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CASE REPORT

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Incidental diagnosis of a lingular pulmonary arteriovenous malformation in a 22-year-old female: a rare case report from Syria

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Abstract

Background Pulmonary arteriovenous malformations (PAVMs) are abnormal communications between pulmonary arteries and veins that bypass the normal pulmonary capillary bed. They are most commonly associated with hereditary hemorrhagic telangiectasia (HHT), while idiopathic cases are rare. PAVMs usually occur in the lower lobes and are more frequent in females, particularly during pregnancy. Although often clinically silent, they can present with dyspnea, hypoxemia, and cyanosis. This case is reported due to its rarity and as, to the best of our knowledge, the first documented surgically resected left lingular PAVM reported in Syria.

Case presentation A 22-year-old Syrian female, nulligravid, presented with unexplained dyspnea persisting for one year. Contrast-enhanced multidetector computed tomography (MDCT) of the chest revealed a PAVM in the left lingula, which was confirmed by histopathology. While endovascular embolization is the preferred treatment for PAVMs, the patient underwent complete surgical resection of the lingula. Postoperatively, vital signs and symptoms improved rapidly, and the procedure achieved a favorable outcome.

Conclusions This case highlights that PAVMs, although rare and often asymptomatic, can cause chronic dyspnea and may require surgical intervention when indicated. Reporting such cases contributes to clinical knowledge and awareness of management strategies in settings where endovascular treatment may not be feasible.

Keywords Case report, Idiopathic pulmonary arteriovenous malformation, Lingular pulmonary arteriovenous malformation, PAVM

Introduction

Pulmonary arteriovenous malformations (PAVMs) are defined as abnormal communication between a pulmonary artery and a pulmonary vein, avoiding the normal pulmonary capillary bed and resulting in a right-to-left shunt [1]. Patients with this condition typically present with dyspnea, hypoxemia, and cyanosis [2]. PAVMs are mostly congenital and considered rare, with a prevalence of 38 cases per 100,000 people, according to estimates, originating from a common plexus from which a pulmonary artery and a pulmonary vein form [1, 3]. To the best of our knowledge, this is the first reported case

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of idiopathic PAVM in Syria, discovered incidentally in a 22-year-old patient whose primary concern was dyspnea. The treatment of choice for this condition is endovascular embolization [3]. This case report aims to help local clinicians recognize and treat an uncommon condition.

Case Presentation

A 22-year-old nulligravid woman from Syria presented with progressive dyspnea for one year, corresponding to MRC grade 3 (as the patient described: "I can only walk for a few minutes before having to stop because I'm out of breath, even on level ground"). On examination, her initial vitals included a temperature of 36.5 °C, blood pressure of 110/60 mmHg, a heart rate of 110 per minute, and a respiration rate of 30 per minute. Her pulse oximetry revealed an oxygen saturation level of 85%. Her past medical history included tachycardia, palpitations, headache, central cyanosis with bluish discoloration of the lips, and digital clubbing of the upper extremities. The patient does not smoke or consume alcohol, and has no underlying illnesses, such as hypertension or diabetes. She did not experience hyperthermia, coughing, hemoptysis, or other discomfort. Cardiopulmonary auscultation of the right chest was unremarkable. Auscultation identified a continuous pulmonary vascular bruit on the left chest, which was most pronounced over the lingula of the left upper lobe. The rest of the physical examination was normal. A contrast-enhanced multidetector computed tomography (MDCT) of the chest, with 3D reconstruction revealed a pulmonary arteriovenous malformation (PAVM) (Fig. 1) between the branches of the left pulmonary artery supplying the left upper lobe, particularly the lingular region (lingula), and the corresponding lingular veins draining into the left pulmonary vein. The lesion demonstrated contrast enhancement, confirming the lesion's vascular nature. The 3D reconstruction further clarified the cluster-like vascular configuration typical of PAVMs (Fig. 2). Additionally, the MDCT showed concentric thickening of the left ventricular wall. The patient was admitted for surgery, subsequently put under general anesthesia, and positioned in the right lateral decubitus position, with a left posterolateral thoracotomy performed at the level of the fifth intercostal space. A large PAVM was identified, specifically involving the lingula. The lingula was isolated by ligating the feeding artery and draining vein. A 28-French chest tube was inserted, and the wound was closed in layers. Arterial blood gas analysis, performed preoperatively showed a pH of 7.31, $PaCO_2$ of 45 mmHg, HCO_3^- of 25 mEq/L, PaO_2 of 55 mmHg, and SaO_2 of 80%. The patient reached 100% oxygen saturation after surgery. She had a 95% oxygen saturation on room air on the second day and was conscious and at ease. For a week, she was prescribed carbocysteine as an expectorant, NSAIDs, and acetaminophen.

Discussion

Our case, identified in Syria, appeared as an abnormal vascular connection between a pulmonary artery and a pulmonary vein, lacking a normal intervening capillary bed, leading to an intrapulmonary right-to-left shunt typical of a pulmonary arteriovenous malformation (PAVM) [2]. It mainly affects women, with a male-to-female ratio of around 1:1.5 to 1.8, and pregnancy is recognized as a predisposing factor that may promote growth [2, 3]. Most PAVMs are hereditary, with hereditary hemorrhagic telangiectasia (HHT), which is an autosomal dominant vascular disorder, being a primary association with the condition [2]. Approximately 70% of PAVMs occur as part of HHT syndrome. The diagnosis of HHT can be evaluated using the Curaçao Criteria, which rely on epistaxis, telangiectasia, visceral lesions, and family history, none of which were present in our patient [3]. Confirmatory molecular testing targeting ENG, ACVRL1, and SMAD4, which are the most commonly implicated genes in HHT, constitutes the primary genetic diagnostic approach because mutations in these three specific genes account for the vast majority of cases where a genetic cause is identified [2]. However, such testing was not performed in this patient. Rarely, PAVMs can be idiopathic or associated with other disorders [4], which was suspected in this case. In around 60–95% of PAVMs, the lower lobes are the most commonly affected sites, whereas idiopathic PAVMs, which are usually simple and fistulous, show no predominance for the lower lobes. The former is attributed to the fact that pulmonary blood flow and pressure increase in the lower lobes. Unlike idiopathic cases, PAVMs associated with HHT are usually multiple [3, 4]. While 50% of patients with PAVMs are typically asymptomatic, they can also present with dyspnea, cough, chest pain, palpitations, intrapulmonary hemorrhage, and neurological symptoms or deficits. A bruit or thrill, clubbing, telangiectasia, polycythemia, cyanosis, or a systolic murmur may also be discovered during a physical examination. Other, more fatal complications include massive hemoptysis and spontaneous hemothorax, besides stroke and intracranial abscess, which are the result of paradoxical systemic embolization. The latter can be explained by the absence of the pulmonary capillary bed, which normally acts as a filter for blood coming from the pulmonary arteries [3]. Early recognition and timely intervention are essential to prevent these potentially life-threatening complications. PAVM typically becomes more apparent in the second and third decades of age [2]. Several common presenting symptoms were present in our patient, a 22-year-old nulligravid woman with an upper-lobe PAVM outside the usual lower-lobe predominance of

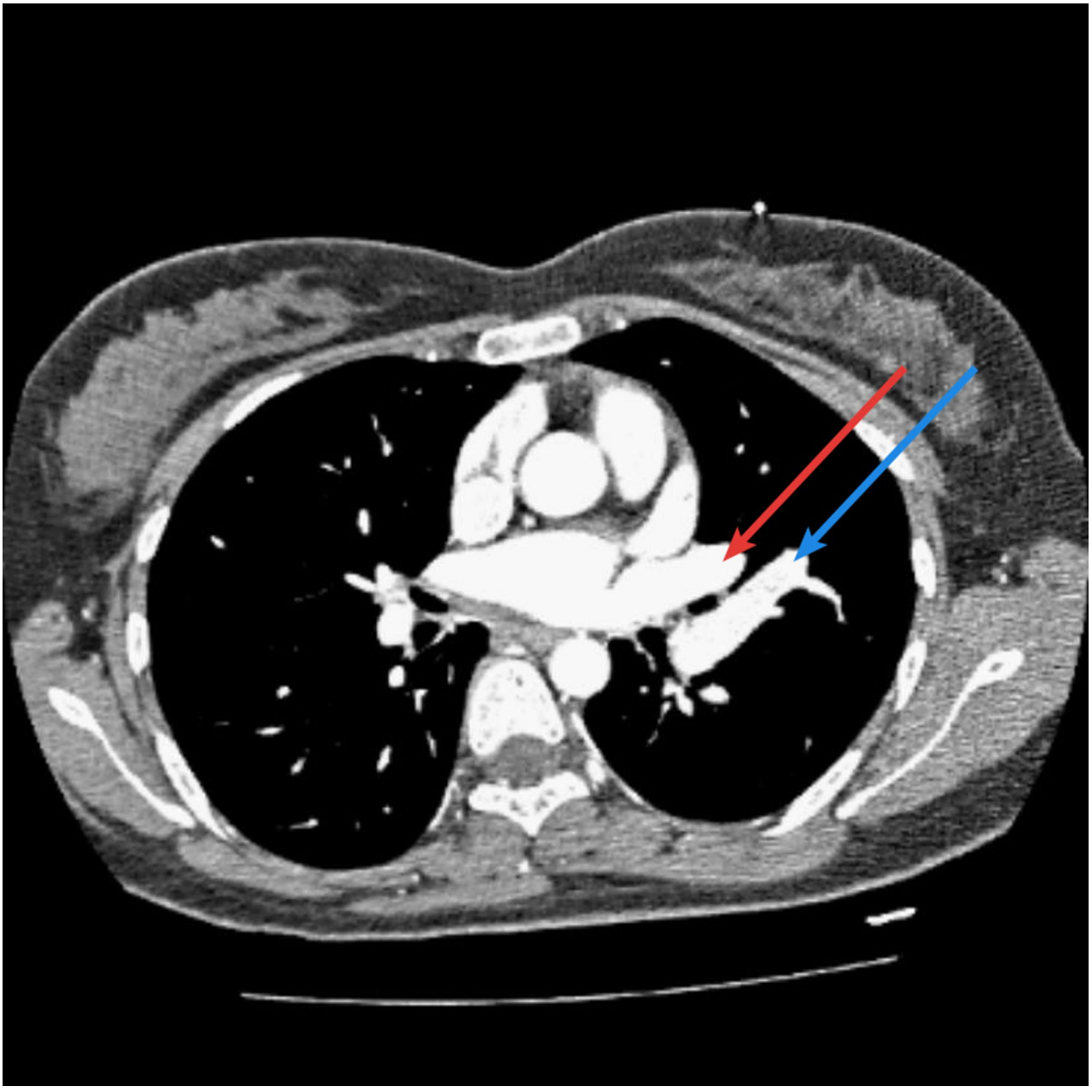

Fig. 1 Contrast-enhanced MDCT showing a lingular pulmonary arteriovenous malformation. Legend: Axial contrast-enhanced multidetector computed tomography (MDCT) image demonstrating a lingular pulmonary arteriovenous malformation, with the feeding artery (blue arrow) and the draining vein (red arrow) clearly visualized. All patient identifiers and hospital information have been removed from the image

HHT-associated PAVMs. These included progressive dyspnea, palpitations, central cyanosis, digital clubbing, and an audible pulmonary vascular bruit, while more serious complications were absent. However, due to her persistent pulmonary symptoms and the need to check for underlying lesions, imaging was sought. The patient stated that she had previously undergone chest X-rays at external facilities, but was unable to recall the results, and the records were not available. In order to provide a quick and thorough assessment of her pulmonary symptoms, the clinical team proceeded directly to contrast-enhanced chest MDCT rather than repeating a chest X-ray at our facility. Although the gold standard for diagnosis is computed tomography pulmonary angiography [3], the guiding imaging method conducted by the doctors relied mainly on contrast-enhanced multidetector computed tomography (MDCT) of the chest, which revealed the lingular location of the PAVM. The rest of the lungs appeared clear, with no other lesions observed. This

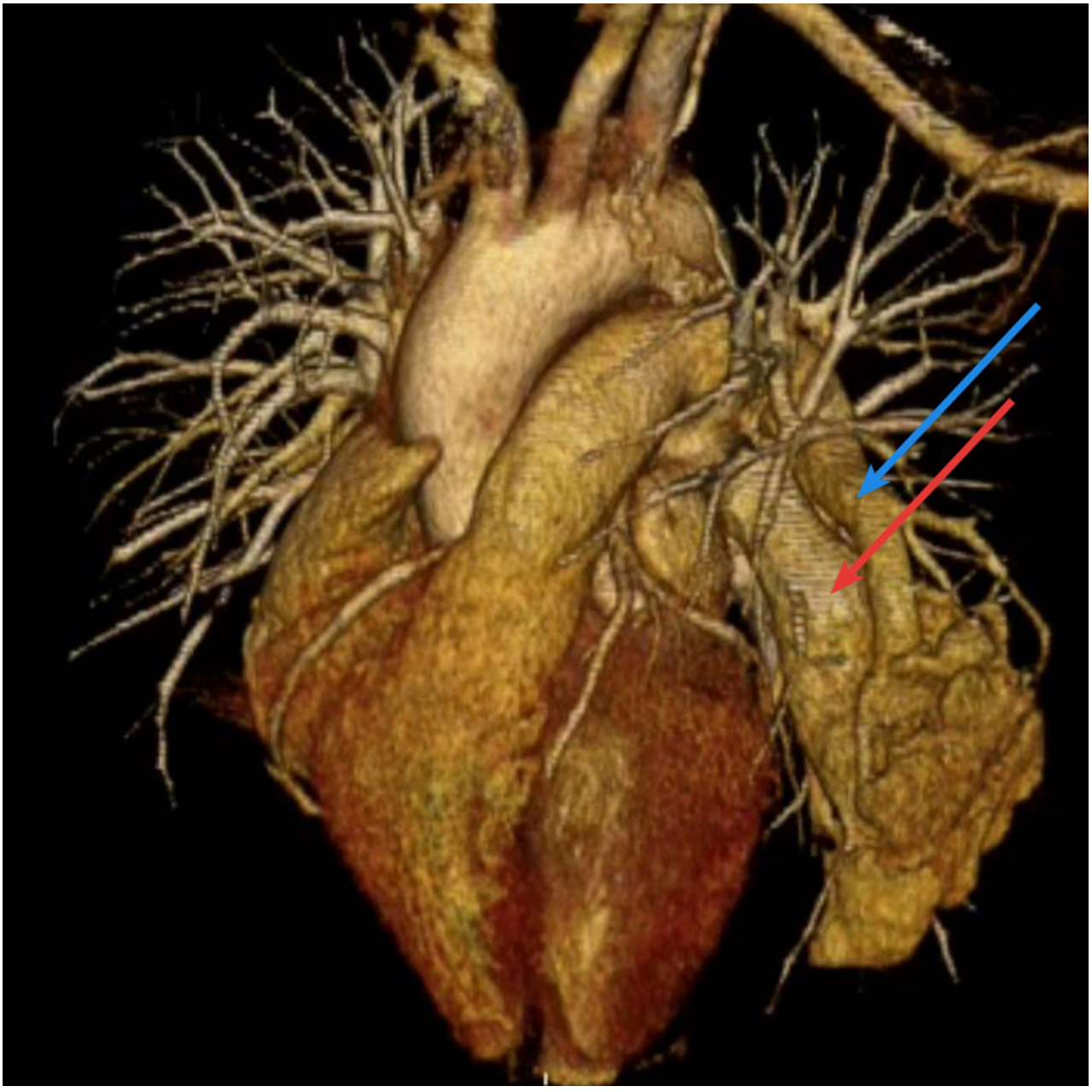

Fig. 2 Three-dimensional reconstruction of the lingular pulmonary arteriovenous malformation. Legend: Three-dimensional reconstruction illustrating the lingular pulmonary arteriovenous malformation. The feeding artery (blue arrow) and the draining vein (red arrow) are shown in their spatial relationship to the surrounding pulmonary vasculature. All patient identifiers and hospital information have been removed from the image

confirms the PAVM was solitary, consistent with non-HHT cases [4]. No echocardiography was performed, and an echocardiogram with a bubble study, which can identify right-to-left shunting, was not done. Differential diagnosis originally included pulmonary malignancy. Instead of a solid neoplastic mass, contrast-enhanced MDCT showed early and uniform enhancement of the lesion with contrast material in a pattern consistent with vascular structures and in continuity with adjacent vessels. These imaging features led the treating physicians to favor a diagnosis of pulmonary arteriovenous malformation over malignancy. Surgical resection was decided based on imaging findings and clinical judgment. To definitively exclude malignancy, histopathology of the resected lingular segment was performed. In line with previous research that describes PAVM as having no malignant potential [2, 4], macroscopic findings revealed a well-circumscribed, spongy, reddish area measuring 4 cm, composed of dilated thin-walled vascular channels with

no distinct mass or necrosis; microscopic description showed clusters of markedly dilated vascular channels of variable caliber lined by a single layer of flattened endothelial cells without atypia or mitotic activity, with direct communication between arteries and veins and absent intervening capillary bed; surrounding alveoli demonstrated mild congestion and focal hemosiderin-laden macrophages, with no indication of inflammation, fibrosis, granuloma, or neoplasia, effectively excluding malignancy in this case. It is widely acknowledged that endovascular embolization or surgery are the two main treatments for PAVM, with many reports suggesting that embolization is generally preferred over surgical resection. Video-assisted thoracoscopic surgery (VATS) has also increasingly been adopted as an effective and less invasive surgical approach for selected cases [2–4]. The final course of treatment in this case was open surgical resection of the entire lingula due to the inability to use less invasive techniques and the need to prevent paradoxical embolism, especially given the size and location of the PAVM and the arrangement of its feeding vessels. A chest X-ray taken about ten days after the surgery showed expected postoperative changes in the left lung, with no evidence of acute complications. At one-month and three-month telephone follow-ups, the patient reported that her symptoms had completely disappeared. She was instructed to attend an in-person follow-up, and if clinically necessary, additional imaging, including CT, would be carried out. In patients with PAVMs, including our case, it is recommended to follow specific lifestyle and procedural precautions [5, 6]. These include prophylaxis with antibiotics before procedures that could cause bacteremia, particularly dental work, for both treated and untreated patients [5]. Careful measures to avoid intravenous air, including the use of air filters on IV lines, are advised to prevent iatrogenic embolism, which can be especially dangerous in untreated patients due to the lack of a pulmonary capillary filter between the arterial and venous beds, allowing venous blood and any emboli it contains to bypass the lungs and enter the systemic circulation. Activities such as scuba diving carry theoretical risks and should be avoided [5, 6]. Adherence to these precautions is essential to reduce serious complications, including stroke and brain abscess [5].

Strengths and limitations

One of the report's primary strengths is the documentation of a single idiopathic PAVM involving the lingula, an uncommon upper-lobe site that deviates from the typical PAVMs with HHT-association and lower-lobe distribution. As far as we are aware, this is the first case of its kind to be reported from Syria, so it serves as a benchmark for the local medical literature. The case also shows that traditional techniques, such as open surgical resection, are still a feasible management approach for reducing symptomatic distress in situations where endovascular interventions cannot be used.

However, this case report has several limitations. First, previous chest X-rays were not available, limiting the ability to establish a radiographic baseline for this patient. Second, it was difficult to rule out a genetic cause because confirmatory genetic testing for hereditary hemorrhagic telangiectasia (ENG, ACVRL1, and SMAD4) was not available locally. Third, open surgical resection was pursued as endovascular embolization and video-assisted thoracoscopic surgery (VATS) were not viable options due to a lack of trained personnel, unavailable equipment, and the patient's financial limitations. Fourth, transthoracic echocardiography with a bubble study, which can detect right-to-left shunting, was not performed because this technique is not routinely practiced at our facility. Fifth, sagittal and coronal CT reconstructions were not available due to technical limitations in retrieving the original imaging data, although all views were reviewed by the treating physicians during clinical assessment. Lastly, there was little follow-up: no in-person evaluations or advanced imaging were performed beyond the chest X-ray at ten days postoperatively, although telephone follow-ups at one and three months showed no recurrence of symptoms.

Conclusion

This case illustrates that pulmonary arteriovenous malformation (PAVM) should be taken into consideration when a patient, particularly a female, presents with unexplained symptoms of dyspnea, hypoxemia, cyanosis, and clubbing. Diagnosing the condition early and treating it appropriately, either with embolization or surgery, can reduce the risk of serious complications.

Abbreviations

HHT	Hereditary hemorrhagic telangiectasia
MDCT	Multidetector computed tomography
PAVM	Pulmonary arteriovenous malformation

Authors' contributions

M.A. and H.A. were responsible for drafting the manuscript, conducting the literature review, and preparing all sections of the paper. B.I., Y.A., and B.D. contributed to the collection of clinical data. S.I. revised the manuscript for clarity and academic quality. B.D. additionally ensured the accuracy of clinical and surgical details. All authors read and approved the final manuscript.

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Data availability

The data supporting the findings of this study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
Ethics approval was not required for this case report according to the guidance provided by the treating institution. Written informed consent to participate was obtained from the patient.

Consent for publication
Written informed consent for publication of the case details and accompanying images was obtained from the patient.

Competing interests
The authors declare no competing interests.

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References

1. Raptis DA, Short R, Robb C, Marlow J, Naeem M, McWilliams S, et al. CT Appearance of Pulmonary Arteriovenous Malformations and Mimics. Radiographics. 2022;42(1):56–8.
2. Cartin-Ceba R, Swanson KL, Krowka MJ. Pulmonary arteriovenous malformations. Chest. 2013;144(3):1033–44.
3. TellapuriS, Park HS, Kalva SP. Pulmonary arteriovenous malformations. Int J Cardiovasc Imaging. 2019;35(8):1421–28. https://doi.org/10.1007/s10554-018-1479-x.
4. Reichert M, Kerber S, Alkoudmani I, Bodner J. Management of a solitary pulmonary arteriovenous malformation by video-assisted thoracoscopic surgery and anatomic lingula resection: video and review. Surg Endosc. 2016;30(4):1667–9.
5. GargN, Khunger M, Gupta A, Kumar N. Optimal management of hereditary hemorrhagic telangiectasia. J Blood Med. 2014;5:191–206. https://doi.org/10.2147/JBM.S45295.
6. Meek ME, Meek JC, Beheshti MV. Management of pulmonary arteriovenous malformations. Semin Intervent Radiol. 2011;28(1):24–31.

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