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Case report

An elderly patient and an adult patients with isolated unilateral pulmonary arterial hypoplasia: Two cases reports and literature review of the literature

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ABSTRACT

Unilateral hypoplasia of a pulmonary artery (UHPA) is a rare condition that is usually associated with cardiac anomalies in childhood. In the absence of cardiac anomalies, patients may not be diagnosed until respiratory symptoms develop. We present two patients who were diagnosed as having isolated unilateral right pulmonary artery hypoplasia, one a woman aged 80 years, the other, a man aged 55 years. To our knowledge, woman is the oldest patient with UHPA in the English medical literature. Awareness of this condition may contribute to the early recognition of these cases and planning of appropriate treatment.

1. Introduction

Unilateral agenesis and hypoplasia of a pulmonary artery (UAPA/UHPA) are rare congenital anomalies. The estimated prevalence of isolated UAPA ranged from 1 in 200 000 to 1 in 300 000 [1,2]. It is caused by involution of the proximal sixth aortic arch resulting in the absence of the proximal pulmonary artery (PA). The distal part of the PA is often present as a small vessel or fibrous cord [3]. Unilateral agenesis of the right PA is more common on the right side, but unilateral agenesis of the left PA is frequently associated with congenital cardiac malformations such as tetralogy of Fallot, atrial septal defect, coarctation of the aorta, and patent ductus arteriosus [1]. Adult patients without cardiac anomalies are rare, and these patients have no symptoms or mild dyspnea during exercise. Therefore, most patients with UHPA who have no cardiac pathology are diagnosed at adult ages. We report 2 patients with isolated UHPA without cardiologic abnormalities who were diagnosed in a late decade.

2. Case presentation 1

A woman aged 80 years was admitted to hospital with dyspnea, cough, and fatigue, which she had had for 2 years. She reported having pulmonary tuberculosis when she was aged 15 years. The patient had no smoking history. She was using bronchodilator drugs for asthma and levothyroxine for hypothyroidism. In the physical examination, decreased breath sounds on the right side and mild wheeze were found. Her complete blood count and biochemistry parameters were normal. Lung function tests revealed a mild obstructive pattern: forced vital capacity (FVC) 1.60 L (92.7% predicted), forced vital capacity in one second (FEV1) 1.22 L (88.7% predicted), and the FEV1/FVC ratio was 0.76.

Chest X-ray showed decreased lung volume on the right lung, shrunken right hilus, a calcific nodule on the right upper lobe, and hyperinflation in the left lung. (Fig. 2a-paag). Computed tomography (CT)-angiography revealed agenesis of the main right pulmonary artery, and bronchiectasis on the right upper and right lower lobes. Bilateral mosaic perfusion was seen on the parenchyma slices of the chest CT (Fig. 2b-bt). Abdominal ultrasonography showed no significant anomalies. Electrocardiography and echocardiography revealed no cardiac abnormalities. The patient was diagnosed as having agenesis of the right pulmonary artery. Her clinical status remained stable during a 16-month follow-up period.

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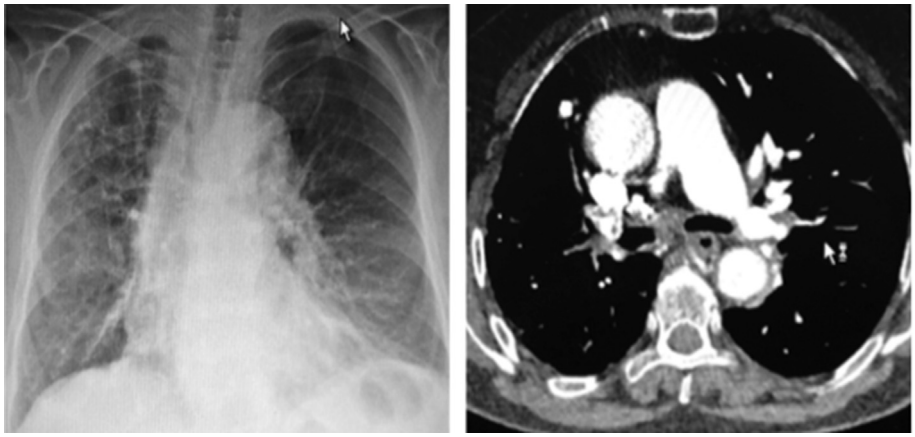

Fig. 1. Chest X-Ray of case 1 showed decreased right lung volume, calcification on the right upper zone and absence of right hilar vascular shadow. Chest CT of case 1 showed that hypoplasia of right pulmonary artery.

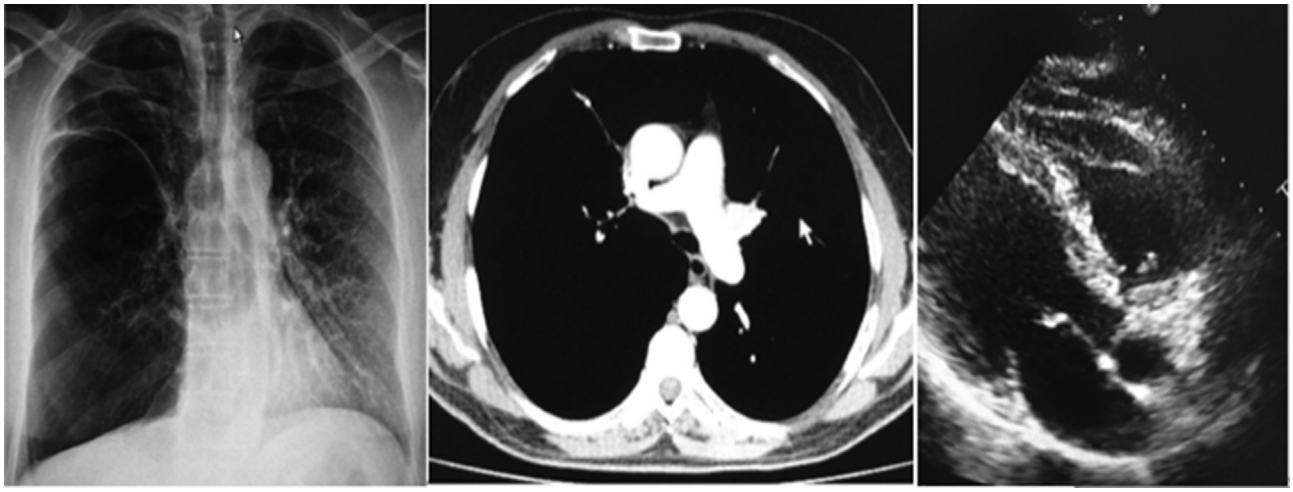

Fig. 2. Chest X-Ray of case 2 revealed that hyperlusent of right lung and absence of right hilarious vascular shadow. Chest CT of case 2 revealed that hypoplasia of right pulmonary artery, fibrotic linear lesion on the right upper lobe.Echocardiography of case 2 was normal.

3. Case presentation 2

A man aged 55 years was admitted to the outpatient clinic of our hospital with cough and dyspnea. He had a 30 pack-year smoking history. He had had exertional dyspnea for 3 years. He was diagnosed as having chronic obstructive lung disease (COPD) and he used bronchodilator drugs. He reported a history of 2 exacerbations of COPD in the last 2 years, one of which required treatment in an intensive care unit. There was no history of any other diseases. His vital parameters were normal. An examination of his respiratory system revealed decreased breath sounds on the right side and mild wheeze on the left side. His complete blood count and biochemistry parameters were normal. Pulmonary function tests were FVC 2.0 L (57% predicted), FEV1 1.1 L (58% predicted), FEV1/FVC ratio 0.82, and his diffusion capacity for carbon monoxide (DLCO) was 18 mm/min.kPA (74% predicted) with mild restrictive respiratory dysfunction and decreased diffusion capacity. Blood gas analysis was normal when breathing room air.

Chest X-ray showed hyperinflation in the right lung, and shrunken right hilus (Fig. 1a). CT of the chest demonstrated that the right pulmonary artery diameter was smaller than the left pulmonary artery (13.5mm vs. 23.5mm, respectively). Minimal bronchiectasis and fibrotic shrinkage were seen in the right upper lobe. Thorax CT showed no parenchymal perfusion on the right middle and lower lobe (Fig. 1b). Therefore, a ventilation/perfusion scan of the lung was performed. A decreased perfusion defect in the right upper lobe and complete absence of uptake in perfusion scans of the right middle and lower lobes were detected. Abdominal ultrasonography showed no significant anomalies in the liver, kidney, spleen, and other organs. To evaluate cardiac abnormalities, electrocardiography and echocardiography were performed. No cardiac abnormalities were detected (Fig. 2c). The patient was diagnosed as having hypoplasia of the right pulmonary artery. His clinical status remained stable during a 30-month follow-up period.

4. Discussion

UHPA or UAPA is a rare anomaly that is frequently associated with cardiac pathology. It was reported that the UHPA was more common on the left side compared with the right [4]. Patients with cardiac pathology are diagnosed and treated at an early age. It is rare for UHPA to remain asymptomatic and be diagnosed in adult age, as in our cases.

The most common symptoms of UAPH/UHPA are shortness of breath, especially during exercise, cough, hemoptysis, and frequent pulmonary infections [1]. Ten et al. reported that the mean age of

patients with UAPA was 14 years (range, 0.1–58 years) and they had symptoms such as dyspnea or limited exercise intolerance (40%), hemoptysis (20%), and recurrent pulmonary infections (37%). Pulmonary hypertension (PHT) was present in 44% of 108 patients with UAPA [4]. Some previous studies reported that 13–30% of patients were asymptomatic [2,4]. Our patients were diagnosed at the ages of 55 and 80 years, presumably due to the absence of disease-specific symptoms or any cardiac abnormalities. To our knowledge, the woman is the oldest patient with UHPA in the English medical literature. This patient had only had shortness of breath for two years and was diagnosed as having asthma. The other patient, a man aged 55 years, had exertional dyspnea and had used bronchodilator drugs for COPD for 3 years. Patients might be misdiagnosed as having asthma, as with ours.

The diagnosis of UAPA/UHPA is very challenging and is based on medical history, physical examination, chest X-ray, thorax CT, ventilation-perfusion scans, and echocardiography to investigate related cardiac anomalies. The gold standard test for the diagnosis of UAPA/UHPA is pulmonary angiography. It can also be diagnosed with CT angiography. Angiography should be performed preoperatively in patients with cardiac pathologies [1,2,4]. Radiologically, it presents as predominant unilateral hyperlucency. Other causes of unilateral hyperlucent lung, such as pneumothorax, idiopathic giant bullae, congenital lobar emphysema, and bronchial obstruction due to a foreign body or endobronchial malignancy, should be considered. Other chest X-ray or CT findings include absence of the pulmonary artery shadow with enlargement of the contralateral pulmonary artery shadow, volume loss of the affected side, and ipsilateral mediastinal shift [5]. Three-dimensional reconstruction of thorax CT can be helpful for outlining abnormalities of the bronchi and associated vascular structures. Ventilation-perfusion scans of the lung can show complete absence or diminished perfusion in the affected side [6].

The clinical presentation is affected by age. Infants usually present with congenital cardiac diseases, such as tetralogy of Fallot, atrial septal defect, coarctation of the aorta, right aortic arch, truncus arteries patent ductus arteries, and pulmonary atresia. Older patients are usually asymptomatic and are incidentally detected during chest X-ray or CT evaluations [1]. To determine cardiac pathology and PHT, all patients should be evaluated using transthoracic echocardiography. Our patients were assessed by a cardiologist and no cardiac pathologies or PHT were detected.

The treatment decision is based upon on the age of patients, symptoms, and the presence of cardiac abnormalities. Asymptomatic patients with no evidence of cardiac diseases do not usually need treatment [1,5]. Pneumonectomy, lobectomy, embolization of collateral arteries, and revascularization are considered for patients with PHT, recurrent pulmonary infections or hemoptysis [1,7,8]. The presence of PHT may affect survival for adult patients with UAPA/UHPA [1]. Pharmacotherapy such as bosentan or parenteral prostacyclins should be considered for the treatment of PHT [1]. The prognosis depends on related cardiac pathologies and is good if there are no another malformations [1].

5. Conclusion

Isolated UAPA/UHPA are rare anomalies and usually diagnosed in childhood. However, it can remain unnoticed until late decades. We presented two patients with isolated UHPA who survived to middle and old age with mild dyspnea and no other congenital cardiac anomalies. Awareness of this condition may contribute to the early recognition of these cases and planning of appropriate treatment.

Conflict of interest

The authors declare that they have no conflicts of interest concerning this article.

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