Swyer-James syndrome in a 7-year-old female

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Abstract

Swyer-James syndrome is a rare syndrome that occurs as a result of repeated bronchiolitis and pneumonitis in childhood. Most cases are asymptomatic, and subsequent diagnosis may not occur until adulthood. We present the case of a 7-year-old female with Swyer-James syndrome, which was initially diagnosed and treated as asthma. The patient developed respiratory distress and atelectasis which were treated with biphasic cuirass ventilation. This case suggests that Swyer-James syndrome should be a concern in patients with chronic cough and wheezing, and highlights the importance of taking a careful history and appropriate radiological investigations for diagnosis. Once Swyer-James syndrome is diagnosed, prophylaxis and appropriate management of respiratory infections becomes important.

Case Report

The female patient, now aged seven years, was born as twin after 34 weeks gestation with a birth weight of 2.286 g. Past medical history was significant for pneumonia twice, at 3- and 5- years of age. The pneumonia at 5 years of age had been thought to be due to Chlamydia pneumoniae infection, as IgM antibodies to C. pneumoniae were elevated. She had taken a leukotriene receptor antagonist (LTRA) for asthma or other respiratory pathologies, in addition to a LTRA. However, these treatments were not effective at resolving, or improving the presented symptoms. Consequently, the patient was treated for a suspected asthma attack. She was therefore administered intravenous prednisolone, inhaled P2-agonist, and supplemental oxygen in addition to a LTRA. However, these treatments were not effective at resolving, or improving the presented symptoms. Consequently, the patient was treated with biphasic cuirass ventilation. This case suggests that Swyer-James syndrome is a rare syndrome that occurs as a result of repeated bronchiolitis and pneumonitis in childhood. Most cases are asymptomatic, and subsequent diagnosis may not occur until adulthood. We present the case of a 7-year-old female with Swyer-James syndrome, which was initially diagnosed and treated as asthma. The patient developed respiratory distress and atelectasis which were treated with biphasic cuirass ventilation. This case suggests that Swyer-James syndrome should be a concern in patients with chronic cough and wheezing, and highlights the importance of taking a careful history and appropriate radiological investigations for diagnosis. Once Swyer-James syndrome is diagnosed, prophylaxis and appropriate management of respiratory infections becomes important.

Discussion

Swyer-James syndrome was first reported in 1953. It is a rare syndrome in which an unilateral hyperlucent lung develops due to bronchiolitis obliterans caused by viral infections such as adenovirus and/or measles.1,2 Bronchiolitis obliterans induces inflammation and fibrosis of the respiratory walls, leading to the narrowing of the bronchiolar lumen. Fibrosis of intra-alveolar septa provokes obstruction of the pulmonary capillary bed, and decreased blood flow to the major pulmonary artery segments, resulting in a hypoplastic pulmonary artery. Consequently, the typical radiological findings are unilateral hyperlucent lung, hypoplasia of pulmonary artery and pulmonary hypoperfusion. Patients may complain of productive cough, shortness of breath and dyspnea on exertion. However, most patients are asymptomatic, thus diagnosis is usually unexpected, and confirmed on screening chest radiographs in adults. There is no radical cure for Swyer-James syndrome. In a case complicated with asthma or other respiratory pathologies, for which treatments exist, effective therapeutic management may lessen the severity of symptoms evoked by Swyer-James syndrome. As such, prophylaxis, and appropriate management of respiratory infections is important. Although the outlook of Swyer-James syndrome is excellent in many cases, a few cases require surgical management, such as pneumonectomy, due to persistent and severe lung infection.1 An anamnestic of recurrent bacterial infections of the affected lung is an indication for surgery.3 A female patient in the present case study had productive cough and wheezing, which was likely due to the narrowing of the bronchiolar lumen caused by Swyer-James syndrome. Lung function testing showed decreased V25/HT, accompanied by normal FVC and FEV1/FVC. These data suggest V25/HT could be a sensitive marker of peripheral airway closure in the early stages of

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obstructive airway disease. At first, the patient received the diagnosis of asthma. There might be a possibility that asthma was complicated by Swyer-James syndrome. However, treatment for asthma, including intravenous administration of prednisolone and inhalation of β2-agonist, were not effective. Thus, the main pathogenesis of this episode is thought to be due to airway obstruction due to Swyer-James syndrome and atelectasis. Narrowing of the bronchiolar lumen caused atelectasis, which BCV dramatically ameliorated.

Bronchoscopy is reported to be useful for removal of mucous secretions. However, it is an invasive procedure, and should be avoided, if possible, especially in children. BCV provides an alternative option to bronchoscopy. Swyer-James syndrome should be a concern in patients with chronic cough and wheezing, and radiological findings should carefully be examined. Once Swyer-James syndrome is diagnosed prophylaxis, and appropriate management of respiratory infections becomes important.

Conclusions

We report a case of Swyer-James syndrome in a 7-year-old female, diagnosed by chest X-ray, CT angiography, lung perfusion scintigraphy and pulmonary function testing. V25/HT in pulmonary function testing could be a sensitive marker of airway closure, and BCV is an effective treatment for atelectasis in Swyer-James syndrome. There might be some individuals with Swyer-James syndrome among patients diagnosed with asthma. Thus a careful examination of chest X-ray radiograms and pulmonary function testing is needed.

References

1. Swyer PR, James GCW. A case of unilateral pulmonary emphysema. Thorax 1953;8:133-6.
2. Macpherson RI, Cumming GR, Chernick V. Unilateral hyperlucent lung: a complication of viral pneumonia. J Can Ass Radiol 1969;20:225-31.
3. Spigelblatt L, Rosenfeld R. Hyperlucent lung: long term complication of adenovirus type 7 pneumonia. J Can Med Ass 1983;128:47-9.
4. Fregonese L, Girosi D, Battistini E, et al. Clinical, physiologic, and roentgenographic changes after pneumonectomy in a boy with Macleod/Swyer-James syndrome and bronchiectasis. Pediatr Pulmonol 2002;34:412-6.
5. Vishnevsky A, Nikoladze GD. New approach to the surgical treatment of Swyer-James/MacLeod syndrome. Ann Thorac Surg 1990;50:103-4.
6. Preciado D, Vergheze S, Choi S. Aggressive bronchoscopic management of plastic bronchitis. Int J Pediatr Otorhinolaryngol 2010;74:820-2.