Isolated left pulmonary artery sling in a 7-month-old female presenting with wheezes after the age of 3 months

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Abstract

A 7-month-old female infant was completely healthy till the age of 3 months, when she started having persistent wheeze associated with bouts of brassy cough and cessation of breathing. Barium swallow and cardiac echocardiogram were inconclusive. Because steroid burst and steroid inhalers failed to clear her problems she was referred to Queen Rania al-Abdallah hospital for children where flexible bronchoscopy showed slit-like narrowing of the lower trachea with pulsation A subsequent chest CT angiogram showed pulmonary sling causing severe tracheal narrowing. She underwent left pulmonary re-implantation and aortopexy and her symptoms resolved. Flexible bronchoscopy should be done in all children with intractable wheeze.

Introduction

Isolated pulmonary artery sling (PAS) is a very rare form of vascular ring.[1] Unlike other types of vascular ring the PAS is more commonly associated with cardiac and tracheal anomalies. The age of clinical and mostly respiratory manifestation depends on the associated tracheo-bronchial and cardiac anomalies.[1,2] Diagnosis and treatment are often delayed.[2] we report the case of a 7-month-old girl with isolated PAS causing severe tracheal compression.

Case report

A 7-month-old female infant was referred to our clinic with wheezes and brassy cough, which she had suffered since the age of 3 months. Her symptoms were worse during the night with a history of frequent suffocation and cyanosis during sleep. She had frequent visits to the emergency room and had been resuscitated frequently with an adrenaline nebuliser and steroid injection. One month previously she had been started on oral prednisolone 7.5 mg every other day, together with a steroid inhaler, without improvement. Becauses her symptoms persisted she was referred to the respiratory clinic at the Queen Rania hospital for children .

The patient was the result of a full-term normal vaginal delivery with a body weight of 3 kg and no perinatal events of note. She was well till the age of 3 months and received all the vaccinations according to the national programme. She wasbeing breastfed and had no history of allergies. Examination showed an awake infant with good eye contact and cushingoid face. Her respiratory rate was 45 breaths with suprasternal and some subcostal recession. She had expiratory stridor difficult to distinguish from wheeze with frequent attacks of brassy tracheal cough. Oxygen saturation was 92% in room air. The airmovement was symmetrical bilateral with monophonic expiratory wheeze all over the chest. Rest examination was normal. Chest X-ray was unremarkable and complete blood count and electrolytes were normal. Due to the association of wheeze with upper airway tracheal cough and poor response to steroid flexible bronchoscopy was done. It showed slit-like narrowing of the lower third of the trachea with external pulsation and complete obstruction on expiration, as shown in Figure 1. There were no complete tracheal rings. CT angiogram showed pulmonary sling with severe compression of the trachea as shown in Figures 2 and 3. She underwent re-implantation of the left pulmonary artery and aortopexy and extubated on the second day. Her symptoms on follow-up disappeared completely and inhalers were no longer required.

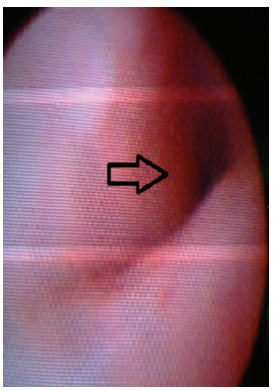

Figure 1 bronchoscopy showing severe narrowing of the lower third of the trachea (black arrow)

Discussion

The striking feature in this case is that the patient was completely healthy for the first 3 months of her life until the appearance of the expiratory wheeze with recurrent attacks of cessation of breathing and tracheal brassy cough. The age of the appearance of the clinical manifestation in our case is the time of the lowest nadir

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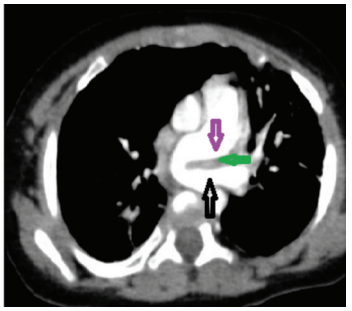

Figure 2 CT-angio showed the left artery pulmonary artery (black arrow) behind the trachea causing severe compression (green arrow). Right pulmonary artery shown in purple arrow.

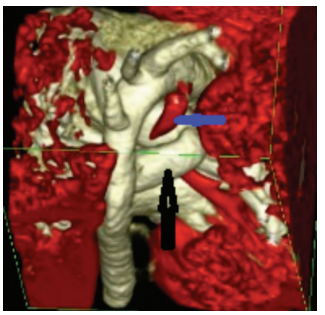

Figure 3 The 3-dimensional CT scan with the trachea compressed (purple arrow) and the pulmonary sling (black arrow)

of pulmonary pressure. This could be explained by the fact that the growth and dilatation of pulmonary artery during this age is disproportionately more than that of the trachea, thus compromising it. Our case also emphasises the necessity of doing flexible bronchoscopy in children with wheeze not responding to anti-asthma treatment, particularly when associated with tracheal brassy cough.

There is no report yet emphasising the exact age of onset of the clinical manifestation in isolated PAS) In fact the initial clinical manifestation varies depending on the associated anomalies such as structural heart disease and tracheal anomalies as complete tracheal ring.[2,3]

PAS is a rare congenital anomaly. [3] In contrast to other types of vascular ring it is more likely to have associated anomalies; mostly tracheal stenosis and cardiac structure-anomalies.[4] The respiratory symptoms (chronic stridor, cough, and wheeze) in more than 90% of the patients are due to tracheal stenosis (TS) and usually appear in the first year of life.[2,4,5] Studying 18 patients with PAS, Chen and colleagues found associated TS in all of them, combined right tracheal bronchus, underdeveloped right lung, persistent left superior vena cava, and left patent ductus arteriosus in 22%,22%, 22%, and 29%, respectively.[6] Ma and colleagues found one case with PAS among seven patients with vascular ring, presenting with respiratory distress. His age was 5 months with main complaints of recurrent cough, stridor; and wheeze; this patient did not undergo surgery.[7]

Because the symptoms and signs are usually non-specific, diagnosis of PAS is usually delayed and frequently as in our case misdiagnosed as asthma. [3,5–7] Furthermore patients with PAS may be asymptomatic for a long time, but failure to recognise those with airway compromise and anomalies can lead to potential mortality and morbidity during exercise and acute infections precipitating sudden airway obstruction.[6] In a large study done on school-aged children in Taiwan, Yu and colleagues found a prevalence of PAS as 59 per million using two-dimensional echocardiography.[2]

Early surgery can improve pulmonary function performance.[6] The surgery depends on the presence of associated anomalies.[6–8]All patients with PAS reported by Oshima and colelagues required left pulmonary re-implantation (LPR); 90% required tracheoplasty and 32% required repair of cardiac anomalies.[8] Our patient required aortopixy in addition to LPR and was extubated on the second day of surgery.

We conclude that patients with intractable wheeze should undergo flexible bronchoscopy to assess the airway and guide proper management.

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