Proximal Pulmonary Artery Aneurysm Secondary to Suspected Pulmonary Hypertension Treated with Conservative Therapy in Limited Resource Setting

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Patient: Male, 56-year-old
Final Diagnosis: Proximal pulmonary artery aneurysm
Symptoms: Atypical bilateral chest pain • shortness of breath
Medication: —
Clinical Procedure: —
Specialty: Cardiology

Objective: Rare disease
Background: Pulmonary artery aneurysm (PAA) is a rare disease in cardiovascular system. This disease is difficult to diagnose and less often considered due to its non-specific clinical manifestations. Until now there are no clear guidelines about its optimal management because of the small number of reported cases.

Case Report: We report a 56-year-old male with chief complain of atypical bilateral chest pain and shortness of breath. Initial electrocardiogram (ECG) and laboratory evaluation showed no sign of ischemic heart disease. After the patient was stabilized, he was evaluated using chest x-ray, transthoracic echocardiography (TTE), and multi slice computed tomography (MSCT). The patient was then diagnosed with PAA secondary to suspicion of pulmonary hypertension (PH) with chronic obstructive pulmonary disease and heart failure. Conservative treatment was chosen because of the limited resources for surgery and patient’s refusal to be referred. The treatment aims to lower the pulmonary artery pressure while monitoring the aneurysm. His 6-month follow-up evaluation showed an improvement in pulmonary artery pressure and persistent of the PAA without any increase of the diameter.

Conclusions: PAA is a rare disease that is difficult to diagnose because of its non-specific nature. Persistent atypical chest pain can be an early symptom of PAA, thus clinicians should be aware in a high-risk patient suffered persistent chest pain, despite normal ECG and laboratory findings. TTE and MSCT evaluation are reliable for diagnosing PH and PAA. With conservative treatment and routine follow-up, patient with PAA secondary to PH could be managed well.

MeSH Keywords: Aneurysm • Hypertension, Pulmonary • Indonesia • Pulmonary Artery

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Background

Pulmonary artery aneurysm (PAA) is a rare disease in the cardiovascular system. It is described as a dilatation of the pulmonary artery. Earlier autopsy study of 109,571 necropsies revealed that the incidence of PAA is only 1 in 13,696 necropsies [1]. To this date, there is no accurate definition for the minimal size of the aneurysm to be diagnosed as PAA. A recent review article mentioned that there have been several proposed cutoff measurements for PAA, i.e., dilatation of the PA beyond its maximal normal caliber; PA with a diameter exceeding 4 cm; enlargement of main PA diameter exceeding 2.9 cm based on the computed tomography (CT) [2]. Recognition of PAA is important because of high morbidity and mortality rates of rupture [3]. Unfortunately, this disease is difficult to diagnose and less often considered because the clinical manifestation is non-specific and can be seen in many other conditions, or even may remain asymptomatic [4]. Moreover, there is no clear guideline about its optimal management because there are only few reported cases. In this report, we present a case of patient with PAA secondary to pulmonary hypertension (PH) treated with conservative therapy.

Case Report

A 56-year-old male came to emergency room with chief complaint of atypical bilateral chest pain and shortness of breath. The chest pain was getting more persistent and increasing gradually without any radiating pain in the past 1 week. Shortness of breath was felt in the last 3 years due to chronic obstructive pulmonary disease (COPD) (spirometry test 1 month before was as follow: FEV1/FVC ratio of 0.48 with FEV1 of 40%) and worsened 5 days before admission to the emergency room. He had been diagnosed with tuberculosis infection 3 years ago and already completed the 6-month treatment. Furthermore, he suffered from heart failure 2 year ago but did not comply to routine check-up. He was an active smoker with daily consumption of 2 packs per day since he was young until he was admitted to the emergency room.

On physical examination, he was fully awake with the vital sign as follow: Blood pressure (BP) of 90/60 mmHg, pulse rate (PR) of 92 beats per minute with strong but irregular pulse, respiratory rate (RR) of 30 breaths per minute, oxygen saturation of 88%, and axillary temperature of 36.9°C. His lung examination revealed slight wheezing in bilateral basal region and coarse crackle on superior lobe of right lung. Cardiac examination showed early diastolic murmur at pulmonary valve and holosystolic murmur at tricuspid valve, shifting of cardiac border, and raised jugular vein pressure. Other examinations were within normal limit.

Electrocardiogram (ECG) evaluation showed an occasional premature ventricular contraction (PVC) and premature atrial contraction (PAC) with right axis deviation (RAD), right bundle branch block (RBBB) and right ventricular hypertrophy (RVH), and spike P wave (Figure 1). Chest x-ray showed cardiomegaly and predominate pulmonary cone. There is also an infiltrate in upper lobe of both lung (Figure 2). Laboratory evaluation showed slight leukocytosis (12.800/mm³) and hyponatremia (124 mmol/L), while the CK-MB titer was normal (20 U/L).

Based on the initial evaluation, the patient was diagnosed with congestive heart failure because of cor pulmonale with suspected PAA and pneumonia. He was then transferred to high care unit where he was given oxygen mask 6 L/min, treated with oral spironolactone 25 mg and bisoprolol 2.5 mg once daily. He also received intravenous levofoxacin 750 mg once daily, furosemide 20 mg thrice daily, and dopamine with dosage of 3 mcg per kg body weight (BW). Five hours later, his blood pressure drops to 80/60 mmHg with sign of cardiogenic shock. The dopamine then was titrated up to 5 mcg per kg BW. He was
scheduled for transthoracic echocardiography (TTE) and multi slice computed tomography (MSCT) on the following morning.

TTE result showed normal kinetic on left ventricular segmental analysis, normal left ventricular (LV) systolic function with ejection fraction (EF) of 89%, grade I diastolic dysfunction on LV, decreased right ventricular (RV) systolic function (TAPSE 1 cm) with right atrium (RA) and RV dilatation, severe tricuspid regurgitation (TR), mild pulmonary regurgitation (PR), and high probability of severe PH (TR peak velocity 5.21 m/s, estimated mean pulmonary artery pressure 68.99 mmHg), accompanied by main pulmonal artery (MPA) aneurysm (4.2 cm in diameter) (Figure 3, Video 1). No congenital or structural heart defect was found. Contrast-enhanced thorax MSCT showed an aneurysm on the main pulmonal artery (MPA) (4.39 cm in diameter) and bilateral pulmonal artery dilatation with wall adherent apposition thrombus (Figure 4).

With additional findings from TTE and MSCT, the patient was then diagnosed with heart failure with preserved EF and RV dysfunction secondary to suspected severe PH due to COPD and CTEPH along with MPA aneurysm. The given therapy was continued with addition of beraprost sodium 20 mcg twice daily and warfarin 2 mg once daily. On the fifth day, he became stable with blood pressure of 90/60 mmHg and the symptoms was improved. He was then scheduled to be referred to tertiary hospital for further evaluation with right heart catheterization (RHC), but he refused. Therefore, conservative treatment with oral therapy of furosemide 40 mg once daily, bisoprolol 2.5 mg once daily, spironolactone 25 mg once daily, beraprost sodium 20 mcg twice daily, and warfarin 2 mg once daily continued.

Six months after regular monthly follow-up, the patient’s condition was improved with stable hemodynamic and without any significant complaint. TTE evaluation showed a decreased in the PH severity grading from severe to moderate PH and TR severity grading from severe to moderate. Moreover, the MPA diameter was not increased, with RA and RV dilatation and normal LV and RV systolic function (Figure 5).

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**Figure 2.** Chest x-ray anteroposterior (AP) position showed cardiomegaly with cardiothoracic ratio (CTR) of 67% and predominate pulmonal cone, suggesting a possibility of pulmonary artery aneurysm. There was also cephalization and prominent left and right pulmonal artery with abrupt narrowing in distal segment.

**Figure 3.** Transthoracic echocardiography evaluation before conservative treatment: (A) parasternal short axis view showed pulmonary artery aneurysm with diameter of 4.2 cm. (B) Apical 4 chamber showed dilatation of right atrium (RA major 6.2 cm, RA minor 4.7 cm) and right ventricle chamber (RVDB 3.0 cm). (C) Severe tricuspid regurgitation (TR maxPG 108.44 mmHg) with high probability of PH (TR peak velocity was 5.21 m/s, with estimated mean pulmonary artery pressure of 68.99 mmHg). RA – right atrium; TR – tricuspid regurgitation; PH – pulmonary hypertension.
Video 1. Transthoracic echocardiography from: (A) apical 4 chamber view, (B) parasternal long axis view, (C) parasternal short axis aortic valve view, (D) parasternal short axis pulmonal artery.

Figure 4. Multi slice computed tomography evaluation before conservative treatment: (A, B) aneurysm of main pulmonal artery. (C) Aneurysm of main pulmonal artery with pulmonary trunk of 4.39 cm in size and dilatation of bilateral main pulmonal artery with wall adherent apposition thrombus, predominantly at right pulmonal artery (black arrow), and an eccentric filling defect at left pulmonal artery (red arrow).
Discussion

The location of PAA can be classified into proximal and peripheral. Proximal PAA involve MPA and right and left PA, while peripheral PAA involve intrapulmonary artery [5]. The reported clinical manifestations of PAA include hemoptysis, shortness of breath, chest pain, palpitations, or syncopal episodes. If bronchial compression due to a large PAA occurs, there will be additional clinical manifestation i.e., cough, worsening dyspnea, cyanosis, or pneumonia [4]. In this patient, the clinical manifestation was chest pain and shortness of breath. Cause of ischemic heart disease was excluded from ECG, cardiac biomarker result, and echocardiography which shown no indication of coronary artery disease.

Potential complications of PAA include rupture, dissection, thrombosis, severe pulmonary valve regurgitation secondary to valve cusp malcoaptation, compression of large airways, and coronary artery compression [6]. In our patient, there was no complication occurred. Although there are various cardiac imaging modalities, pulmonary angiography still remains as the gold-standard for diagnosing PAA because of its capability to evaluate pressure gradients within the right side of the heart and the extension of aneurysmal structure into the vascular structure [7]. However, we could only evaluate using TTE and CT because of the limited resources in our hospital.

The etiology of PAA could be differentiated into congenital, acquired, and idiopathic causes. Congenital cardiac anomalies (mostly PDA) or pulmonal valvular stenosis leading to pulmonary arterial hypertension, are the most common cause of PAA [2]. In this case, PAA occurred because of one of the acquired causes, PH. Congenital causes were less likely because of the presenting patient’s age and the absence of congenital heart defect from TTE evaluation (neither PDA, ASD, nor pulmonary stenosis suspicion were found). Although our patient had a history of lung TB 3 years ago, the symptoms in our patient and the location of the aneurysm did not match with the description of Rasmussen aneurysm, a pseudoaneurysm secondary to pulmonary TB [8,9]. For PAA to be considered as caused by idiopathic causes, it has to fulfill several criteria: 1) simple dilatation of the pulmonary trunk; 2) absence of abnormal intra or extracardiac shunt; 3) absence of chronic cardiac or pulmonary disease; and 4) absence of arterial disease [10]. In this patient, idiopathic criteria were not fulfilled because of chronic pulmonary disease.

Gold standard for diagnosing PH is by using RHC to determine the pulmonary arterial pressure at rest (normal level is 10 mmHg) or greater than 30 mmHg during exercise (normal level is 15 mmHg) [11]. In this study we could not confirm the diagnosis of PH due to limited facilities and patient refusal to be referred for RHC. However, high probability of PH could be suspected using TTE and MSCT evaluation. The presence of PH should be suspected when the diameter of the MPA on MSCT is greater than 29 mm or the TR velocity is more than 3.4 m/s [12,13]. In our patient, the MPA diameter from MSCT evaluation was 42 mm. From TTE evaluation, the TR peak velocity was 5.21 m/s with estimated mean pulmonary artery pressure of 68.99 mmHg. These findings suggest high probability of PH.

Etiology of PH is categorized into 5 groups according to the World Health Organization (WHO), which are: pulmonary arterial hypertension (group 1); pulmonary hypertension due to left heart disease (group 2); pulmonary hypertension due to lung disease and/or chronic hypoxia (group 3); pulmonary hypertension due to blood clots in the lungs (group 4); and pulmonary hypertension due to blood and other disorders (group 5) [14]. In our patient, we suggest that the cause of PH was COPD (group 3) and chronic thromboembolism (CTEPH) (group 4). The COPD was confirmed from the
latest spirometry evaluation. CTEPH was suspected from the presence of bilateral apposition thrombus found in MSCT, supported by clinical examination and TTE evaluation findings (WELL score: 0, revised GENEVA score: 3, no cardiac thrombus and McConell sign was found, IVS septum was not flattened). This finding suggest that the thrombus was because of CTEPH rather than acute pulmonary embolism. However, we could not confirm using pulmonary perfusion scintigraphy due to the limited facilities in our hospital.

To this date, there are no definite guidelines on the optimal treatment for patients with PAAs because there are only a few reported cases. It is suggested that patients with PAA caused by PH should be considered for surgical treatment because of the risk for impending dissection and rupture. Moreover, surgery seems to be the only treatment with the possibility of effective long-term survival [15]. Recent review article propose an indication for surgery as follows: 1) absolute PAA diameter ≥5.5 cm; 2) increase the diameter of the aneurysm in 6 months; 3) compression of adjacent structures; 4) thrombus formation in the aneurysm sack; 5) appearance of clinical symptoms; 6) valvular pathologies or shunt flow; 7) verification of PH; and 8) sign of rupture or dissection [2]. In this case, we realize that the patient fulfills the criteria for surgery (PAA with PH and thrombus formation in the aneurysm sack). However, due to lack of resources in our hospital to perform the surgery and the patient’s refusal to be referred to tertiary referral hospital, we opted to treat the patient with conservative treatment. The treatment aims to lower the pulmonary artery pressure while monitoring the aneurysm.

The latest guideline recommends giving supportive therapy (oral anticoagulants, diuretics) and specific drug therapy (calcium channel blocker, endothelin receptor antagonist, phosphodiesterase type 5 inhibitor, or prostacyclin analogues) for treating PH [16]. In this case, the patient was given prostacyclin analogues because it was the only available specific drug therapy which was covered by Indonesia National Health Insurance. Other treatments given include beta blocker for the heart failure, diuretics for supportive therapy, and oral anticoagulant for both supportive PH therapy and chronic thromboemboli.

Six months conservative treatment showed a desirable outcome marked by an improvement of patient’s condition and PH severity. Moreover, the PAA was still intact without any increase in diameter. The successfullness of the conservative treatment also showed in previous reports. Hernández et al. reported 4 patients with proximal PAA with severe PH that had been treated conservatively (2 patients with sildenafil and the other 2 patients treated with the combination of sildenafil and subcutaneous teprostatin). After 2 to 3 years of follow-up, the aneurysms were persistent and showed no sign of complication [17]. Other case report of 65-year-old patient with proximal PAA and PH which had been treated conservatively with intravenous eproprostenol showed that after 20 months of treatment, pulmonary artery pressure was nearly normalized and the size of PAA remains the same [18]. However, recent studies found that the progression of PAA dilatation is independent of the change in pulmonary artery pressure [19,20]. We argue that the difference between our findings or the previous case reports and the previous cohort studies was because of the difference in the follow-up period. The follow-up period in our study was 6 months, while the previous studies follow-up period was longer (one study with mean follow-up period of 942 days, other study with median follow-up period of 3.6 years). Thus, even though the pulmonary artery pressure was already normalized, clinician should still encourage the patients to get intervention therapy or surgery because the risk of PA diameter increase and rupture still exist.

Conclusions

PAA is a rare disease that is difficult to diagnose. Persistent atypical chest pain can be an early symptom of PAA; thus, clinicians should be aware in a high-risk patient suffered persistent chest pain, despite normal ECG and laboratory findings. TTE and MSCT evaluation are reliable for early detection of PH and PAA when the gold-standard modalities are not available. Even if the pulmonary artery pressure had been normalized, the risk of PA diameter increase and rupture still exist and must still be concerned.

Conflict of interest

None.

Department and Institution where work was done

The work was done in the Department of Cardiology and Vascular Medicine Dr. Koesma General Hospital, Tuban, Indonesia.
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