CASE REPORT

Paediatric case of ductal origin of right pulmonary artery presenting with exertional dyspnoea and mimicking asthma and primary ciliary dyskinesia

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SUMMARY

Ductal origin of pulmonary artery is a rare anomaly that is frequently misdiagnosed. Patients may present with exertional dyspnoea, recurrent respiratory infections and pulmonary hypertension. The presence of pulmonary hypertension can adversely affect clinical outcome in these patients; hence, early identification and intervention is the key to improve survival. A case of a 3-year-old child presenting with exertional dyspnoea is presented in this report. Chest radiograph revealed right-sided pulmonary hypoplasia and mediastinal shift to the right. Pulmonary artery agenesis was suspected when CT of the chest demonstrated right-sided pulmonary artery agenesis. Cardiac catheterisation revealed the correct diagnosis of ductal origin of right pulmonary artery. The most striking feature of this case is that the clinical presentation is mild compared with the findings on imaging.

BACKGROUND

Ductal origin of pulmonary artery (DOPA) is a rare vascular anomaly with an estimated incidence of 1:200 000.¹ It is erroneously known as pulmonary artery agenesis (PAA).² The condition may manifest with hypoplasia of the lung on the same side. The clinical symptoms may be subtle, and this condition can commonly be misdiagnosed. Moreover, frequent respiratory infections and the appearance of dextrocardia/cardiac shift can mislead the clinician to suspect primary ciliary dyskinesia (PCD).

Figure 2 Chest radiograph a year later: persistent findings as noted at initial presentation.

Chest radiograph may not reveal the diagnosis, especially when suspicion is low. This case reports a 3-year-old patient whose imaging findings were out of proportion to the clinical presentation. CT imaging raised the suspicion of right PAA, echocardiogram demonstrated mild pulmonary hypertension (PH) and cardiac catheterisation confirmed the diagnosis of DOPA. Hence, this case underlines the need for a high level of suspicion to diagnose a rare congenital lung anomaly that can mimic a common illness, such as asthma.

Figure 1 Chest radiograph at initial presentation: hypoplasia of right lung and overinflation of left lung with dextrocardia.

Figure 3 Chest CT demonstrates the absence of right pulmonary artery.

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Figure 4 Chest CT demonstrates volume loss of right lung and compensatory hyperinflation of the left lung.

CASE PRESENTATION

A 3-year-old female child, born full term from uncomplicated pregnancy, was referred for persistent right lung volume loss with mediastinal shift towards right on chest X-ray. The parents reported that the child had experienced breathing difficulties from infancy. The patient often required albuterol via nebuliser for wheezing; she was also using inhaled fluticasone with little improvement in her symptoms. Neither parent smoked or described a family history of asthma or atopy. The patient had a respiratory infection secondary to influenza during infancy, but she did not require hospitalisation at that time. She was later admitted to the hospital when she was 2 years old for respiratory syncytial virus pneumonia and was found at that time to have right lung volume loss with mediastinal shift to the right on chest X-ray (figure 1). After recovery, the parents of the child noticed that the patient had increased frequency of dry coughing episodes. They also described respiratory distress on exertion with reduced exercise tolerance. A follow-up chest radiograph at the age of 3 years showed similar findings of right lung volume loss with mediastinal shift, which prompted the referral to pulmonology (figure 2). Prior to that, she had been evaluated at a different facility by a pulmonologist. The appearance of dextrocardia on imaging with a few episodes of otitis media had raised

Figure 5 Nuclear scan showing no perfusion to right lung (anterior view).

Figure 6 Nuclear scan showing no perfusion to right lung (posterior view).

a concern for PCD. However, the work-up was not done for PCD as the patient was lost to follow-up. On presentation to the clinic, the patient had vital signs within normal limits and was in no respiratory distress. She had symmetric chest movement with good air exchange. No wheezing, rhonchi or rales were observed. Cardiovascular examination was normal. No clubbing, cyanosis or oedema was noted.

INVESTIGATIONS

Chest radiographs from initial presentation and after 1 year respectively are shown (figures 1 and 2). Asymmetric lung volumes with mediastinal shift to the right were described on both the films. CT on chest with contrast was performed; absence of the right pulmonary artery (RPA) was observed in addition to hypoplasia involving the entire right lung with compensatory

Figure 7 An aortogram showing an occluded beak of the ductal origin of the right pulmonary artery from the right innominate artery (arrow).

Figure 8 After recanalisation, angioplasty and stenting of the ductus, an angiogram in the right innominate artery shows good flow through the ductus arteriosus (arrow) to the right pulmonary artery. Also seen is the expected immediate reperfusion injury to the right lung.

hyperinflation of the left lung (figures 3 and 4). Left main pulmonary artery (PA) was anteroposteriorly 10.5 and 12.2 mm cranio-caudally. Main PA was 13.7 mm in transverse diameter. The right lung demonstrated mosaic attenuation and mild scattered linear scarring, which was likely related to the chronic volume loss. A shift of the mediastinal contents towards the right secondary to the hypoplastic right lung was observed. A left-sided aortic arch was seen. No other cardiovascular abnormalities were identified. Echocardiogram was subsequently performed. A small atrial septal defect was noted with left to right flow. No discernable RPA could be identified. Concern for PH arose due to noticeable flattening of the interventricular septum. Perfusion scan was recommended to assess the pulmonary blood flow. The study demonstrated no perfusion to the right lung and normal perfusion to left lung (figures 5 and 6). Bronchoscopy with ciliary

Figure 9 Right-sided pulmonary venous wedge angiography via a transeptal approach was used to identify the distal right pulmonary artery (arrow) and facilitate recanalisation of the ductus. No significant aortopulmonary collaterals were noted.

brushings was initially planned to rule out PCD; however, it was deferred as CT chest and then cardiac catheterisation established the diagnoses and hence the benefits did not outweigh the risks.

TREATMENT

Since the patient had PH and exertional dyspnoea, she underwent a cardiac catheterisation at Texas Children's Hospital at Houston. The proximal RPA was absent with an occluded ductal origin. There was a long segment occluded patent ductus arteriosus (PDA) from the right innominate artery, which only had a short proximal stump visible. Pulmonary venous angiography identified distal RPA. No significant aortopulmonary collaterals were identified by angiography. The PDA was recanalised and stent was placed from it to the RPA, which resulted in good flow to the RPA. Mean pulmonary arterial pressure and pulmonary vascular resistance were normal at 17 mm Hg and 1.8 WU, respectively. After her first procedure, repeat echocardiogram revealed the stent to be occluded. Repeat cardiac catheterisation was performed and the stent was recanalised. This was done to facilitate the patient's RPA to grow so that the surgeons could reconnect the PAs at a later date. During the second catheterisation, the patient had a mild PH crisis as her mean left pulmonary arterial pressure increased from 17 to 24 mm Hg and the right ventricular pressure increased from 29 to 41 mm Hg. Hypoxaemia (PaO_2 70 mm Hg on arterial blood gas) was noted during cardiac catheterisation and it improved thereafter (PaO_2 on capillary blood gas was 98 mm Hg). Images from the cardiac catheterisation are demonstrated in figures 7–9. Since the patient was stable prior to the procedure, blood gas was not done. However, in retrospect, it would have been helpful to compare oxygenation before and after the procedure.

OUTCOME AND FOLLOW-UP

The patient is currently on enoxaparin, aspirin and clopidogrel. The plan is to repeat cardiac catheterisation in 3 months.

DISCUSSION

DOPA is a congenital condition in which the right or left PA supplying all bronchopulmonary segments is connected with the distal end of arterial duct (AD) instead of the main PA. The main PA is derived from truncus arteriosus, the proximal PAs are derived from proximal sixth aortic arch and distal PAs are derived from lung buds. Hence, DOPA occurs when there is an involution of the ipsilateral proximal sixth aortic arch with persistent connection between distal PA and ductus arteriosus (distal sixth aortic arch). The intrahilar PA persists despite involution of proximal PA because intrahilar PA branches form independently. Closure of the AD results in reduction in perfusion of the affected lung. Hence, the non-visualisation of the effected PA on imaging studies often leads to the incorrect diagnosis of PAA. Usually, if the ductal origin is on the same side of arch, then it arises from the inferior aspect of aorta at the usual site of ductus arteriosus, and if the ductal origin is on the side contralateral to the arch, the PA arises from innominate artery base, which was noted in the patient reported.³ PAA has been used interchangeably with DOPA in several published reports in the past. A postmortem study was done in 18 cases, which were originally diagnosed as PAA. With the exception of one case of agenesis, all others were confirmed to be DOPA.⁴

Clinical symptoms and course are similar in patients with DOPA and true PAA. The first case of PAA was reported in 1868.⁵ There have been 119 cases of PAA documented since then. However, this number may underestimate the true incidence as 30% of these

were asymptomatic.^{1 6-8} Those with milder agenesis may remain asymptomatic because of compensatory hyperplasia of the existing pulmonary vasculature. A retrospective analysis of 108 cases was reported between 1978 and 2000 ranging from 0.1 to 58 years of age. Most patients had frequent pulmonary infections (37%), dyspnoea (40%) and haemoptysis (20%).⁷ Similar to the presented case, a 4-year-old male child with asthma symptoms and right-sided PAA has been reported.⁹ Symmetric air entry on pulmonary examination was an unexpected finding for the patient, but it could be explained by overinflated left lung being auscultated on the right side.¹⁰ The complication of PH is reported in 19%–44% of patients.^{6 7} Clinically important PH developed only in a minority of patients, usually in those who had large intracardiac shunts.¹⁰ The studies in the patients with isolated unilateral pulmonary artery agenesis (UPAA) have shown that one lung can accept increased blood flow with little or no change in normal pressure. Only a small percentage of these patients have severe PH,¹⁰ likely from increased pulmonary blood flow from collaterals.¹¹ Another study reported 32 patients, 5 of whom died in infancy due to severe heart failure and PH.¹² Patients with isolated unilateral pulmonary artery agenesis (UPAA) who develop PH usually do so at an early age and die from right-sided heart failure. However, if PH does not develop at an early age, it is unlikely that it will develop later.¹² The reported patient had mild PH based on the findings of echocardiogram. Hence, cardiac catheterisation was performed to assess the anatomy of the PA, evaluate for collaterals, accurately identify the severity of PH and determine the need for intervention. The diagnosis of PAA can be made by chest radiograph, echocardiography, CT, MRI, ventilation-perfusion scan and cardiac catheterisation.¹³ Radiologically, displacement of the mediastinum, heart and large vessels towards the affected hemithorax is commonly seen. Hilar arteries can be seen during cardiac catheterisation and pulmonary venous wedge angiography. This is important because revascularisation may improve PH.⁷ It is likely that many of these reports of PAA included patients who actually had DOPA and were not diagnosed in these reports correctly. Unilateral DOPA can occur in an otherwise normal heart and may escape detection. In a series of 45 patients correctly diagnosed as having DOPA, cyanosis (with associated cardiac defects), heart failure, pulmonary infections, failure to thrive, chest pain and feeding difficulties were present. PH was seen in 7 out of the 45 patients at presentation.³ For this patient, chest radiograph was non-diagnostic. However, the abnormal CT of the chest resulted in cardiac catheterisation, which ultimately led to the correct diagnosis of DOPA. This highlights the importance of a multimodal diagnostic approach.

For all patients specifically diagnosed with DOPA, surgical/catheter-based correction by restoring flow to affected PA is recommended with eventual surgical reconnection of the PAs. Failure to treat this condition may result in hypoplasia of the affected PA, ipsilateral development of aortopulmonary collaterals, increased susceptibility to pulmonary infections, pulmonary haemorrhage and scoliosis. Many cases diagnosed with DOPA are never offered surgical options or ultimately undergo pneumonectomy as diagnosis of PAA creates a misunderstanding that the lesion is uncorrectable. Hence, it is very important to do cardiac catheterisation in order to make the correct diagnosis.²

Favourable outcomes are reported in a case series which included 10 patients, median age 2 years with unilateral DOPA. Symptoms were present in six patients. Cardiac catheterisation was performed in all and showed a patent duct or a ductal stump in most patients. Two patients underwent single-stage centralisation. The other eight underwent ductal stenting or a systemic-to-PA shunt as the first stage before centralisation. Two patients with ductal stenting

developed pulmonary oedema. Two patients with a shunt developed early thrombosis requiring reintervention. Nine patients have undergone centralisation. There have been no deaths. Symptoms and lung hypoplasia have improved in all patients.²

Many patients with DOPA remain asymptomatic for a long period of time. However, the development of pulmonary haemorrhage or PH may preclude long-term survival. Hence, recognition of this lesion (distinguishing it from PAA) and its potential treatment is of utmost importance.

Learning points

- Diagnosis of ductal origin of pulmonary artery could be delayed as it is a rare disease, symptoms may be subtle and it can be easily confused with more common respiratory problems like asthma.
- It is important to establish an early diagnosis of ductal origin of pulmonary artery, as the condition can lead to serious consequences like pulmonary hypertension, which may preclude long-term survival. Further, it is a treatable condition since it is distinctly different from true pulmonary artery agenesis.
- Multimodal diagnostic approach is of key importance, and various medical and surgical options are available.

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