Case Report

Hemoptysis caused by the unilateral absence of a pulmonary artery: a noteworthy diagnosis

Sophie Pohlen [a], Valentine Weber [b], Nicolas Arribard [c], Nasroola Damry [d], Christine Quentin [b]

[a] Pediatric Fellow. Hôpital Universitaire des Enfants Reine Fabiola (HUDERF). Brussels, Belgium.
[b] Pediatric Pulmonology. Hôpital Universitaire des Enfants Reine Fabiola (HUDERF). Brussels, Belgium
[C] Pediatric Cardiology. Hôpital Universitaire des Enfants Reine Fabiola (HUDERF). Brussels, Belgium
[d] Pediatric Radiology. Hôpital Universitaire des Enfants Reine Fabiola (HUDERF). Brussels, Belgium

sophie.pohlen@ulb.be

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Abstract

Unilateral absence of a pulmonary artery is a rare congenital abnormality. The diagnosis is complicated because it remains asymptomatic or exists with non-specific symptoms, unless the abnormality is associated with other cardiac malformations. Hemoptysis is rarely the initial symptom of the malformation, especially in the pediatric population. Computed tomography angiography is the gold standard to confirm the diagnosis. There is no consensus on how the pathology should be managed or treated.

We report a rare clinical case with an initial atypical symptom and review the literature.

Case report

A 13-month-old girl suffering from hemoptysis for two days, during an episode of bronchiolitis was referred to our pediatric service. Clinical examination showed fresh blood in the back of her throat and pulmonary auscultation revealed wheezing. The patient's vital parameters were within normal limits. No other bleeding points were noticed. The anamnesis was able to exclude foreign body aspiration, contact with an irritant product, direct contact with tuberculosis patients and hereditary hemorrhagic diseases prior to the onset of the illness.

The child had been born at full-term by vaginal delivery following a normal pregnancy. Her post-natal adaptation was normal. Upon physical examination at birth three vascular spots were observed on the baby's skin: one on the right knee and two on the left buttock. An ultrasound scan did not reveal any vascular malformation. The patient's medical history mentioned bronchial hyperreactivity treated by inhaled corticosteroids.

Complementary investigations showed hemoglobin to be at 10,4 g/dL (normal value between 13 and 18 g/dL) while platelet and coagulation panel were normal, ruling out the possibility of a systemic hemorrhagic disease. A chest radiograph did not reveal any focus or malformation. Fibroscopy revealed the presence of fresh blood on the vocal cords but no visible active bleeding. Bronchoscopy showed very dilated vessels in the right bronchus, which may have explained the hemoptysis. In addition, a contrast-enhanced computed tomography (CT) of the chest revealed complete absence of the right main pulmonary artery and the presence of collateral circulation arising from the bronchial arteries, intercostal arteries, right internal mammary artery, right subclavian artery and diaphragmatic arteries vascularizing the right lung (fig. 1 and fig. 2). Catheter angiography confirmed the diagnosis of isolated absence of the right pulmonary artery (fig. 3) and showed two important collaterals: a collateral from the right subclavian artery and one originating from the descending aorta. There was no associated heart defect. Pulmonary pressure in the left pulmonary artery was normal (average 18 mmHg).

In summary, only the left lung was involved in oxygenation. The right lung was vascularized by aortic collaterals performing a left-right shunt and, as a result, it only received blood that was already fully saturated. The flow of these aortic collaterals was modest because the child's growth was good. In view of the normal pulmonary pressures and the child's normal growth, the decision was taken to abstain from therapy for the time being. In the event of massive hemoptysis, collateral embolization would have been considered, provided the risk of pulmonary infarct was limited. The child will have to follow a vaccination schedule against pneumococcus and influenza.

One month later, the child returned for recurrence of hemoptysis, caused by an upper respiratory tract infection with influenza. The hemoptysis was limited and did not require any medical intervention.

Discussion

Unilateral absence of a pulmonary artery (UAPA) has been known since 1868 and was first described by Frantzel (1). The malformation is generally associated with other cardiac anomalies and is mostly located on the left side (80 %) (2). Occasionally, the malformation is isolated and in such cases it usually affects the right pulmonary artery (60 %) (2). The prevalence of this rare pathology is estimated at 1/200000 and the mortality can reach 7% caused by massive hemorrhage, right heart failure or respiratory failure (2,3). In case of UAPA, there is an increase in blood flow in the contralateral pulmonary artery resulting in shear stress on the endothelium with subsequent release of vasoconstrictor agents such as endothelin. This results in chronic vasoconstriction of the pulmonary arterioles, leading to remodeling and thus increased resistance in the pulmonary vascular system. The development of pulmonary arterial hypertension leads to right ventricular hypertrophy that can result into right heart failure which can be fatal.

Between 1978 and 2000, Jan Ten Harkel AD et al. have found 108 cases of isolated UAPA that were reported in the literature (4). The authors used the database of the National Library of Medicine. We have used the same database focusing exclusively on the last 10 years, using the key words "isolated absence of pulmonary artery", "children" and "case report". With these search criteria, only 8 cases have been reported over this time period. All cases concerned children under 7 years old, whereas the median age of the cases in Jan Ten Harkel et al. was 14 years (range 0.1 to 58 y), which suggests that this malformation is rarely identified in young children.

When UAPA is associated with other cardiac anomalies, the symptoms may appear early in childhood usually consisting of cyanosis, heart failure, heart murmur or growth retardation. By contrast, isolated UAPA remains

asymptomatic during childhood and goes generally undiagnosed. The median age of diagnosis in the pediatric population is 4 years old (5)range 6 months to 10 years. Most of the cases reported in literature concern adults whose diagnosis is complicated because the symptoms are not very specific: recurrent respiratory infections (37%), dyspnea during exercise (40%) and pulmonary arterial hypertension (25%) (4)diagnostic procedures, and therapeutic strategies of patients with an isolated unilateral absence of a pulmonary artery (UAPA. I.Boudard et al. conducted a retrospective study that included children with UAPA, 75% of them had a symptomatology suggesting asthma, as was the case in our patient (5)range 6 months to 10 years.

Hemoptysis only occurs in 10-20% of cases and is most often seen after observation of systemic collateral vascularization, which has evolved over a number of years (4,6). This explains its absence in the pediatric population. The alveoli are in close contact with the pulmonary arteries, in which pressure is very low, even under stress (20mmHg). In UAPA, vascularization of the lung occurs through collaterals from the systemic circulation, which are subject to higher pressure than normally measured in a pulmonary artery leading to a rupture of their thin walls in case of respiratory tract infection and hemoptysis occurs. To the best of our knowledge, our case report describes the youngest child with hemoptysis as initial symptoms of this pathology reported in the literature to date. We would like to underline the importance of considering pulmonary arterial malformations in the differential diagnosis of hemoptysis in children. In the other few reported cases of infant diagnosis of isolated UAPA reported in literature, the initial signs were cyanosis or murmur (7, 8).

The diagnosis can be suspected on a chest radiograph (3,4)diagnostic procedures, and therapeutic strategies of patients with an isolated unilateral absence of a pulmonary artery (UAPA. This sometimes occurs unintentionally when a different diagnosis was being pursued. The following anomalies can be observed in the adult population: an asymmetry between the two pulmonary fields, a mediastinum shift attracted to the affected side of the lung, an elevation of the ipsilateral diaphragm and a narrowing of the intercostal spaces (2). Differential diagnosis should consider unilateral emphysema, embolism of a pulmonary artery, coarctation of the pulmonary artery and Swyer James syndrome.

Since 1952, angiography has been used as the gold standard to confirm the diagnosis (5). It is the examination method of choice to study the systemic revascularization of the affected lung. Such revascularization is of particular interest in the case of hemoptysis since it directs embolization. Hemoptysis is usually limited, but it can be life threatening when it is substantial. Two approaches are available : embolization and pneumonectomy. Pneumonectomy should be considered in cases of massive hemoptysis, congestive heart failure, bronchiectasis or pulmonary arterial hypertension. In patients having co morbid conditions or for critically ill patients, selective embolization is indicated for the treatment of hemoptysis. This therapeutic approach is safe, effective, well tolerated, minimally invasive and the operation can be repeated. Embolization can also be useful to delay pneumonectomy, as it delays long-term side effects such as scoliosis, thoracic deformity and overdistention of the remaining lung (9). Nonetheless, the embolization procedure can result in complications such as pulmonary infarction, spinal artery embolization, post-embolization syndrome and the procedure has a long-term recurrence rate of 25% (2).

Another promising treatment is the revascularization of the isolated lung at a young age in order to prevent life threatening complications.

The embryologic cause is thought to occur during the involution of the proximal branches of the 6th aortic arch, leading to the absence of the proximal part of the pulmonary artery (7). During embryonic life, there is a distal pulmonary artery which is supplied by an ductus arteriosus. Thanks to this duct, the fetal pulmonary development is normal. At birth, the ductus closes, which leads to homolateral pulmonary hypoplasia and to the development of collateral circulation. A retrospective study has demonstrated that reperfusion of the affected lung before the age of 6 months appears to improve lung growth and to prevent the development of pulmonary hypertension and of collaterals, thus preventing hemoptysis (47%) (10). The techniques that have been used are: persistent ductus arteriosus stenting and the creation of an anastomosis between the main pulmonary artery and the distal part. The retrospective

Figure 1: *Chest CT scan, frontal view.*
Arrows point to an enlarged right internal mammary artery and intercostal arteries.

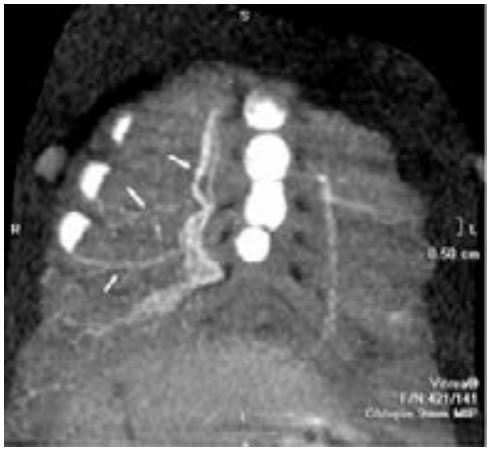


Figure 2: *Frontal CT scan reconstruction shows an unusual collateral vessel (arrows) from the right subclavian artery penetrating the right hilum.*

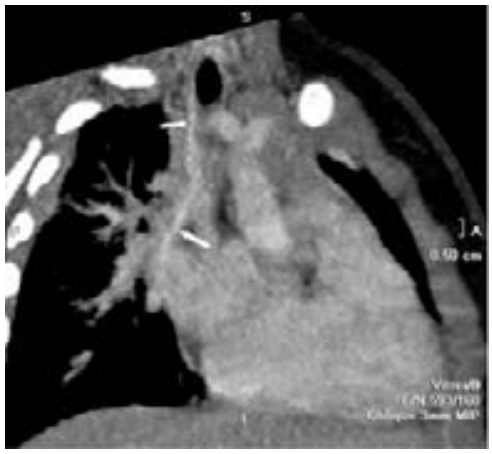
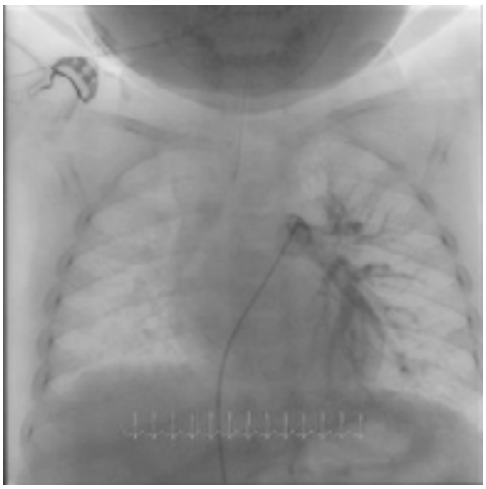


Figure 3: *Catheter angiography showing absence of the right main pulmonary artery.*



study shows that good post-natal lung growth requires adequate pulmonary vascularization.

The youngest case reported in recent literature concerns a 2-day-old child. The baby had a heart murmur on clinical examination, which lead the doctors to perform a cardiac ultrasound with Doppler. On further examination, the doctors discovered the absence of the right pulmonary artery, without any other cardiac defect. No medical or chirurgical intervention was deemed necessary in light of the fact that the malformation was isolated, that the child was asymptomatic and that she had regular growth rates at one week of age. Close monitoring of pressure in the left pulmonary artery was recommended. Three years later, the child is still symptom-free and is growing well (8) dyspnea, chest pain, hemoptysis and recurrent pulmonary infections. As patients may remain asymptomatic or have vague symptoms, the diagnosis of isolated UAPA can be difficult to make in infancy. Indeed, most cases described in literature are adults. Due to the rarity of neonatal presentation, there is no consensus regarding the treatment of this malformation. Case presentation: Herein, the case of a two-day-old term female infant, born after uneventful pregnancy, who required a cardiological assessment for a light murmur, is reported; an echocardiogram demonstrated an isolated unilateral absence of the right pulmonary artery, confirmed by means of magnetic resonance imaging (MRI.

Given the rare cases described in pediatrics, there is no consensus yet on treatment. This will have to be the subject of a multidisciplinary discussion. Any decision should take factors into account such as the child's age, symptoms, growth and cardiopulmonary anatomy (2). A decision to rely on monitoring without immediate intervention can work very well: cardiac ultrasound allows us to detect changes in pulmonary arterial pressure as well as changes in the right ventricular function. Even though the median age of onset of the development of pulmonary arterial hypertension is not known, the literature reports that it occurs early on in life (5). For this reason, close monitoring is even more important. Complications would be discovered early on, allowing for prompt action. The disadvantage of this strategy is that a child may develop comorbidities or insufficient lung reserve and then become inoperable. This should be taken into account in the monitoring program. In all cases, close monitoring is essential.

Conclusion

We report this case to raise awareness of this rare malformation. When faced with a child with hemoptysis, one should consider UAPA in the rare etiologies of hemoptysis. Early recognition of the pathology allows proper management and follow-up, which can prevent the development of associated complications.

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