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A rare manifestation of pulmonary artery agenesis

Ретка манифестација агнезије плућне артерије

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SUMMARY

Introduction Unilateral absence of pulmonary artery is rare vascular malformacy. Because of this anomaly the lungs are supplied by the system of collateral arteries.

Case outline We report a case of right pulmonary artery agenesis in female patient. She was admitted to hospital because of hemoptysis. CT scan revealed the congenital malformation -right lung smaller dimensions, the right principal pulmonary artery has not been developed, and aberrant tortuous blood vessels.

Conclusion In the case of our patient was applied to symptomatic therapy. There was no need for any surgical treatment. However, in case of massive hemoptysis will probably applied embolization or lobectomy/pneumonectomy.

Keywords: pulmonary arteries; hemoptysis; computed tomography; angiogenesis

САЖЕТАК

Увод Једнострано одсуство плућне артерије је ретка васкуларна малформација. Због ове аномалије плућа се васкуларизују системом колатералних артерија.

Приказ болесника Приказујемо редак случај агнезије десне плућне артерије код пацијенткиње. Болесница је иницијално хоспитализована због манифестних хемоптизија. Компјутеризована томографија грудног коша је показала конгениталне малформације - мању димензију десног плућа, десна главна плућна артерија није била развијена, као и аберантне крвне судове.

Закључак У случају наше пацијенткиње примењена је симптоматска терапија. Није било потребе за било каквим хируршким третманом. Међутим, у случају масовних хемоптизија вероватно ће се применити емболизација или лобектомија/пнеумонектомија.

Кључне речи: плућне артерије; хемоптизије; компјутеризована томографија; ангиогенеза

INTRODUCTION

Unilateral absence of pulmonary artery (UAPA) was first described by Frentzel in 1868 and was demonstrated angiographically in 1952[1]. The most likely theory of UAPA is due to the developmental failure of the left sixth arch, resulting in the absence of the pulmonary artery [2].

Pulmonary artery agenesis can be localized to a single lobe, also can affect an entire lung or in very rare case both lungs. Because of that the lungs are supplied by the system of collateral arteries from bronchial, subclavian, intercostal and coronary arteries . The increased pressure in the collateral arteries may lead to a damage of pulmonary arteries, endothelial tissue damage and pulmonary arterial hypertension. These submucosal collaterals hypertrophy with time and may rupture causing hemoptysis, which is the most common symptom [3 – 7].

Congenital unilateral absence of any of pulmonary artery (UAPA) is an extremely rare anomaly (approximately 0.39% of all congenital heart diseases), with a prevalence of 1 in 200,000 young adults [3]. This malformation occurs less often as compared to total anomalous pulmonary venous connection (TAPVC), that is 0.7-1.5% of all congenital heart malformations [8]. Anatomical characteristic of TAPVC is an abnormal connection of pulmonary veins with systemic venous circulation, but so far no association has been described with UAPA [8].

UAPA is usually associated with other cardiovascular abnormalities (tetralogy of Fallot, ventricular septal defect, coarctation of the aorta, subvalvular aortic stenosis, scimitar syndrome).

It is, in most cases, diagnosed in childhood. The average age of UAPA patients is 14 years.

However, because of the atypical symptoms some patients can be diagnosed in adulthood with the symptoms: hemoptysis, dyspnea, reduced exercise tolerance, recurrent bronchopneumonia. In some patients chronic infection can lead to bronchiectasis [9 – 12].

Treatment options include revascularisation surgery, pulmonary vasodilator therapy (for pulmonary hypertension), pneumonectomy or lobectomy, embolisation of collateral hemorrhage [11 – 17].

CASE REPORT

A 28-year-old, non-smoker female, was admitted in Clinic for Pulmology, Clinical Center of Serbia for evaluation of hemoptysis, which have occurred for the first time 4 years ago. Hemoptysis, mostly in an effort, repeated again at the end of 2013. and early 2014. but patient did not visit a doctor. In a meantime, until the present admission to the hospital, the patient led a normal life, she had a healthy baby by a normal delivery.

Physical examination was unremarkable. She had normal vital signs with oxygen saturation of 97% on room air. Her lungs were clear to auscultation, only over the basal part of the right lung weakened sound, without wheezes or rales. Her cardiovascular exam revealed normal heart rate and rhythm. Laboratory data showed normal complete blood count and chemistries.

On posteroanterior chest x-ray the mediastinal shadow and heart were shifted to right, volume of the right lung was smaller with hyperinflated left lung (Fig.1).

Total lung capacity of 106% of predicted, RV 101%, and a diffusion capacity for carbon monoxide of 72% of predicted.

A perfusion scintigram showed the absence of perfusion in the right lung.

Bronchoscopy was normal (no deformity in the bronchial tree, normal arborization of the airways), but . incidental finding during bronchoscopy was nasal polyps tend to bleed.

Echocardiography showed that dimensions of all cardiac chambers were normal. Atrial septum was normal with suspected ductus arteriosus persistent, and there was absence of the right pulmonary artery. There was no pulmonary hypertension.

Hemodynamic examination (coronary angiography, right ventriculography revealed) tricuspid regurgitation 2+, pulmonary angiography revealed aplasia of the right pulmonary artery while left pulmonary artery arborization was normal. Aortography revealed ductus arteriosus persistent. There was normal pressure in the right and left heart.

Another notable point of this case is that CT scan revealed the congenital malformation (Fig.2, Fig3, Fig 4) -right lung smaller dimensions, developed with all three lobes, the left lung was hyperinflated. Pulmonary trunk after leaving the right atrium continues to the left principal pulmonary artery while the right has not been developed. Immediately below the aortic arch are separating two aberrant tortuous blood vessels. Medial blood vessel is

dominant and supply the greater part of the right lung. All four pulmonary veins drain into the left atrium.

Selective aortography we found the existence of multiple aortopulmonary collateral artery (MAPCA). The three main collateral comes from descending aorta and supplying all three right lung lobes, and there is also a secondary collaterals from the brachiocephalic trunk which supplies the smaller parts of the upper right lobe. All collateral are very tortuous, but at no time does not display the right pulmonary artery (Fig 5, Fig 6).

DISCUSSION

We report a case of young woman, who was admitted in hospital because of haemoptysis. During the treatment in hospital, we found that patient had UAPA.

Patients with UAPA can remain asymptomatic for a long period , or may include hemoptysis (in 20% of patients), dyspnea in physical activity, recurrent respiratory infections, chest pain, or pleural effusion. The most common symptom is haemoptysis but massive and life-threatening hemoptysis could also occur. Pulmonary hypertension was diagnosed in 25% of the patients with UAPA and it's a bad prognostic sign [9].

Sometimes haemoptysis can be provoked by factors such as exercise or in pregnancy. Although our patient had one term deliverie, she had no complaints.

In case of our patient, that was isolated UAPA without any other cardiovascular anomaly.

Optimal management requires a multi disciplinary approach for diagnostic and treatment.

For diagnostic it can be ussed X ray chest radiography, MSCT scan, magnetic resonance imaging (MRI), ventilation-perfusion scintigraphy and angiography.

On chest X-ray can be reduction in the volume of hemithorax, an elevation in the hemidiaphragm and mediastinal shift in the affected side [3].

Ventilation-perfusion scintigraphy is rarely performed today and it can show absence of perfusion on the affected lung with normal ventilation [3].

Angiography is the golden standards for establishing a definitive diagnosis and identify the collaterals to the affected lung [14]. In our case the blood supply to the affected lung comes from a branch of the artery from the descending aorta, forming collateral circulation for the aorta-pulmonary artery.

Symptomatic treatment consists of medications such as antibiotics, expectorants and bronchodilators, the treatment of pulmonary hypertension and any other treatments for complications. Today in this cases it is very important prophylaxis for respiratory syncytial virus, pneumococcus, influenza infections [15].

Surgical UAPA treatment methods are revascularization (a systemic-pulmonary shunt involving hilar arteries), lobectomy, pneumonectomy and embolization of developed aorto-pulmonary collateral arteries. Also, re-anastomoses of peripheral pulmonary arteries and the pulmonary trunk have been described in literature [11,17].

According to the our council decision (pulmonologist, thoracic and cardiovascular surgeons) we applied only symptomatic therapy. There was no need for any surgical treatment in this moment because that was not massive hemoptysis.

We advised her to visit the otolaryngologist because it is possible that the cause of hemoptysis was nasal polyps that bleed easily when touched. The mucosa of the polyp that was seen by bronchoscopy is very fragile and bleeds easily.

The patient will be followed by-pulmonologist, thoracic and cardiovascular surgeons. Given that it is innate birth defect that is detected in a patient at an older age, symptoms (hemoptysis) are minimal and mainly on exertion and that her had a successful birth, can be

expected that massive hemoptysis is not coming. However, we advised her that in case of massive hemoptysis should report immediately to the hospital to decide on further treatment. In case of massive hemoptysis will probably applied embolectomy blood vessel that bleeds. As a final option takes into account the eventual lobectomy or pneumonectomy.

Unilateral absence of pulmonary artery is very rare vascular malformacy and may remain undiagnosed for prolonged periods of time. Although this malformation occurs in childhood, in our patient showed after her 25 years and after she became a mother. The delivery is like any kind of activity, risk factor for the hemoptysis because of that the lungs are supplied by the system of collateral arteries. Therefore, it is necessary to think about UAPA in patients with other heart anomalies and if there is a reduced transparency or volume of one lung on X chest radiography. Optimal management requires a multi-disciplinary approach for diagnostic and treatment. In the case of our patient was applied only symptomatic therapy. There was no need for any surgical treatment. But in case of massive hemoptysis can be applied embolisation or surgical treatment of that lung.

Conflict of interest: None declared.

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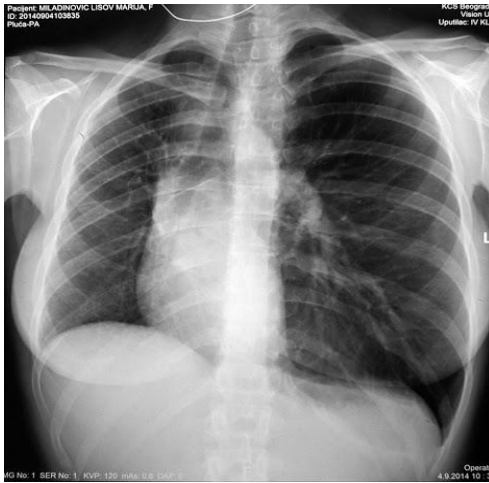


Figure 1. Chest x-ray radiography- the mediastinal shadow and heart shifted to right, volume of the right lung is smaller with hyperinflated left lung

Paper accepted



Figure 2. CT scan - pulmonary trunk after leaving the right atrium continues to the left principal pulmonary artery while the right has not been developed (regular CT scan);

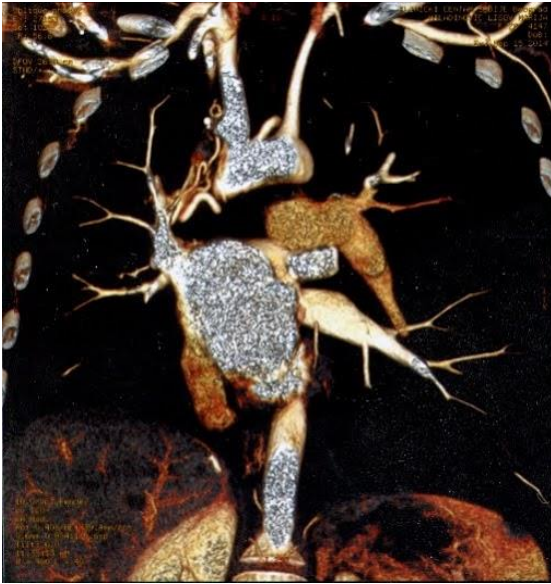


Figure 3. CT scan - pulmonary trunk after leaving the right atrium continues to the left principal pulmonary artery while the right has not been developed (colour 3D CT scan)

Paper accepted



Figure 4. CT scan- three main collateral from descendent aorta

Paper accepted

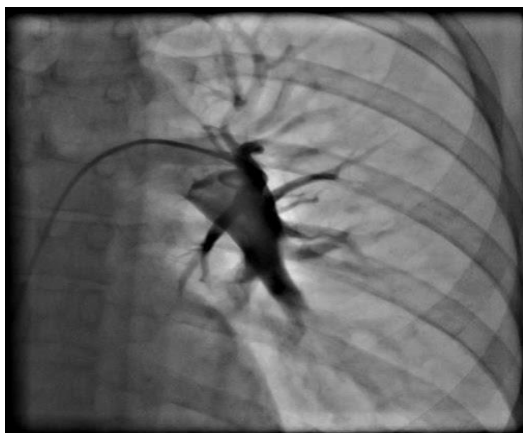


Figure 5. Selective aortography: multiple aortopulmonary collateral artery (MAPCA); A-MAPCA; B-absence of right pulmonary artery on aortography

Paper accepted

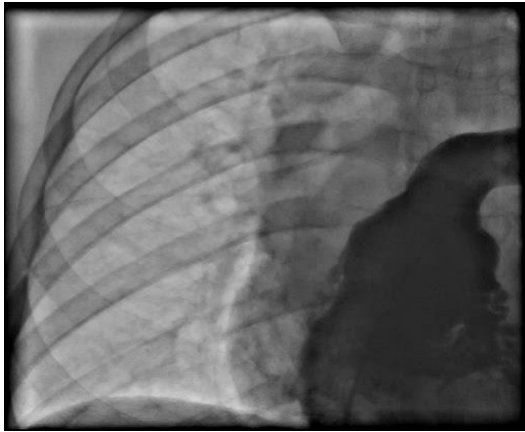


Figure 6. Selective aortography - absence of right pulmonary artery on aortography

Paper accepted