



## Case report

# Unilateral pulmonary artery agenesis manifested by hemoptysis in pregnancy

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## ABSTRACT

Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly which can be symptomatic or even asymptomatic. Most of patients with isolated UPAA have mild symptoms and it is difficult to be diagnosed, especially when abnormal findings of chest radiograph are the first presentation. It is often misdiagnosed and is not considered during differential diagnosis. To make a diagnosis of UPAA, various imaging modalities including chest radiograph, computed tomography (CT), and angiography are used. We report a 33-year-old woman in pregnancy presented recurrent hemoptysis whose CT was postponed due to her pregnancy. Although CT is a useful diagnostic tool, chest radiograph could be used instead in pregnancy suggesting UPAA with a lot of information.

## 1. Introduction

Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly. Since it was first described in 1868 [1], more than 420 cases have been reported [2]. It is frequently accompanied with congenital cardiovascular abnormalities, such as tetralogy of Fallot, atrial septal defect or patent ductus arteriosus [3]. On the other hand, it can also occur as a single finding, isolated UPAA, which can be symptomatic or even asymptomatic. It is diagnosed in adulthood using various imaging modalities including chest radiograph, computed tomography (CT), and angiography [4]. Here, we report a 33-year-old woman in pregnancy presented recurrent hemoptysis whose CT was postponed due to her pregnancy.

## 2. Case

A 33-year-old woman in the third trimester of pregnancy presented at the emergency department with hemoptysis of unknown cause. The amount of hemoptysis was small and she had never had any hemoptysis or dyspnea before. Her vital signs were stable including those of fetus. No past medical history was reported and she was a never-smoker. She did not report any other symptom except hemoptysis. On the physical examination, slightly deviated trachea was found. Lung auscultation and the other physical examination were unremarkable. Chest X-ray showed the displacement of mediastinal structure to the right,

decreased volume of the affected lung, and decreased hilar vasculature of ipsilateral side, suggesting UPAA (Fig. 1).

At the emergency department, she refused to take chest CT because of her pregnancy. A week later, on the second postpartum day, she took chest CT for hemoptysis. It showed absence of right pulmonary artery (Fig. 2A) with marked hypertrophy of ipsilateral bronchial arteries (Fig. 2B), right lung hypoplasia with mild bronchiectasis, and consolidation on right upper lobe due to hemorrhage. Echocardiography demonstrated mild pulmonary hypertension and right ventricular hypertrophy without other congenital cardiovascular abnormalities. Her symptoms were improved after conservative treatment, and she was scheduled to regular follow-up.

## 3. Discussion

UPAA is difficult to be diagnosed, especially when abnormal findings of chest radiograph are the first presentation. It is often misdiagnosed and is not considered during differential diagnosis. In 1995, Bouros et al. emphasized that a high index of suspicion is needed to diagnose UPAA in patients with unilateral hyperlucent lung [5]. Several studies also have reported that awareness of UPAA may help to make the diagnosis [6,7].

In addition to high index of suspicion, the diagnosis of UPAA is based on history taking, physical examination and imaging work up. It has various presentations which can be symptomatic or even

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**Fig. 1.** 33-year-old woman with isolated pulmonary artery agenesis. Chest X-ray shows displacement of mediastinal structure to right, nodular opacity on right upper lung and decreased volume of right lung.

asymptomatic. Some patients present dyspnea because of accompanying congestive heart failure or pulmonary hypertension while other asymptomatic patients are diagnosed incidentally. In a previous study, 108 cases of isolated UPAA were reviewed and about 15% of patients were asymptomatic who were diagnosed during preoperative examination, tuberculosis survey or examination for enlistment in the army [4]. On the contrary, symptomatic patients present dyspnea, exercise intolerance, or recurrent infection. About 10–20% of patients with UPAA show inconsequential hemoptysis. Pulmonary hypertension was also reported in 25% of patients. Physical examination usually unremarkable except for deviated trachea, unilaterally small hemithorax, and decreased breathing sound on the affected lung [4–6,8,9].

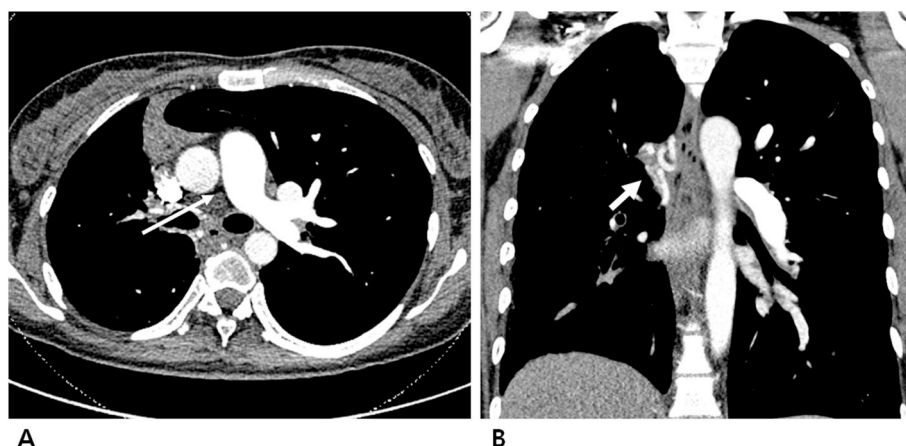
Chest radiograph shows decreased volume of the affected lung, mediastinal displacement to ipsilateral side and decreased hilar vasculature. If there are suspicious findings on chest radiograph, definite diagnosis can be made with CT scan or with magnetic resonance imaging (MRI) [7,10]. Transthoracic echocardiography is used to evaluate other congenital cardiovascular anomalies or pulmonary hypertension. If there was no pulmonary hypertension, follow-up echocardiography is recommended to detect early pulmonary hypertension [11,12]. Although angiography has been considered as a gold standard to diagnose UPAA, CT replaces the role of angiography with its advance. Instead, recent angiography is used to embolize selected arteries when patients present massive hemoptysis [6,13].

There is no agreement on the treatment of isolated UPAA and it depends on symptoms, collateral vasculatures and the anatomy of pulmonary artery. Regular follow-up is required in asymptomatic patients to monitor the development of pulmonary hypertension [12]. Hemoptysis is treated with occlusion of collateral vessels using intervention or surgery. Pneumonectomy and surgical revascularization are reserved for recurrent hemoptysis, recurrent respiratory infection, and pulmonary hypertension. Considering that 8% of patients were required to undergo either pneumonectomy or lobectomy for recurrent hemoptysis and recurrent pulmonary infections [4], it is significant for these patients to be diagnosed early and follow-up closely. Heart-lung transplantation may also be considered to improve oxygenation [14].

The diagnosis of UPAA is difficult which is usually diagnosed with CT or angiography. In our case, radiological imaging modalities were limited to chest radiograph due to her pregnancy. Her chest radiograph, however, demonstrated UPAA with its typical manifestations. Although chest radiograph is considered as an insufficient diagnostic tool in UPAA, it could be an important clue to diagnosis in case of pregnancy.

#### 4. Conclusion

UPAA is a rare congenital anomaly which is difficult to be diagnosed. In pregnancy, CT is not available and a simple chest radiograph could be used instead as a significant modality to diagnosis of UPAA.



**Fig. 2.** (A) The sagittal section of CT angiography shows absence of the right pulmonary artery (long white arrow) and (B) the coronal section shows right bronchial artery hypertrophy (short white arrow).

## Conflicts of interest

None declared.

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## Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.rmcr.2018.10.028>.

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