

Bilateral Pulmonary Arteriovenous Malformation in Female Patient Based on Computed Tomography Imaging: A Rare Case Report

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ABSTRACT

Background: Pulmonary arteriovenous malformations (PAVMs) is an extremely rare vascular anomaly due to a direct connection between the pulmonary artery and pulmonary vein, potentially causing irreversible damages to the systemic circulation. This condition requires a prompt diagnostic approach to ensure early diagnosis and treatment.

Case Report: A 53-year-old Indonesian female was referred to our department with unexplained dyspnoea and cyanosis. Physical examination revealed low oxygen saturation and remarkable finding on pulmonary auscultation. Further investigation revealed the findings suggesting PAVM based on contrast-enhanced chest computed tomography (CT) scan, with multiple nidus on bilateral lungs with feeder arteries from the pulmonary artery and draining veins in the pulmonary vein. Hence, this case emphasizes the rare finding of a female PAVMs patient with bilateral PAVMs.

Conclusion: CT scan is a reliable and effective imaging approach to establish the diagnosis of PAVMs. This modality should be first considered to visualize PAVMs lesions, particularly in adult patients with unexplained dyspnoea and cyanosis.

Keywords: arteriovenous malformations, CT scan, rare case, embolization

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BACKGROUND

Pulmonary arteriovenous malformations (PAVMs) are a rare formation of an abnormal vascular connection between the pulmonary artery and pulmonary vein through a thin-walled aneurysmal sac. It is estimated to occur in 2-3 per 100.000 people, with a higher incidence among males than females. The clinical consequences of PALMs are related to the entry of deoxygenated blood into the systemic

circulation (right-to-left shunt), resulting in hypoxemia, fatigue, dyspnea, and cyanosis (Circo et al., 2014).

Classically, PAVMs manifest as a simple malformation consisting of a single feeding and draining vessel, or in more infrequent cases, as complex malformations consisting of multiple feeding vessels with a predominant area in the bilateral lower lobes of the lung (Othpost et al., 2019). The diagnosis modality can be readily

established by imaging such as computed tomography (CT) scan providing characteristic finding. Unfortunately, previous studies explained that 80% of patients with PAVMs are related with Rendu Osler Weber Syndrome, also known as hemorrhagic hereditary telangiectasia. This condition may contribute to being misdiagnosed (Pollack et al., 2006).

Our case presented a 53-year-old female with chief complain unexplained dyspnoea and cyanosis, which subsequently established as bilateral PAVMs. This case report was rarely finding and usually misdiagnosed by imaging. Herein, we emphasized the clinical findings based on CT scan, which is an excellent approach in diagnosing PAVMs.

CASE PRESENTATION

A 53-year-old Indonesian female was referred to our department with chief complain unexplained dyspnoea on moderate exertion for one months ago with central and peripheral cyanosis, she had no relevant medical and family history. She was housewife and denied previous smoking and alcohol consumption. Physical examinations revealed the patient was in a hypoxic state (oxygen saturation 88%). Pulmonary auscultation examination was unimpressive, and blood laboratory tests showed mild anemia. The patient was admitted for hospitalization with respiratory distress and decreased level of consciousness.

A chest plain radiography suggested suspicion of PAVMs. Therefore, we decided to perform contrast-enhanced computed tomography (CT) scan on the thorax region. The result showed abnormal vasculature at the right lower lobe, with multiple nidus on bilateral lungs with feeder arteries from the pulmonary artery and draining veins in the pulmonary vein (see Figure 1 and 2).

RESULTS

Further investigation revealed the findings suggesting PAVMs based on contrast-enhanced chest computed tomography (CT) scan, the result showed multiple nidus on bilateral lungs with feeder arteries from the pulmonary artery and draining veins in the pulmonary vein. On the right lung showing nidus 2.71 x 2.03 x 1.40 cm, feeder artery of 0.55 cm, and draining vein of 0.48 cm), and lower lobe of the left lung revealed nidus 1.91 x 1.78 x 1.43, feeder artery of 0.36 cm, draining vein of 0.30 cm). Abnormal vasculature at the right lower lobe. On the left middle lung with coronal section revealed 2.48 x 1.46 x 0.97, feeder artery of 0.50 cm, draining vein of 0.28 cm. (see Figure 1 and 2). Hence, this case emphasizes the rare finding of a female PAVMs patient with bilateral PAVMs.

These findings were confirmed of the diagnosis of bilateral PAVMs. Subsequently, the patient was admitted for further treatment by performing embolization and closure of PAVMs. Unfortunately, the patient refused to complete the procedure. The patient has died after a few hours of refusing the procedure.

DISCUSSION

Pulmonary arteriovenous malformation (PAVMs) is a rare vascular anomaly due to inappropriate connection between two main vessels of pulmonary circulation, with reported incidence ranging from 2-3 cases per 100,000 population. Congenital anomalies account for more than 80% of patients, and the remainder is acquired, mostly due to chest trauma (McDonald et al., 2015). The estimated incidence showed that PAVMs were more common in women, with a male-to-female ratio of 1.5:1 (Nakayama et al., 2012).

Mutations in the endoglin and activin receptor-like kinase 1 (ACVRL1 or ALK1)

become a specific genetical cause of PAVMs

(Wehner et al., 2006).



Figure 1. A chest computed tomography scan of (A) Right lung, showing the approximate size of nidus 2.71 x 2.03 x 1.40 cm, feeder artery of 0.55 cm, and draining vein of 0.48 cm; (B) Lower lobe of the left lung, showing the approximate size of nidus 1.91 x 1.78 x 1.43, feeder artery of 0.36 cm, draining vein of 0.30 cm.

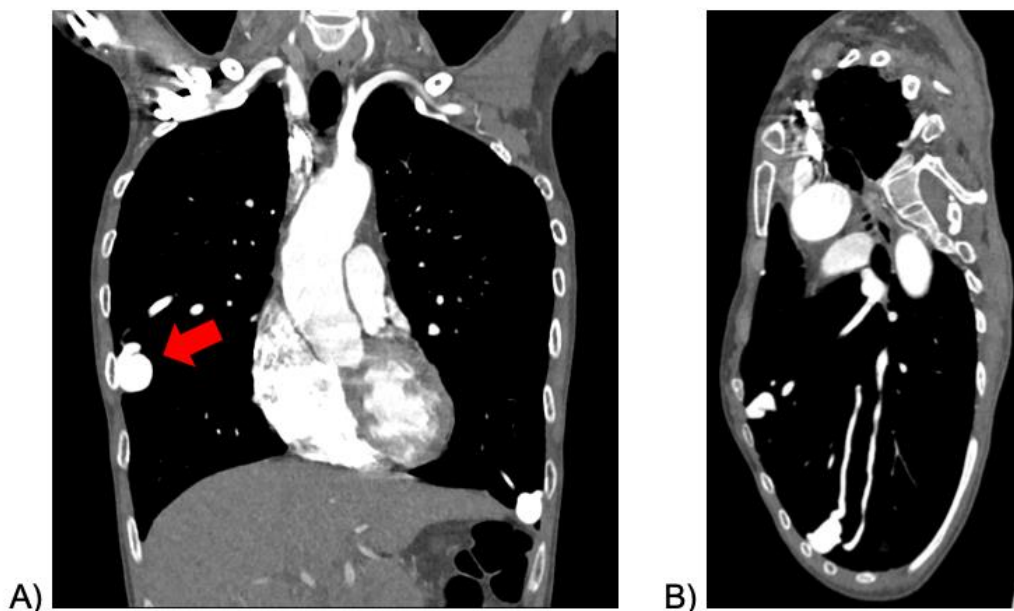


Figure 2. A chest computed tomography scan in (A) Coronal section, which demonstrates the abnormal vasculature at the right lower lobe (arrow); (B) On the left middle lung, showing the approximate size of nidus 2.48 x 1.46 x 0.97, feeder artery of 0.50 cm, draining vein of 0.28 cm.

The clinical manifestations of PAVMs often present with dyspnoea on exertion, leading to hypoxemia with the signs of cyanotic appearance and nail clubbing (Mohammed et al., 2018). Nevertheless, PAVMs patients are typically asymptomatic until the four-decade of life (Kuhajda et al., 2015).

Our patient demonstrates classic imaging findings of PAVMs based on contrast-enhanced CT scan, with multiple nidus on bilateral lungs connected to serpiginous arteries feeders and draining veins. Chest computed tomography imaging is a standard diagnostic procedure, especially to assist the evaluation of feeding and draining vessels. The diagnosis is established based on the finding of an aneurysm sac (nidus), which appears as a well-defined, homogeneous round or serpiginous nodule connected to the blood vessels. In complex PAVMs cases, two or more feeding arteries can be found. A contrast-enhanced CT scan provides a more accurate anatomical picture through enhancement of the vessels. This method is beneficial and is generally performed prior to therapeutic embolization. Pulmonary arteriography is another method in diagnosing complex PAVMs cases; however, it has been reported less sensitive compared to CT-scan (Karavdic et al., 2018).

The differential diagnosis of PAVMs includes the isolated meandering pulmonary vein (MPV) and scimitar syndrome. Isolated MPV presents with asymptomatic features, with the absence of systemic arterial supply and associated anomaly. Meanwhile, the scimitar syndrome often manifests with dyspnea and associated recurrent pulmonary infection due to congenital hypoplastic right pulmonary artery and right lung (Ko et al., 2021).

An abnormal connection between the pulmonary artery and vein causes impair-

ment of the systemic circulation due to the right-to-left shunting. The lungs act as a filter preventing the passage of blood clots into the systemic circulation. Direct connection from the pulmonary artery to systemic circulation increases the risk of paradoxical embolism, leading to stroke and brain abscess due to bacterial invasion. Furthermore, the dilated, thin-walled aneurysmal sac may also lead to hemorrhage, which accounts for the manifestation of hemoptysis and hemothorax in approximately 30% of PAVMs cases (Silva et al., 2016).

Although rare, the presence of PAVMs may lead to fatal complications. Therefore, a multidisciplinary approach for managing the disease and its complications must be performed carefully and comprehensively. Catheter embolization and surgery are two main modalities in managing PAVMs. Simple PAVM with nodule less than 2 cm in diameter should be treated percutaneously or surgically, regardless of the presence or absence of symptoms (Woodward et al., 2013). Narsinh et al. (2013) stated that PAVMs with arterial feeder diameter of less than 3 mm should also receive adequate treatment.

Transcatheter coil embolization is an ideal percutaneous treatment of choice that has taken over most of the need of surgery among PAVMs patients because it offers a less invasive approach than the surgical resection (Hsu et al., 2015). This technique improves the oxygen saturation and decreases in right-to left-shunting by using coaxial or triaxial catheters through the site of the arterial feeder artery to push fibered coils, which are subsequently placed into the PAVM site (Verhelst et al., 2018; White et al., 2007).

Studies showed that this technique provides a good outcome, with only 3% recurrence rate (White et al., 2007).

Embolization has a high success rate (95-100%) to establish the occlusion of the arterial feeder (Andersen et al., 2010). A significant decrease of the pulmonary shunt was reported based on echocardiography, followed by the increased partial pressure of oxygen in arterial blood flow. Additionally, only 10% of cases occur as compromising complications from the embolization method.

The post-embolization follow-up for the patients with PAVMs is performed to re-evaluate the lesion based on CT scan imaging. The current recommendation suggests the follow-up should be done in six to 12 months after embolization, followed by every three years. In PAVMs patients with an indicative result on CT imaging or positive contrast echocardiographic examination, the follow-up should be done every one to five years (Shovlin, 2014). In case of therapy failure, surgical treatment, such as local excision, segmental resection, lobectomy or ligation, and pneumonectomy, may be considered to perform (Sharifah et al., 2009).

Bilateral PAVMs in female patients is a relatively uncommon finding in our radiology department. A comprehensive history taking and a high degree of suspicion are immensely required to establish a proper confirmed diagnosis. CT scan examination is considered the best option to visualize PAVMs clearly, particularly in patients with unexplained dyspnoea on moderate exertion with central and peripheral cyanosis and failure on initial conventional therapy.

AUTHOR CONTRIBUTION

Each author individually made significant contributions to the development of this manuscript.

CONFLICT OF INTEREST

The authors declare that the study was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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