Plastic bronchitis with life-threatening respiratory failure: a case report

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Abstract
Plastic bronchitis is characterised by obstruction of the airway by bronchial casts. Depending on the completeness of blockage, symptoms could range from chronic cough to acute respiratory failure. It is classified into type 1 and type 2. Bronchial cast in type 1 consists of inflammatory cells. Type 1 cast is uncommon though the diagnosis should be entertained in a difficult to treat wheezing patient. We presented a case, a 16-month-old girl with type 1 plastic bronchitis who presented with cough and dyspnoea with rapidly progressive respiratory failure. Its pathogenesis and treatment were discussed.

Keywords: Bronchitis, child, respiratory failure

Introduction
Plastic bronchitis is characterised by obstruction of the airway by bronchial casts. Depending on the completeness of blockage, symptoms could range from chronic cough to acute respiratory failure. It is classified into type 1 and type 2. Bronchial cast in type 1 consists of inflammatory cells. Type 1 cast is uncommon though the diagnosis should be entertained in a difficult to treat wheezing patient. We presented a case, a 16-month-old girl with type 1 plastic bronchitis who presented with cough and dyspnoea with rapidly progressive respiratory failure.

Case presentation
A 16-month-old girl with Cri-du-chat syndrome and global developmental delay was admitted to hospital for cough and dyspnoea. She had no known drug allergy. She had wet cough for 3 weeks. There was no fever. She visited private doctor 4 times and was told to have a clinical diagnosis of "allergic airway", though no obvious wheeze was noted by parents in the previous illnesses. No steroid or inhaled bronchodilator was prescribed before. She was noted to have increasing dyspnoea with wheeze on the day of admission. There was no history of choking and foreign body inhalation. She had no travel history nor sick contact. Her mother had allergic rhinitis. There was no personal history of atopy including eczema and allergic rhinitis. There was no smoker, pet, carpet and incense burning at home.

On admission, she was in significant respiratory distress with respiratory rate 48 per minute, suprasternal and substernal insucking. Her SpO₂ was 79% in room air. Heart rate was 170/min. Chest auscultation showed poor air entry bilaterally with diffuse wheeze. Her SpO₂ was 88% with 100% oxygen via a non-rebreathing mask. She was managed as severe bronchiolitis. Trial of salbutamol 10 puffs, ipratropium bromide 4 puffs and nebulized 1:1000 adrenaline and hypertonic saline were given, but there was no improvement. ABG showed respiratory acidosis (pH 7.176 pCO₂ 8.33 HCO₃ 22.6 BE -6.3). Bilateral chest hyperinflation was seen in chest X-ray (CXR) (Figure 1). She was transferred to paediatric intensive care unit for further management.

She was put on continuous positive airway pressure (CPAP) 15 cmH₂O and FiO₂ 65% was needed via oral-nasal mask to keep SpO₂>90%. In view of the life threatening wheezy attack, a regime of status asthmaticus treatment was tried and antibiotic was given. Intravenous magnesium sulphate 40 mg/kg, intravenous methylprednisolone, continuous nebulized salbutamol, augmentin, azithromycin and montelukast were given. Her breath sound was improved slightly after CPAP. There was still poor breath sound over left chest. ABG worsened with mixed respiratory and metabolic...
acidsis (pH 7.001 pCO\textsubscript{2} 14.57 pO\textsubscript{2} 10.30 HCO\textsubscript{3} 26.4 BE -6.9) and PaO\textsubscript{2}/FiO\textsubscript{2} ratio was only 123. She was intubated with microcuff size 3.5 mm endotracheal tube, guided by bronchoscopy (Figure 2). A dose of fentanyl was used for sedation. No muscle relaxant was needed.

She was put on mechanical ventilation with PRVC mode (Tidal volume 6 ml/kg Rate 25 PEEP 7 Ti 0.4). However there was no improvement with type 2 respiratory failure (pH 6.992 pCO\textsubscript{2} 13.44, pO\textsubscript{2} 12.80, BE -9.1). Oxygenation index was 10 and PaO\textsubscript{2}/FiO\textsubscript{2} ratio was 160. Examination showed poor breath sound over left chest especially expiration phase.

Emergency bronchoscopy was performed and showed (1) A plug obstructing left upper bronchus (Figure 3). (2) Thick mucus lining left lower bronchus, obstructing <50% of bronchus. (3) Normal trachea, main right bronchus, RUL, RML, RLL bronchi. Removal of the plug was failed at that time since patient’s condition was not stable.

She was then put on high frequency oscillation ventilation (Max amplitude 7 MAP 18 Freq 8Hz). Intravenous sulbutamol was also started. Blood gas was gradually improved (pH > 7.2 and PaCO\textsubscript{2} 7-8 kPa). FiO\textsubscript{2} decreased from 60 to 50%.

Bronchoscopy with bronchoalveolar lavage (BAL) was performed next morning. It showed thick mucus plug over right upper lobe, which was cleared by normal saline lavage. There was a cast over left upper bronchus which could not be removed by 10 ml normal saline BAL. Another aliquot of BAL of 10 ml solution of 8.4% NaHCO\textsubscript{3}+Normal saline at a ratio of 1 to 4 was given with successful removal of the cast. A whitish rubbery mass was carried to trachea by scope which could only be removed by repeated suction via a 12F suction catheter. There was thin mucus over left lower bronchus and was removed by suction. The rest of bronchi were patent. BAL showed raised eosinophils (15%) and pus
cells. There was presence of Charcot-Layden crystals in the cast. There was no intracellular organism. BAL culture was negative. A diagnosis of plastic bronchitis was made.

After clearance of the cast, her ventilator setting could be changed from high-frequency to conventional ventilation with PRVC mode (TV 6 ml/kg, RR 30, PEEP 8, I:E 1:40). Her blood gas further improved (PaCO\textsubscript{2} 8.2 \rightarrow 7.39 \rightarrow 6.95 \rightarrow 4.57). Acidosis was resolved. Oxygenation index decreased from 14 to 7.8. Patient was getting more awake with own respiratory effort. In view of the improved lung compliance, she was extubated to non-invasive ventilation (pressure 17/13 cmH\textsubscript{2}O). We could step down NIV to CPAP 5 cmH\textsubscript{2}O followed by heated humidified high flow oxygen 8L/min. Further workup included the following: Negative NPA respiratory virus, mycoplasma PCR and blood culture. Viral culture was negative. Her IgE level was elevated. Both sputum and nasal smear collected after starting steroid were negative. Aspergillus antibodies and skin prick test were also negative.

For the treatment of plastic bronchitis, we gave the patient five days of systemic steroid (two days of intravenous methylprednisolone and 3 days of oral prednisolone) followed by inhaled corticosteroid (fluticasone 250 mcg daily). She was also put on montelukast. After extubation, chest physiotherapy and in-exsufflator therapy were commenced for sputum clearance.

Patient could finally be weaned off high flow oxygen to room air after 17 days of hospital stay. She was discharged on Day 23. In view of the life threatening wheezy attack and elevated IgE, she was put on inhaled corticosteroid and montelukas on discharge.

Discussion

Wheezy attack is a common presentation in respiratory emergency. Common causes included asthma, acute bronchiolitis, foreign body aspiration and croup etc. In a case with acute and severe wheezy attack not nonresponsive to conventional treatment including bronchodilator and nebulizing adrenaline, rare condition including plastic bronchitis should also be considered.

Plastic bronchitis is characterised by mechanical obstruction of the bronchial tree by casts. These casts are of cohesive and rubbery consistency and conformed to the shape of tracheobronchial tree. Occasionally patients cough out casts of variable size from a small segment to the whole tree-bud appearance, which can be an unpleasant and unforgettable experience. They may sometimes mistake the cast as other substances, most commonly described as chicken meat. Degree of dyspnoea depends on the completeness of bronchial obstruction. When the cast completely occludes the bronchi and patient had difficulty in expectoration, life-threatening acute respiratory failure could have occurred.\textsuperscript{1,2}

Physical examination reveals poor breath sounds and wheeze if airway is partially blocked; or silent chest if completely obstructed. CXR may show atelectasis or infiltrates predominantly located in the lower lobes, and contralateral chest hyperinflation. CT thorax is not required for diagnosis of plastic bronchitis, but it can help demonstrate a level of obstruction especially when the diagnosis was not clear.\textsuperscript{3} In our case, the CXR did not correspond to the clinical severity. We postulated that it is because the case presented acutely with thick mucus and mucus plug obstructing the airway and the lung was not yet collapsed.

Bronchoscopy is important in three aspects. First, it is a diagnostic tool by direct visualisation of the airway and the cast. Second, therapeutically, it allows mechanical clearance of the cast. Moreover, it increases the chance of successful intubation in emergency conditions. Failure to intubate is not uncommonly encountered in intensive care unit especially in critical patients who are unstable, uncooperative and with no prior airway assessment.\textsuperscript{4} The use of bronchoscopic intubation, as shown in our case, is useful for evaluation of upper airway, insertion of endotracheal tube, verification of its placement and position. In contrast to rapid sequence induction in conventional intubation, intubation under bronchoscopy can be achieved without paralyzing the patient by muscle relaxant. Patient can breathe spontaneously, and reduces chance of respiratory compromise should the procedure fail.\textsuperscript{5}

Plastic bronchitis is categorised into two groups according to their histological appearance.\textsuperscript{6} Type 1 casts consisted of inflammatory cells. Histology showed fibrin with eosinophilic inflammatory infiltrates. They were often associated with bronchial diseases, for example, asthma, bronchiectasis, cystic fibrosis and allergic bronchopulmonary aspergillosis.\textsuperscript{7} It was proposed that the underlying inflammatory process promotes hypersecretion of airway mucus, subsequently, cast...
formation. These casts tend to diffusely involve the lung, and presents more acutely than chronic. It is less commonly reported than type 2 plastic bronchitis.

Type 2 casts are acellular casts. They composed mainly of mucin with no inflammatory cell. Associated conditions included cyanotic congenital heart disease, disorders of lymphatics such as lymphangiomatosis, and idiopathic plastic bronchitis. For those of cardiac causes, patient usually suffered from plastic bronchitis after corrective surgery e.g. Fontan procedure, Blalock-Taussig shunt. It is believed that with diversion of systemic blood flow into the pulmonary circulation, there will be an alteration of lymphatic drainage, resulted in endobronchial lymphatic leakage. Type 2 plastic bronchitis tends to present chronically or recurrently, and carries a poorer prognosis than type 1 casts. One of the suggested reasons for this poor prognosis is the limited cardiopulmonary reserve of patients.

Our reported patient presented acutely with left main bronchus obstruction. BAL demonstrated predominant inflammatory cells with eosinophils and Charcot-Layden crystals, elongated spindle shaped crystal resulting from degradation of eosinophil. These are typical of type 1 inflammatory plastic bronchitis. We tried to explore its association with bronchial disease, and the only positive finding is elevated IgE levels. We propose that our patient has underlying allergic inflammation of the airway, leading to increased secretion of thick mucus which could not be cleared by expectoration. With time, these became obstructing casts and caused respiratory failure.

Treatment of plastic bronchitis could be divided into acute and chronic. Acute management aimed at removal of the casts. This could be done mechanically (by bronchoscopy or chest physiotherapy or pharmacologically by mucolytics such as inhaled acetylcysteine, recombinant human DNAase and antifibrin therapy). Inhaled acetylcysteine was unavailable in this hospital and therefore was not given in this case. Chronic treatment targets at the underlying diseases. To tackle with type 1 inflammatory plastic bronchitis, we try to address the underlying hypersecretory process by the use of anti-inflammatory agents including corticosteroid (systemic / inhaled). Low dose azithromycin is also suggested because of its immunomodulatory properties. For type 2 acellular plastic bronchitis, it has been suggested that surgery for thoracic duct ligation may be helpful.

To conclude, plastic bronchitis should be entertained in a case of respiratory distress and wheeze refractory to conventional treatment. In such cases, early bronchoscopy for diagnosis and cast removal would be important to improve clinical outcome.

References