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Case Series

A Case Series of Plastic Bronchitis: Uncommon Cause of Respiratory Distress in Children

Gawahir Mukhtar[1]*, Wasan Almazyad[1], Safa Eltahir[1], Khalid Al-mobaireek[2] and Wadha Alotaibi[3]

[1]*Pediatric Pulmonary Division, King Fahad Medical City (KFMC), KSA*

[2]*Pediatric Pulmonary Division, King Khalid Hospital/ King Saud University, KSA*

[3]*Pediatric Pulmonary and Sleep Physician, Head of the Pulmonary Unit, KFMC, KSA*

***Corresponding Author:** Gawahir Mukhtar, Pediatric Pulmonary division, King Fahad Medical City (KFMC), KSA.

Received: August 25, 2021; **Published:** August 30, 2021

Abstract

Background: Plastic bronchitis (PB), which is a rare pulmonary disease, characterized by the formation of endobronchial casts of undefined etiology. The pathogenesis is still not fully understood, however It is characterized by the presence of large, thick, mucofibrinous plugs filling the broncho-pulmonary tree, leading to severe respiratory distress.

Case-Series: In this article we reported 4 cases of plastic bronchitis with common manifestations (respiratory distress) for uncommon disorder, as a first serial cases from Middle East Area according to our Knowledge. The gold standard for the diagnosis of PB is the detection of bronchial dendritic casts with flexible bronchoscopy. These casts were detected in all our case with flexible bronchoscopy except in the case number 3 his bronchoacopy showed clear airways, however he expectorated a lot of casts during his admission.

Conclusion: In patients with persistent atelectasis or recurrent respiratory distress, PB should be considered as a differential diagnosis. Flexible bronchoscopy should be performed early to confirmation and clearance of airway obstructions.

Keywords: *Plastic Bronchitis; Respiratory Distress; Children; Case Series; Saudi Arabia*

Introduction

Plastic Bronchitis (PB) is a rare pulmonary disease, characterized by formation of large rubbery gelatinous like casts that fill the tracheobronchial tree. It was first described by Galen as expectorated veins and arteries [1].

The pathogenesis is still not fully understood, however It is characterized by the presence of large, thick, mucofibrinous plugs filling the broncho-pulmonary tree, leading to severe respiratory distress. While commonly seen among children, few cases have been described among adults. There is a hypothesis of cast formation due to lymphatic drainage defects leading to accumulation of lymph in bronchial airways [2].

Plastic bronchitis can cause either partial or complete obstruction of airways which commonly manifest as wheezing. Other presentations like, coughing up bronchial shape casts, shortness of breath, and fever. Radiography might show atelectasis of affected segment and hyperinflation [3-6].

There are two types of Plastic bronchitis as proposed by Seer et al; Type I (inflammatory) which are mainly inflammatory infiltrates consist of fibrin, eosinophils and Charcot–Leiden crystals, and type II (non inflammatory) which are mucin casts mainly seen in cardiac structural disorders [7].

Several associations have been observed with plastic bronchitis including; congenital heart disease, Post Fontan procedure, asthma, bronchopulmonary aspergillosis, sickle cell acute chest syndrome, and lower airway infections including adenovirus and mycoplasma organisms. Also, some of reported syndromes such as Turner and Noonan syndromes [3-6,8-10].

Plastic bronchitis can lead to rapid fatal outcomes in case of complete obstruction of larger airway segments and respiratory failure. The best diagnostic and therapeutic approach is direct visualization and removal by Bronchoscopy (flexible and rigid) [9]. Other line of treatment include, fibrinolytics such as aerosolized heparin, urokinase, and tissue plasminogen activator. Also, adjunct therapies like mucolytics (dornase alpha) and macrolides as anti-inflammatory agents showed some benefit in some cases [10-12].

Here, we reported case series of plastic bronchitis from Saudi Arabia, with common manifestations (respiratory distress) for uncommon disorder. This is a first serial cases from Middle East Area according to our Knowledge.

Case Series

Case 1

A-3 years old girl, who was previously healthy and duly vaccinated to age, presented with history of chronic cough for the last 5 months, which was non productive, no diurnal variation or clear triggering factors, the cough associated with mild intermittent dyspnea. During this period, she received multiple courses of antibiotics and asthma medications as labeled asthmatic. Her symptoms were getting worse with time. In the last 5 days prior to admission she developed fever with increase work of breathing and left side chest pain. There was no history of contact to sick person or recent travelling, no family history of atopy or chronic lung diseases.

Physical examination revealed well thriving female child with mild respiratory distress and markedly diminished chest movement and breath sound on left side. The rest of examination were unremarkable. Basic investigations revealed leukocytosis, mainly neutrophilia and raised inflammatory markers.

The diagnosis of foreign body in left main bronchus with complete atelectasis of left lung was made based on radiological images. Her chest x ray (Figure 1) showed cut off sign of left main bronchus with complete atelectasis of left lung, which may suggest foreign body. CT scan chest (Figure 2) showed the same findings. The patient underwent rigid bronchoscopy by ENT Surgeon and reported there was cheesy like material occluded the left main bronchus, which they could not remove completely. Thus, the patient underwent both fixable and rigid bronchoscopy, a thick whitish dense plug in the lingula and left basal segment branch, which was difficult to be removed completely, so dornase alfa was installed and she was subjected to aggressive chest physiotherapy and nebulized hypertonic saline solution to open residual atelectasis of left lower lobe (LLL). The diagnosis of plastic bronchitis was confirmed by tissues and bronchoalveolar lavage (BAL) pathology which reported that; mucus, fibrin (Image slide 1), inflammatory exudate with ghost of inflammatory cells (Image slide 2) and charcot crystals (Image slide 3). No evidence of granuloma or malignancy. Special fungal stain and oil red stain were negative. Patient received iv antibiotics based on sensitivity result from bronchoalveolar lavage. The patient progressed satisfactorily with improvement of cough and expectoration. She was discharged with combined treatment, nebulized medications.

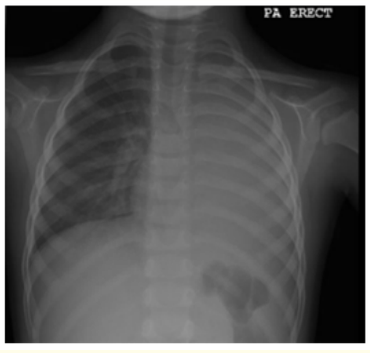

Figure 1: *Chest X-ray.*

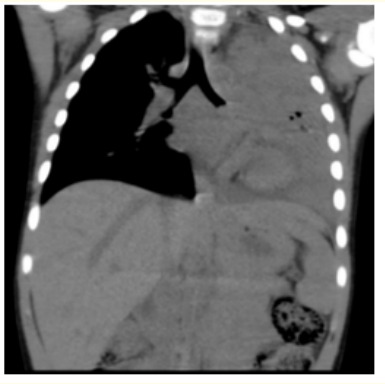

Figure 2: *CT scan chest coronal view.*

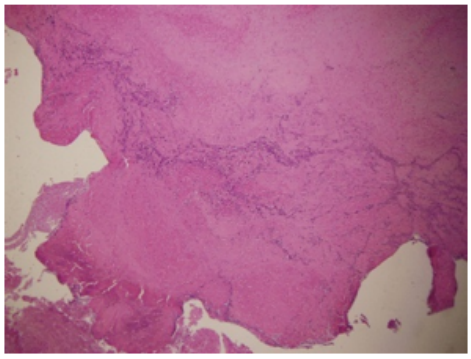

Image (Slide) 1: *Plastic H&E 10 Showing fibrin.*

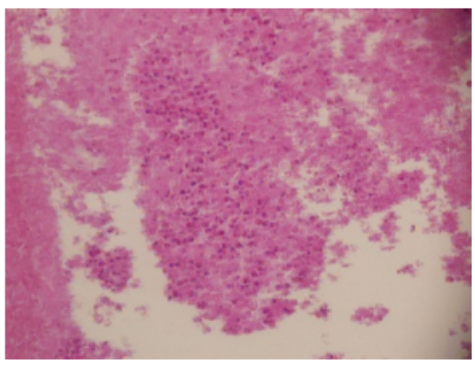

Image (Slide) 2: *Eosinophilic infiltrate.*

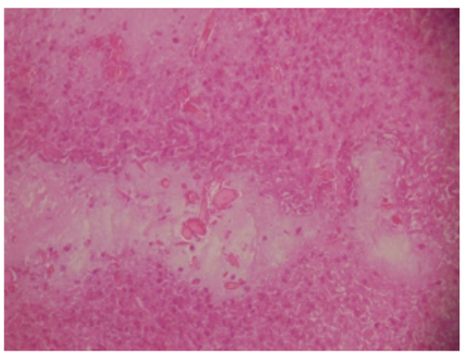

Image (Slide) 3: *Showing charcoat crystals (arrows).*

Case 2

9-year-old girl, previously healthy apart from mild shortness of breath from time to time when the house is being painted. She never used asthma medication.

One month prior to presentation, she suddenly developed shortness of breath and apnea, she went to a local hospital in their area where she was admitted for a total of 3 weeks. Initially 7 days at the ward. The patient was deteriorating so she was transferred to PICU and electively intubated for 7 days, then transferred to the ward again for 7 days. The patient was discharged against medical advice (DAMA) and came to our hospital through emergency department. She stayed for a few hours then transferred to ICU in a severe respiratory distress. She was intubated for 3 days. The parents gave a history of recurrent expectoration of a material which was hard and yellow in color.

On clinical examination, she was in respiratory distress. No dysmorphic features or clubbing were noted. Her growth parameters were normal and her vital signs showed tachypnea and hypoxemia, and she had reduced air entry mainly on the left side and no added sounds up on auscultation. Her other systemic examination including cardiovascular examination was unremarkable. The patient initial basic laboratory workup showed leukocytosis mainly neutrophilia and increased IgE level (901 Ku/L). Chest x-ray (Figure 3) showed minimal left basal atelectasis, peribronchial thickening is noted. HRCT showed left lower lobe subsegmental consolidation, and evidence of small airway disease (Figure 4). Bronchoscopy was done for the patient, and it revealed a thick secretion mainly in the right upper lobe, right lower lobe and left lower lobe. There was a cast which was seen in the superior segment of the right lower lobe. A few casts were expectorated during suctions, which were sent to histopathology. The pathology report came to confirm the diagnosis of plastic bronchitis. The patient was put on Dornase alfa plus chest physiotherapy. Tazocin and Vancomycin were prescribed based on the microbiology result for the bronchoalveolar lavage (BAL). Patient showed complete recovery and was discharged home in good condition.

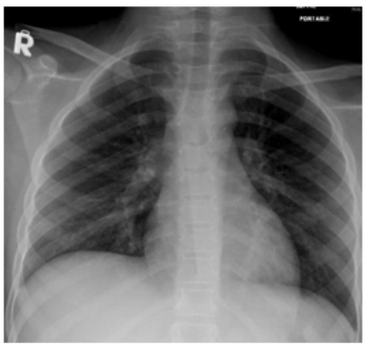

Figure 3: *CXR showed left basal atelectasis consolidation.*

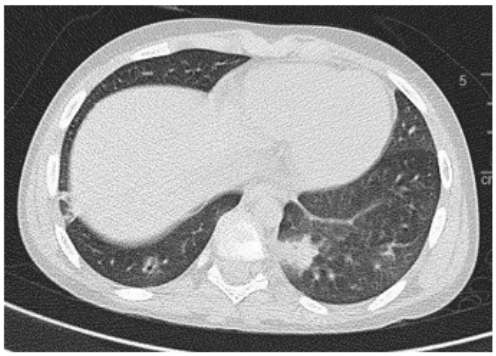

Figure 4: *HRCT showed LLL subsegmental.*

Case 3

6 years old boy case of Complexed Congenital Heart Disease: in the picture of (dextrocardia, double outlet Right ventricle, pulmonary atresia, malposition aorta, retro aortic innominate vein, bilateral pulmonary artery stenosis). Post multiple procedures include Glenn and Fontan procedures. He was labelled also as having asthma, and he was on regular asthma medication including Salbutamol, Fluticasone 125 mcg and Muntelukast. Admitted to pulmonary ward with history of Shortness of breath and productive cough for around 3 weeks, Started shortly to have thick sputum then patient expectorated a yellow material with his cough at the day of admission to hospital. Patient had no fever, and there was no symptom related to other system. He was born full term, normal vaginal delivery, and was diagnosed antenatally with the above mentioned complex congenital herat disease and was treated initially with prostaglandin and later he underwent the above mentioned procedures.

On clinical examination, he was in mild respiratory distress. No dysmorphic features or clubbing were noted. His growth parameters were normal and his vital signs showed mil tachypnea, and he had reduced air entry bilaterally, but no added sounds up on auscultation. His other systemic examination including cardiovascular examination was unremarkable The patient initial basic laboratory workup showed leukocytosis mainly neutrophilia. Chest x-ray (Figure 5) showed Multiple sternotomy wires and surgical clips noted in the mediastinum. There was sign of the lower respiratory tract inflammatory process. No pneumothorax or pleural effusion. The cardiomediastinal silhouette was unremarkable.

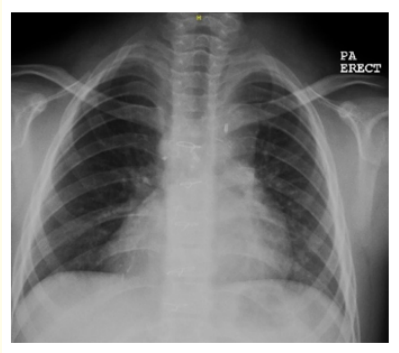

Figure 5: *CXR showed Multiple sternotomy wires and surgical clips an sign of the lower respiratory tract inflammatory process.*

The patient persisted with cough, mobilization of pulmonary secretions, moderate dyspnea and expectoration of casts of gelatinous material for 4 days (Figure 6).

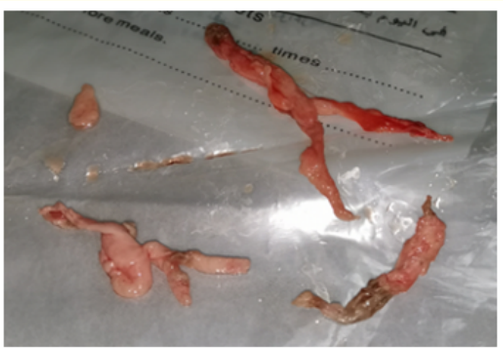

Figure 6: *Expecturated casts form the patient during his admission.*

The diagnosis of Plastic Bronchitis was established based on the history and clinical findings. The patient underwent Flexible Bronchoscopy which showed thick secreations but no casts. Rigid bronchoscopy was not needed. His bronchoalveolar lavage (BAL) culture came back negative. His Pharyngeal C/S showed Staph Aureus. He was started on Cefuroxime for total of 5 days and discharged home in good condition.

Case 4

A 3- year – old boy previously healthy presented with history of fever, cough and shortness of breath of 1-week duration associated with left lower chest pain progressive and increasing with effort and preventing him from sleep and partially resolved by paracetamol. He had already been diagnosed as having pneumonia and was already taking antibiotics, but had returned to the hospital because of worsening of the respiratory distress. He was admitted and received IV antibiotics for 5 days without significant improvement. The mother gave history of expectorated whitish material few days prior to his admission. No GI symptoms or any symptoms related to other systems. He was outcome of uneventful pregnancy and delivery, no NICU admission. Fully vaccinated without complications. On clinical examination, the patient has normal growth parameters, no dysmorphic features. He was in mild respiratory distress but no cyanosis. His vital signs showed tachypnea, and he had reduced air entry on the left upper and middle zone, and there was no wheezes or crackles. other systemic examination revealed no abnormalities.

The patient initial basic laboratory workup showed high WBC count mainly neutrophils. ESR and C-reactive protein (CRP) were normal. A chest X- ray (Figure 7) showed persistent left upper lobe collapse (Figure 8). CT chest with iv contrast revealed bulging soft tissue density within the left upper lobe bronchus origin is suspicious for mass lesion. The differential of bronchial tumors such as papillomatosis needs to be considered particularly in presence of left lower lobe bronchial dilatation. The other differential includes mucous plugging or fungal process.

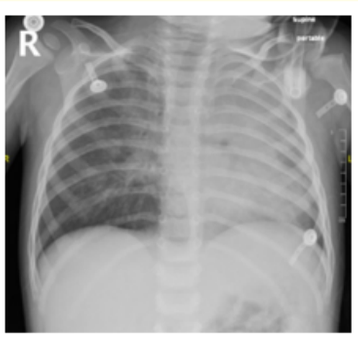

Figure 7: *CXR showed persistent left upper lobe collapse.*

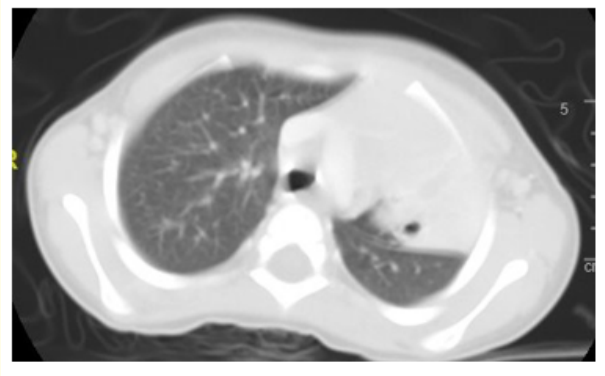

Figure 8: *CT chest shows bulging soft tissue density noted within the left upper lobe.*

The diagnosis of plastic bronchitis was made based on clinical history and radiology findings, he underwent Flexible Bronchoscopy which showed a thick secretion mainly in the left upper lobe, lingual and right lower lobe. There was a cast which was seen in the left upper lobe. A few casts were expectorated during suctions, which were sent to histopathology. The pathology report came to confirm the diagnosis of plastic bronchitis. The patient was put on Dornase alfa, nebulized hypertonic saline solution plus aggressive chest physiotherapy. He showed complete recovery and was discharged home in good condition.

Discussion

Madsen et al. propose a model in which the inflammatory processes activated in asthma would cause dysregulation of the formation of mucus that, together with reduced mucociliary clearance and infiltration of the airway by neutrophils and eosinophils, would explain the pathophysiology of PB [13]. Atelectasis is commonly observed on imaging tests, providing a differential diagnosis with foreign-body bronchoaspiration. All our cases showed significant atelectasis in their images.

The gold standard for the diagnosis of PB is the detection of bronchial dendritic casts with flexible bronchoscopy [14]. These casts were detected in all our case with flexible bronchoscopy except in the case number 3 his bronchoacopy showed clear airways, however he expectorated a lot of casts during his admission.

Histopathology examination of specimens revealed mucus, fibrin, inflammatory exudate with ghost of inflammatory cells, charcot crystals, and eventually, a diagnosis of PB was rendered in all patients.

Hence, in patients with persistent atelectasis or recurrent respiratory distress, PB should be considered as a differential diagnosis.

Treatment of plastic bronchitis consists of bronchoalveolar lavage and FB aspiration. Rigid bronchoscopy was required for aspiration and extraction of the material in 3 cases. The patient with congenital heart disease did not require rigid bronchoscopy as he expectorated the all casts before the procedure. Other treatment options are available, such as administration of a fibrinolytic agent (tissue-type plasminogen activator (t-PA))13 via flexible bronchoscopy, but we have no experience of this technique. Most studies, however, indicate inhaled t-PA for disrupting the bronchial cast [14]. Other treatments include aerosolized fibrinolytics, such as urokinase, or inhaled mucolytics, such as acetylcysteine and dornase alfa [15]. In patients with hyper reactive airway disease, treatment is based on the use of inhaled and systemic corticosteroids [15].

Prognosis is generally good, except in cases with congenital heart diseases, in which mortality can be as high as 29%, 13 and 41% [16].

All our cases had good progress with satisfactory recovery and no death (Table 1).

Patient	Age (year)	Underlying disease	Indication for bronchoscopy	Bronchoscopic findings	Pharmacological treatment	Out come
Case 1	3	Asthma	Complete atelectasis in left lung	Cheesy like material occluded the left main bronchus	Dornase alfa	Residual atelectasis of left lower lobe
Case 2	9	Healthy	History of recurrent expectoration of a material	A cast which was seen in the superior segment of the right lower lobe	Dornase alfa Tazocin and Vancomycin were prescribed based on the microbiology result for the bronchoalveolar lavage (BAL)	Complete recovery
Case 3	6	Complexed Congenital Heart Disease	History of expectorated a yellow material	Thick secretions but no casts	Cefuroxime based on his culture	Complete recovery
Case 4	3	Pneumonia	Persistent left upper lobe collapse	Cast which was seen in the left upper lobe	Dornase alfa	Complete recovery

Table 1: *Clinical indication, bronchoscopic characteristics and outcome of patients.*

Conclusion

Plastic bronchitis is an uncommon process. However, it must be taken into account in patients with recurrent/persistent atelectasis and in case of a suspected foreign body. Patients with asthma are at great risk, other risk factors include underlying congenital heart disease, bronchopulmonary aspergillosis, sickle cell acute chest syndrome, and lower airway infections including adenovirus and mycoplasma organisms. Flexible bronchoscopy should be performed early to confirmation and clearance of airway obstructions.

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Volume 10 Issue 9 September 2021
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