


# Descending aortopulmonary vein fistula: an unusual cause of recurrent hemoptysis in a young female: a case report

Parul Mrigpuri<sup>1\*</sup> , Balakrishnan Menon<sup>1</sup>, Sonam Spalgais<sup>1</sup>, Amitesh Gupta<sup>2</sup>

European Journal of Medical Case Reports

Volume 4(8):255-258

© EJMCr. <https://www.ejmcr.com/>

Reprints and permissions:

<https://www.discoverpublish.com/>

<https://doi.org/10.24911/ejmcr/>

173-1589523246

## ABSTRACT

**Background:** Hemoptysis is an alarming symptom. Its etiology may vary from mild infections to malignancies. Pulmonary infections are a common cause of hemoptysis in young adults. However, vascular malformations are also enlisted among the sources of hemoptysis.

**Case Presentation:** A 33-year-old female presented to the institute with hemoptysis. The etiology of hemoptysis was ascertained to be an anomalous vascular connection between the descending aorta and inferior pulmonary vein draining into the left atrium.

**Conclusion:** Descending aortopulmonary fistula (DAPVF) is a rare congenital vascular malformation representing a left-to-left shunt, and cardiac failure is a presenting feature as described in some reported cases in children. DAPVF may present as hemoptysis in adults.

**Keywords:** Hemoptysis, anomalous connections, descending aorta.

**Received:** 18 May 2020

**Accepted:** 24 June 2020

**Type of Article:** CASE REPORT

**Specialty:** Respiratory Medicine

**Funding:** None.

**Declaration of conflicting interests:** The authors declare that there is no conflict of interest regarding the publication of this case report.

**Correspondence to:** Parul Mrigpuri

\*Department of Pulmonary Medicine, Vallabhnbhai Patel Chest Institute, University of Delhi, New Delhi, India.

**Email:** parul57\_mrigpuri@yahoo.com

*Full list of author information is available at the end of the article.*

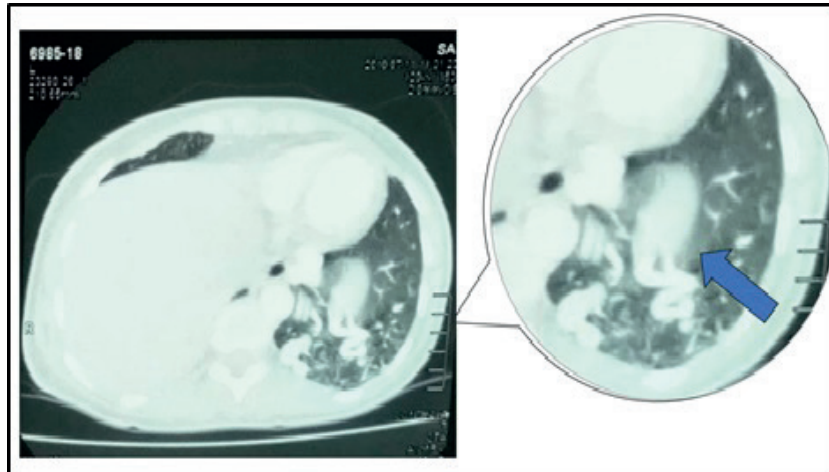
## Background

Hemoptysis is defined as the expectoration of blood from the tracheobronchial tree. It is an alarming symptom and matter of concern for both the patient and the treating physician. Hemoptysis can be caused by various mechanisms depending on the underlying etiology [1]. A detailed history and thorough examination followed by a focused investigation are required for diagnosis. We report a case of descending aortopulmonary fistula (DAPVF) as the cause of recurrent hemoptysis in a 33-year-old female. To the best of authors' knowledge, only very few cases of DAPVF as a cause of hemoptysis in adults have been reported in the literature.

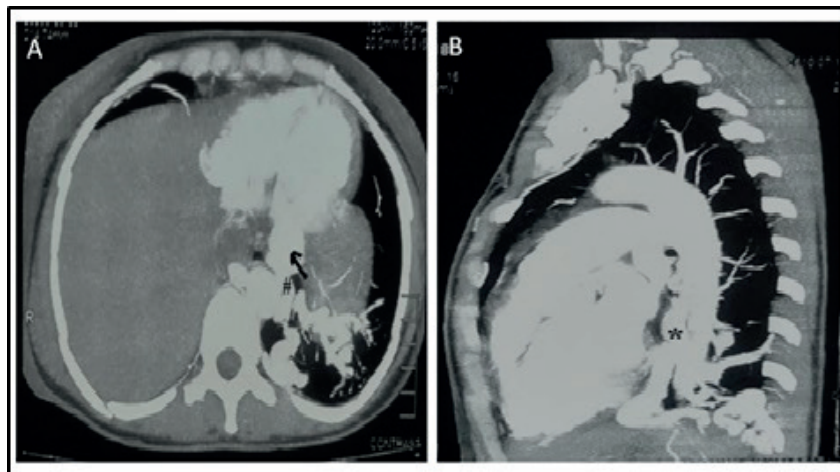
## Case Presentation

A 33-year-old female presented with the chief complaints of mild hemoptysis, 1-2 episodes/month for 2 years which increased to 3-5 episodes/month for the past 2 months. She was a non-smoker, and there was no history of thoracic trauma, anti-tubercular drug intake, hereditary disorder, and anticoagulant drug use. Menstruation had no effect on the episodes of hemoptysis. There was no other significant history, and on general examination, she had a pulse of 96/minutes, BP of 122/84, respiratory rate of 18/minutes, and SpO<sub>2</sub> of 98% at room air. Respiratory system examination was within normal limits. On cardiovascular system examination, a continuous murmur was heard over the fourth intercostal space along the left sternal border.

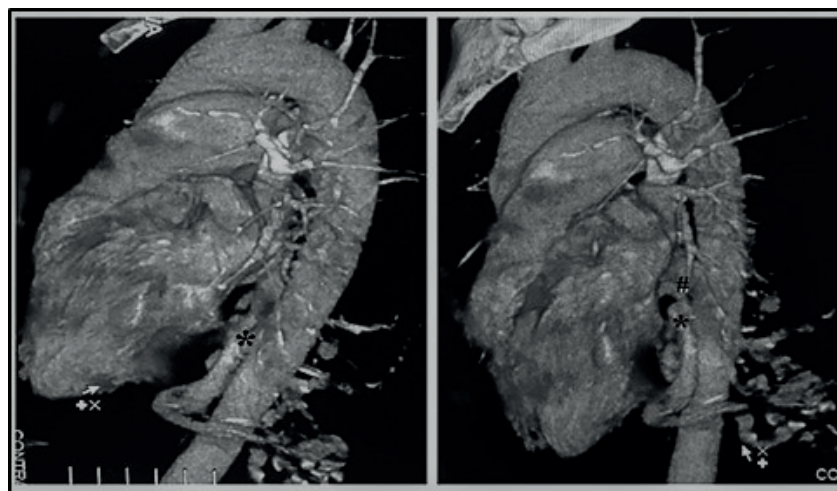
Laboratory tests revealed that normal white blood cell counts were 8,980 cells/μl. Kidney function tests, liver function tests, random blood sugar, and rest biochemistry were also normal. A chest X-ray and echocardiography was done and found to be within normal limits with echocardiography showing a normal left ventricular systolic function and an ejection fraction of 64%. A chest X-ray and echocardiography had no changes suggestive of pulmonary hypertension. The patient was planned for contrast-enhanced computed tomography (CECT) of the chest when she revealed that a previous CECT abdomen has been done at other centers for an unrelated complaint. On reviewing these CT films, some sections showed serpiginous structures consistent with enlarged vessels in the left lower lobe (Figure 1). Hence, CT pulmonary angiography was performed which showed an anomalous systemic artery arising from the descending aorta which was connected to the left inferior pulmonary vein representing the structure of DAPVF (Figure 2). An anomalous large vessel originated from the descending aorta and then descended below the diaphragm coursing along the left diaphragm before entering the left inferior lobe and finally to the left atrium via the inferior pulmonary vein. The three-dimensional reconstruction of the same is shown in Figure 3. The rest of the aorta was normal. The patient was referred to the department of cardiothoracic surgery for ligation of the fistula; however, the patient refused for any intervention.



**Figure 1.** Section of CECT abdomen showing serpiginous structures (arrow) consistent with enlarged vessels in left lower lobe of lung.



**Figure 2.** (A) CT angiography showing the anatomic structure and the course of the anomalous dilated vessel. An anomalous large vessel (#) originated from the descending aorta, then descended below the diaphragm coursing along the left diaphragm before entering the left inferior lobe and finally to left atrium via inferior pulmonary vein (black arrow). (B) Sagittal section showing the course of DAPVF. The anomalous systemic artery (\*) arises from the descending aorta.



**Figure 3.** Three-dimensional reconstruction of DAPVF. The anomalous systemic artery (\*) arises from the descending aorta and joins the left inferior pulmonary vein (#). The dilated inferior pulmonary vein drains into the left atrium.

## Discussion

Usually, the only communication between the systemic and pulmonary arterial systems is represented by the connection between the bronchial and pulmonary artery systems. The earliest descriptions of anomalous systemic to pulmonary communications dates to 1,777 [2]. These anomalous connections can be congenital or acquired [3]. Acquired causes mainly include aortic aneurysm, aortic dissection, infective endocarditis, syphilitic aortitis, Marfan's syndrome, and trauma. The origin of the fistulae can be from a normal systemic artery, e.g., bronchial or internal mammary artery. On the other hand, the origin can be from an aberrant systemic artery arising from the aorta. The adjoining lung parenchyma may be normal or sequestered in the latter case [4]. DAPVF is a rare congenital anomaly representing a left-to-left shunt in the absence of pulmonary involvement [5]. The left-to-left shunt is responsible for the presence of murmur. This shunt can cause the enlargement of the left atrium and left ventricle and, in due course, may lead to heart failure. Cardiac failure may be the presenting feature as described in some reported cases in children [5–8]. Very few cases of DAPVF as a cause of hemoptysis in adults have been reported in the literature [9,10]. In a review by Jariwala et al. [7] on anomalous systemic artery to pulmonary vein fistula out of the eight cases where the anomalous vessel originated from descending thoracic aorta, only one was reported in an adult. In another review by Yamanaka et al. [11] on cases of anomalous systemic arterial supply to normal basal segment of lower lobe, out of the 11 cases, where the anomalous vessel originated from descending thoracic aorta, only two had hemoptysis as one of the clinical manifestations. Hemoptysis is a consequence of persistently elevated venous pressure, resulting in segmental pulmonary arterial hypertension, which further leads to the rupture of proliferating and dilated vessels of the basal segments. In this case, although the patient had hemoptysis and murmur was present on cardiac examination, the etiology could not be established on preliminary investigations.

## Conclusion

DAPVF is a rare congenital vascular malformation representing a left-to-left shunt, and cardiac failure is a presenting feature as described in some reported cases in children. Hemoptysis could be the only symptom of DAPVF in adults. This case highlights the importance of imaging in the diagnosis of such rare anomalies.

### What is new?

DAPVF is a rare congenital vascular malformation representing a left-to-left shunt, and cardiac failure is a presenting feature as described in some reported cases in children. Very few cases of DAPVF as a cause of hemoptysis in adults have been reported in the literature.

## List of Abbreviations

DAPVF	Descending aortopulmonary fistula
CECT	Contrast-enhanced computed tomography

## Consent for publication

Written informed consent from the patient was obtained including publication of images.

## Ethical approval

Ethical approval is not required at the institution to publish an anonymous case report.

## Author details

Parul Mrigpuri<sup>1</sup>, Balakrishnan Menon<sup>1</sup>, Sonam Spalgais<sup>1</sup>, Amitesh Gupta<sup>2</sup>

1. Department of Pulmonary Medicine, Vallabhbhai Patel Chest Institute, University of Delhi, New Delhi, India

2. Department of Pulmonary Medicine, National Institute of Tuberculosis and Respiratory Diseases, New Delhi, India

## References

1. Corey R. Hemoptysis. In: Walker HK, Hall WD, Hurst JW, editors. *Clinical methods: the history, physical, and laboratory examinations* [Internet]. 3rd ed. Boston, MA: Butterworths; 1990:211-3. [cited 2020 May 4]. Available from: <http://www.ncbi.nlm.nih.gov/books/NBK360/>
2. Sade RM, Clouse M, Ellis FH. The spectrum of pulmonary sequestration. *Ann Thorac Surg.* 1974;18(6):644–58. [https://doi.org/10.1016/S0003-4975\(10\)64417-7](https://doi.org/10.1016/S0003-4975(10)64417-7)
3. Lawler LP, Fishman EK. Arteriovenous malformations and systemic lung supply: evaluation by multidetector CT and three-dimensional volume rendering. *Am J Roentgenol.* 2002;178(2):493–5. <https://doi.org/10.2214/ajr.178.2.1780493>
4. Wolf WJ, Casta A, Swischuk L. Aberrant systemic artery-pulmonary vein fistula: detection of an occult lesion by contrast echocardiography. *Am Heart J.* 1985;110(2):480–2. [https://doi.org/10.1016/0002-8703\(85\)90172-3](https://doi.org/10.1016/0002-8703(85)90172-3)
5. Jayan R, Prakashini K, Shetty RK, Chaitanya RK. A rare case of congenital anomalous/aberrant systemic artery to pulmonary venous fistula in a 4-month-old child with co-existent mitral valve prolapse and mitral regurgitation. *BMJ Case Rep* [Internet]. 2015 [cited 2020 May 5]; 2015. Available from: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4577702/> <https://doi.org/10.1136/bcr-2015-210865>
6. Kösecik M, Doğan N, Elmas B. Arteriovenous fistula between descending aorta and left inferior pulmonary vein: closure with vascular plugs. *Turk Kardiyol Dern Ars.* 2016;44(2):151–3. <https://doi.org/10.5543/tkda.2015.28235>
7. Jariwala P, Ramesh G, Sarat Chandra K. Congenital anomalous/aberrant systemic artery to pulmonary venous fistula: closure with vascular plugs & coil embolization. *Indian Heart J.* 2014;66(1):95–103. <https://doi.org/10.1016/j.ihj.2013.10.009>
8. Shebani SO, Khan MD, Tofeig MA. A congenital fistula between the descending aorta and the right pulmonary vein in a neonate presenting with heart failure. *Cardiol Young.* 2007;17(5):563–4. <https://doi.org/10.1017/S1047951107000583>
9. Hu Y, Zhong Q, Li Z, Chen J, Shen C, Song Y. An occult congenital fistula between the descending aorta and the left pulmonary vein in an adult presenting with recurrent

- episodes of hemoptysis. *Chest*. 2013;143(2):549–53. <https://doi.org/10.1378/chest.11-3229>
10. Wu T, Yu Y, Zhang Y, Deng Z. Congenital descending aorta-pulmonary vein fistula. *Am J Med Sci*. 2018;355(1):97–8. <https://doi.org/10.1016/j.amjms.2017.02.005>
  11. Yamanaka A, Hirai T, Fujimoto T, Hase M, Noguchi M, Konishi F. Anomalous systemic arterial supply to normal basal segments of the left lower lobe. *Ann Thorac Surg*. 1999;68(2):332-338. doi:10.1016/s0003-4975(99)00533-0

### Summary of the case

1	<b>Patient (gender, age)</b>	33 years/ Female
2	<b>Final diagnosis</b>	Descending aortopulmonary fistula
3	<b>Symptoms</b>	Hemoptysis
4	<b>Medications</b>	None
5	<b>Clinical procedure</b>	None
6	<b>Specialty</b>	Respiratory Medicine