

Hidden in plain sight: A rare abnormality detected on computed tomography pulmonary angiography

Ankita Rana¹, Pooja Abbey¹, Rama Anand¹, Ankit Gupta²

¹Department of Radiodiagnosis, Lady Hardinge Medical College, Smt. Sucheta Kriplani Hospital, New Delhi, India, ²Department of Medicine, Lady Hardinge Medical College, Smt. Sucheta Kriplani Hospital, New Delhi, India

Address for correspondence: Dr. Pooja Abbey, B-106, Doctors CGHS Apartments, Vasundhara Enclave, New Delhi - 110 096, India.
E-mail: pooja_abbey@yahoo.co.in

Submitted: 21-Oct-2020 **Accepted:** 24-Jun-2021 **Published:** 30-Aug-2021

CASE

A 31-year-old male presented to the emergency with acute-onset shortness of breath and palpitations for 5 days. He had had two episodes of hemoptysis in the last 2 days. There was no history of fever or cough. The patient gave a history of congenital heart disease but no history of past surgery or long-term medications for the same. On examination, blood pressure was 120/92 mm Hg, heart rate was 110 bpm, and SpO₂ was 88% on room air and 95% on O₂. He had central cyanosis and bilateral upper and lower limb clubbing. Blood workup revealed polycythemia with hemoglobin of 19.3 g/dL, hematocrit of 57.4%, mean corpuscular volume of 92.9 fL, mean corpuscular hemoglobin (MCH) of 31.2 pg, and MCH concentration of 33.6 g/dL. Platelet count was reduced (84,000 mcL). Chest radiograph [Figure 1] was reported as normal. Electrocardiography revealed sinus tachycardia and right-axis deviation. Computed tomography (CT) pulmonary angiography (CTPA) ruled out pulmonary thromboembolism [Figure 2]. CT lung window (not shown) showed a wedge-shaped peripheral area of ground glassing with some consolidation in the right lower lobe of lung, suggestive of possible viral (?COVID-19) pneumonia. However, the patient tested negative for COVID-19 by reverse transcription polymerase chain reaction.

QUESTION

What is the diagnosis?



Figure 1: Chest radiograph was reported as essentially normal

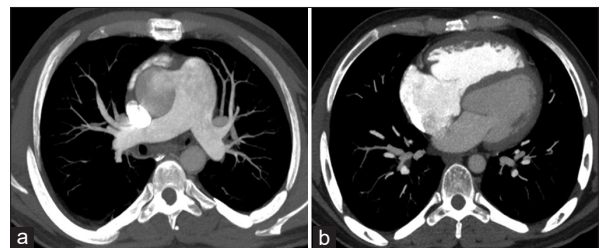


Figure 2: Computed tomography, axial Maximum intensity projection (MIP) images (a and b) showing no evidence of pulmonary thromboembolism.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Rana A, Abbey P, Anand R, Gupta A. Hidden in plain sight: A rare abnormality detected on computed tomography pulmonary angiography. Lung India 2021;38:491-3.

Access this article online	
Quick Response Code: 	Website: www.lungindia.com
	DOI: 10.4103/lungindia.lungindia_842_20

ANSWER

Aortopulmonary window (APW) or aortopulmonary (AP) septal defect (with pulmonary arterial hypertension and Eisenmenger syndrome).

In this patient, CTPA showed an abnormal communication between ascending aorta and main pulmonary artery [Figures 2 and 3]. There was right as well as left ventricular hypertrophy (more marked on right). The right atrium was dilated, which was also appreciated on the chest radiograph, on review. The main pulmonary artery was dilated (3.6 cm) as compared to aortic root (3.4 cm), suggestive of pulmonary hypertension. Pulmonary valve and aortic valve orifices appeared normal. Aortic arch and its branches appeared normal. No evidence of atrial or ventricular septal defects or patent ductus arteriosus was seen. Echocardiography was planned, but unfortunately, the patient rapidly deteriorated during his hospital stay and succumbed to a sudden cardiac arrest.

APW, also known as AP septal defect, is a very rare form of congenital heart disease, in which there is a deficiency in the septum between aorta and pulmonary artery, resulting in an abnormal communication between them.^[1] In approximately 50% of cases, it is associated with other disorders such as tetralogy of Fallot, interrupted aortic arch, coarctation of aorta, ventricular septal defect, or tricuspid atresia.^[2] It usually presents in the neonatal period, with symptoms of pulmonary overcirculation, such as feeding diaphoresis, tachycardia, tachypnea, and increased propensity toward respiratory viral infections. Uncommonly, presentation in adulthood has been described. The mean survival of uncorrected APW is 33 years.^[3] There are three types of APW, depending upon the location of the communication:

1. Type I – Defect in proximal part of AP septum, between aorta and main pulmonary artery. It is the most common type
2. Type II – Defect in distal part of AP septum extending into right pulmonary artery
3. Type III – Combination of I and II. It is the least common type of APW.

The entity must be differentiated from truncus arteriosus, where aorta and pulmonary artery do not differentiate during the fetal life, giving rise to a single large vessel arising from the ventricles with a single arterial or truncal valve. Thus, the presence of two separate semilunar valves is necessary to establish the diagnosis of APW.^[4]

APWs are generally diagnosed easily in the pediatric age group using echocardiography. In case when echo visualization is poor, angiography is used to demonstrate the abnormal communication. However, CT or CT angiography can also delineate the anatomical defect, as seen in our case. CT has the advantage of being noninvasive (as compared to angiography). Cautious

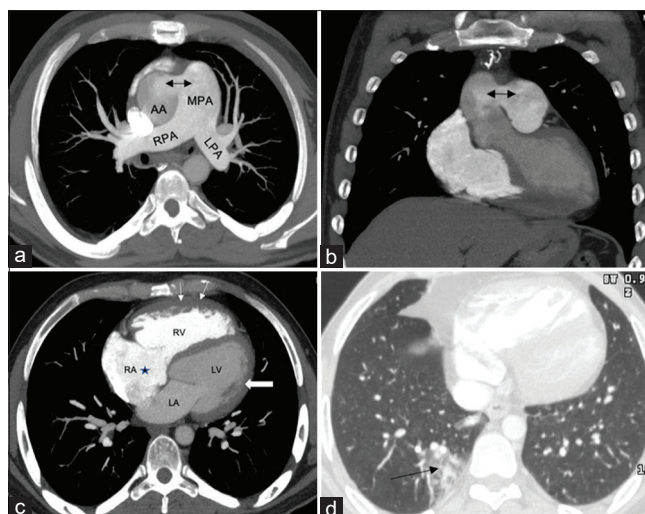


Figure 3: Computed tomography axial oblique and coronal reformatted maximum intensity projection images (a and b) revealing the defect (double-sided arrow) between the ascending aorta and main pulmonary artery, which is dilated. Right and left pulmonary arteries appear normal. Four-chamber view of the heart (c) showing right ventricular hypertrophy (small arrows) and mild left ventricular hypertrophy (block arrow) with enlarged right atrium (star). Axial image of the lung window (d) showing a wedge-shaped peripheral ground-glass opacity (arrow) in right lower lobe

interpretation should be done in the presence of cardiac motion artifacts, which may give rise to false positive results.

Early diagnosis of an APW is important, as surgery or cardiac catheterization device closure during childhood can avoid irreversible pulmonary hypertension, which invariably develops if left untreated. Development of Eisenmenger syndrome in APW is a contraindication to surgical closure of defect. In adult patients with APW, even if correction of defect is done before shunt reversal, the long-term outcomes remain unpredictable.^[5] In our case, the patient presented late with the development of cyanosis, clubbing, dyspnea, and hemoptysis, which are features of Eisenmenger syndrome. Hemoptysis is a common occurrence in patients with Eisenmenger syndrome and can lead to death in 11%–29% of cases.^[6] The presence of wedge-shaped ground-glass opacity in the right lower lobe in this patient could be due to aspiration of blood or due to pulmonary hemorrhage.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Sridhar PG, Kalyanpur A, Suresh PV, Sharma R, Maheshwari S, Hrudayalaya N. Helical CT evaluation of aortopulmonary window. *Indian J Radiol Imaging* 2006;16:847-9
2. Law MA, Mahajan K. Aortopulmonary septal defect. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2020.

3. Kose M, Ucar S, Emet S, Akpınar TS, Yalın K. A case of aortopulmonary window: Asymptomatic until the first pregnancy. *Case Rep Cardiol* 2015;2015:935253.
4. Goo HW, Park IS, Ko JK, Kim YH, Seo DM, Yun TJ, *et al.* CT of congenital heart disease: Normal anatomy and typical pathologic conditions. *Radiographics* 2003;23 Spec No:S147-65.
5. Myers PO, Lador F, Hachulla AL, Bouchardy J, Noble S, Licker M, *et al.* Unrestrictive aortopulmonary window: Extreme presentation as non-Eisenmenger in a 30-year-old patient. *Circulation* 2016;133:1907-10.
6. Jensen AS, Iversen K, Vejstrup NG, Sondergaard L. Images in cardiovascular medicine. Pulmonary artery thrombosis and hemoptysis in Eisenmenger syndrome. *Circulation* 2007;115:e632-4.