1477

Selected Reports

Isolated Unilateral Absence of a Pulmonary Artery: A Case Report and Review of the Literature

A. Derk Jan Ten Harkel MD, PhD [a] ... Jaap Ottenkamp MD, PhD [b]

PRESENTATION

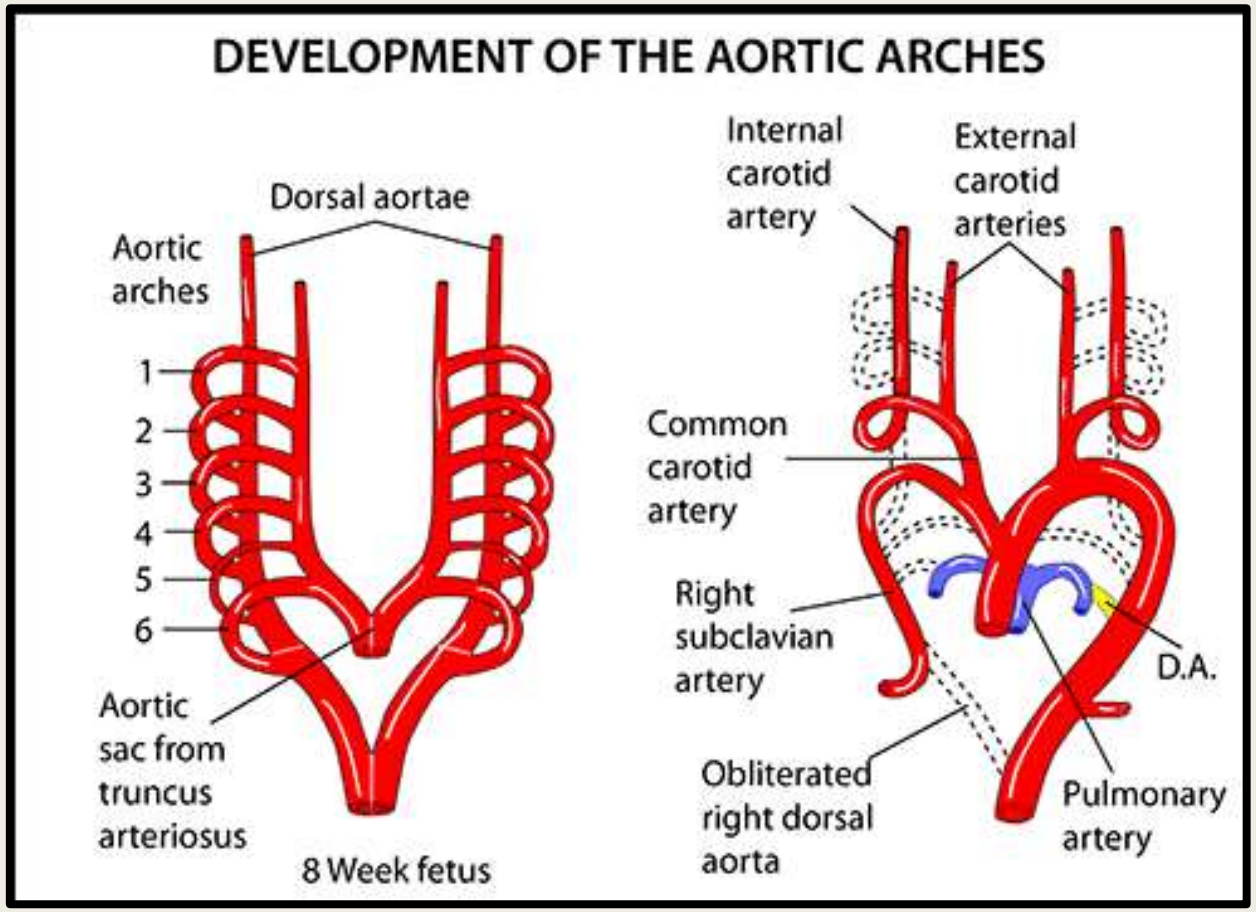
37%- Chest infections and pleural effusion

40%- Dyspnea on exertion

20%- Hemoptysis

Largest retrospective study with 108 cases from 1978-2000

DISCUSSION



Altered development of the **sixth aortic arch** (ductal origin to a pulmonary artery)

⇩

Distal intrapulmonary arteries of the affected artery will be intact

⇩

Supplied by **intercostal arteries**, **bronchial arteries** and **internal mammary arteries**, **subclavian** and even **coronary arteries.**

DISCUSSION

Diversion from Absent pulmonary artery → **Increased blood flow**

↓

Shear Stress on the endothelium

Endothelin release

Chronic vasoconstriction

↓

Remodeling and increased resistance

↓

Pulmonary hypertension

Collaterals formation

↓

Hypertrophied bronchial, intercostal and internal mammary arteries

↓

HEMOPTYSIS

MANAGEMENT

Angiography is the gold standard for diagnosis but is invasive and typically unnecessary unless it is being used as a preoperative test

Oral phosphodiesterase inhibitors and endothelin receptor antagonist - 50% had relief.

Original Article

Isolated unilateral absence of pulmonary artery in adulthood: a clinical analysis of 65 cases from a case series and systematic review

Ping Wang[1], Ling Yuan[2], Juhong Shi[1], Zuojun Xu[1]

- Embolization – 7 cases were done
- Pneumonectomy – 2 cases were done
- Selective embolization of collateral arteries (SECA) - 83% recurrence
- Pneumonectomy

  5 lost to follow up

  7 never had recurrence

ACKNOWLEDGMENT

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DPU

THANK YOU