



Plastic bronchitis in a child with thalassemia alpha

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Abstract

Objective: Plastic bronchitis is an unusual condition in children, associated with formation of mucofibrinous casts and mucous plugging of the tracheobronchial tree. Given that this illness is part of the differential diagnosis of acute respiratory failure, early treatment is important for improved prognosis. The aim of this report is to describe a case of plastic bronchitis in a child with alpha-thalassemia that was treated successfully with endoscopy.

Description: A 3 year old, black, male child, previously healthy, presented with acute respiratory failure and a chest x-ray showing pulmonary atelectasis. There was no evidence of respiratory symptoms or previous allergy state. The diagnosis of plastic bronchitis was made using flexible and rigid bronchoscopy, and confirmed by histopathologic findings. The child progressed well, treatment was based on supportive care and antibiotics were not used. Ten days after discharge, radiographic appearance was normal. Alpha thalassemia was diagnosed through hemoglobin electrophoresis.

Comments: Plastic bronchitis is clinically important because has similar presentation to other prevalent diseases, such as foreign body aspiration and asthma. When plastic bronchitis is suspected, endoscopy is indicated in order to confirm diagnosis and define treatment. Plastic bronchitis has been previously described in patients with cystic fibrosis, cardiac surgery and sickle cell disease. In this case, an association with alpha-thalassemia was observed.

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Introduction

Plastic bronchitis (PB) is an uncommon disease, rarely found as a primary condition, in which there is impaction or plugging of the tracheobronchial tree by mucofibrinous cylinders.¹ In the pediatric age group, PB is most often associated with inflammatory lung disease or increased

secretions, such as in asthma, cystic fibrosis and pulmonary infections.² Recent reports also demonstrate an association with sickle-cell anemia patients (acute chest syndrome), congenital heart disease and those subjected to heart surgery.^{2,3}

Although PB is an uncommon condition, its importance is based on the fact that it may exhibit clinical status very similar to diseases with elevated prevalence rates and early and specific diagnosis is required.

The present report describes a case of a pediatric patient admitted for acute respiratory insufficiency, of sudden onset, with suspicion of foreign body aspiration, which resulted in a diagnosis of PB associated with thalassemia alpha.

Case description

A three-year-old, black patient, previously healthy was admitted to the emergency room at the *Hospital São Lucas da PUCRS* on the 18th of January 2005 with respiratory

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distress and fever over the last 24 hours. There were no previous viral symptoms or any history of allergy or recurrent respiratory infections. On physical examination he presented tachypnea (respiratory rate of 50 bpm), subcostal and intercostal retractions, increased expiratory period and absent vesicle breath sounds in the left hemithorax. He was also suffering from hypoxemia, with oxygen saturation at 88-90% in room air.

Chest x-ray revealed bilateral hyperinflation, partial atelectasis of the left lung, mediastinum displaced ipsilaterally and discrete bilateral interstitial infiltration (Figure 1).

The blood test performed on admission revealed microcytic anemia (hematocrit: 27%, hemoglobin: 9.1 g/dl, mean cell volume: $64\mu^3$), with total white blood count at $16,500\text{ cells/mm}^3$ (basophils: 38.5%, neutrophils: 49.5% and lymphocytes: 8%). Arterial blood gas analysis in room air was pH: 7.37, $p\text{CO}_2$: 21 mmHg, $p\text{O}_2$: 61 mmHg, HCO_3 12 and SaO_2 99%.

The patient developed respiratory failure and required mechanical ventilation at the intensive care unit. The ventilator was a Sechrist IV with parameters set to PIP 35; PEEP 6; FR 20 bpm and FiO_2 40%. Due to the suspicion of foreign body aspiration, the patient underwent prompt flexible bronchoscopy in the intensive care unit, which demonstrated total obstruction of the main left bronchus by extremely thick secretions with a "caseous" appearance. After repeated attempts at bronchial aspiration, including with topical, diluted N-acetyl-cysteine, the secretion on the lumen of the bronchus was partially removed, revealing the presence of large quantities of the same secretion along the entire distal region of the left bronchial tree, with the characteristics of bronchial cast. Rigid bronchoscopy was

performed 24 hours later, with further removal of the bronchial cast. When fixed in formalin at 10%, the material removed for histological investigation took on a cylindrical shape with a length of 4 cm and an average thickness of 0.2 cm. Histopathology revealed a large quantity of eosinophils in fibrin (Figure 2). The bronchial lavage from the flexible endoscopy was negative for acid and alcohol fast bacilli, fungi and bacteria.

After the endoscopic interventions, the patient progressed satisfactorily and was weaned off ventilation in 72 hours and discharged from the intensive care unit. Hospital discharge took place on the tenth day, the patient had no fever, was breathing well and had no further respiratory complaints.

Some tests were performed at the end of the patient's stay in hospital: cystic fibrosis sweat test (negative), sickle-cell test (normal), simple chest x-ray (normal), cardiologic assessment normal). Supplementary to the anemia test, hemoglobin electrophoresis was run using the saponin hemolysate technique.⁴ The result demonstrated the presence of hemoglobin H (HbH) at 2%, characterizing thalassemia alpha.

From January 2005 to date the patient has had no recurrence of the condition.

Discussion

Plastic bronchitis is an uncommon condition, and has been given other names previously, such as fibrinous bronchitis or croup bronchitis.⁵ It generally occurs in patients suffering from asthma, cystic fibrosis, or sickle-cell anemia (as an acute chest syndrome), and during post-op for heart surgery.⁶ No previous description of PB in thalassemia alpha



Figure 1 - Chest x-ray with bilateral hyperinflation, partial atelectasis of the left lung, mediastinum displaced ipsilaterally and discrete bilateral interstitial infiltration

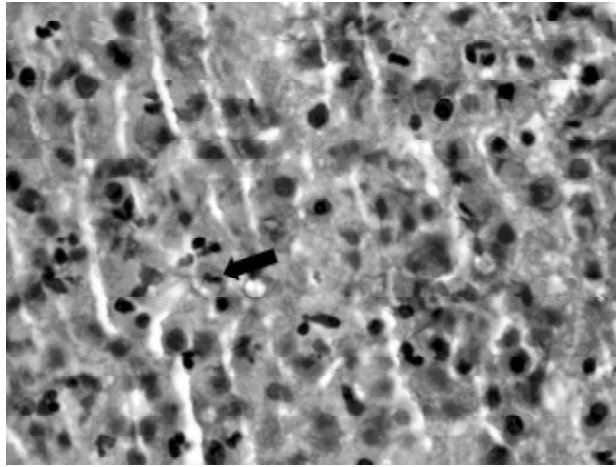


Figure 2 - Histopathology of the bronchial cast revealed a large quantity of eosinophils in fibrin, cellular debris and other polymorphonuclear cells (hematoxylin and eosine, 500X). The arrow shows eosinophils in the histologic section

patients had previously been published. It is possible that this combination occurs in other cases, since in approximately 20% of PB cases etiology is unrelated to the conditions cited above.⁶ The physiopathogenesis of PB in patients with sickle-cell anemia appears to be related to microvascular occlusion of sickle erythrocytes, increased cell adhesion to the vascular endothelium and liberation of inflammatory cytokines.² Thalassemia alpha, diagnosed in this case, can present with manifestations ranging from asymptomatic cases to varying forms of anemia, splenomegaly and increased susceptibility to infections.

It is possible that this patient could have been suffering from a viral infection, which was not tested for. The child presented with fever, there was interstitial infiltrate on chest x-ray and the endobronchial cast was type I – inflammatory. Seear *et al.* undertook a histopathological study that classified bronchial casts as type I or II.⁷ Type I casts exhibit a predominance of fibrin and eosinophil infiltrate and are more associated with inflammatory diseases. Type II casts have low cell content and are made up of mucin. They are more associated with cardiac patients. Etiologies that have been postulated for type II casts include low cardiac output (creating elevated venous pulmonary pressure), lymphatic drainage disorders and hypersecretion of mucus in the airways.⁸

Plastic bronchitis exhibits variable clinical manifestations that are non-specific. It may progress through dyspnea, respiratory difficulties, coughing, wheezing and/or fever. The similarity in presentation to foreign body aspiration or severe acute asthma has been described by Noizet *et al.*⁹ As was reported in this case, the symptoms of acute onset, extensive atelectasis and no previous morbidity, result in an

initial impression of foreign body aspiration. The presentation resulted in the immediate decision to explore the airways with endoscopy. The use of a flexible bronchoscope was justified by the initial severity of the condition and the ease with which it can be performed in the intensive care unit. Plastic bronchitis can be diagnosed by bronchoscopy or in response to the expectoration of bronchial casts.⁶

Treatment is based on the removal of the endobronchial mold by endoscopy, which is very often enough to result in satisfactory resolution, as was the case here. There are, however, proposals that additional treatment be given, which could include corticosteroids (2 mg/kg/day, in the case of type I molds),⁹ antibiotics⁹ for associated bacterial conditions, broncho-alveolar lavage, respiratory physiotherapy and topical N-acetyl-cysteine (2 to 4 ml of 10% solution).¹⁰⁻¹² More recently, the use of thrombolytics in aerosol form (urokinase) and the endotracheal instillation of recombinant immunoglobulin (Dornase Alpha) have been described.¹² Mechanical removal of the molds under direct observation (bronchoscopy) can be very useful in cases of children with acute respiratory insufficiency. The best treatment alternative has not been established because of the lack of controlled studies into a disease with low prevalence.

Cases with similar clinical presentation and x-ray findings should alert pediatricians to the possibility of a diagnosis of sickle-cell anemia or other blood disorders. Since presentation is atypical, very often with no severe anemia or other symptoms, cases of H-hemoglobinopathy associated with PB may have gone unnoticed. It appears to be recommendable, based on this case, that special care should be given to patients with pulmonary infection

and atelectasis to consider diagnoses of other blood disorders.

Plastic bronchitis is an uncommon finding in pediatrics, but its importance is recognized to the extent that it can mimic foreign body aspiration or severe acute asthma, both of which are commonly found in emergency rooms and intensive care units. Furthermore, the search for concomitant diseases must be part of the evaluation of this type of patient. If a pediatrician suspects PB and indicates immediate flexible bronchoscopy, in cases such as the one reported here, etiologic diagnosis can be made earlier and the prognosis of patients with this condition can be improved.

References

1. Bowen A, Oudjhane K, Odagiri K, Liston SL, Cumming WA, Oh KS. Plastic bronchitis: large, branching, mucoid bronchial casts in children. *Am J Roentgenol.* 1985;144:371-5.
2. Raghuram N, Pettigano R, Gal AA, Harsch A, Adamkiewicz TV. Plastic bronchitis: an unusual complication associated with sickle cell disease and the acute chest pain. *Pediatrics.* 1997;100:139-42.
3. Moser C, Nussbaum E, Cooper DM. Plastic bronchitis and the role of bronchoscopy in the acute chest syndrome. *Chest.* 2001;120:608-13.
4. Naoun PC. *Eletroforeses, técnica e diagnóstico.* 1ª ed. São Paulo: Livraria e Editora Santos; 1974.
5. Bowen A. Plastic or cast bronchitis? *Am J Dis Child.* 1990;144:1075-6.
6. Brogan TV, Finn LS, Pyskaty DJ Jr, Redding GJ, Ricker D, Inglis A, et al. Plastic bronchitis in children: a case series and review of the medical literature. *Pediatrics Pulmonol.* 2002;34:482-7.
7. Seear M, Hui H, Magee F, Bohn D, Cutz E. Bronchial casts in children: a proposed classification based on the nine cases and a review of the literature. *Am J Resp Crit Care Med.* 1997;155:364-70.
8. Quasney MW, Orman K, Thompson J, Ring JC, Salim M, Schoumacher RA, et al. Plastic bronchitis occurring later after the Fontan procedure: treatment with aerosolized urokinase. *Crit Care Med.* 2000;28:2107-11.
9. Noizet O, Leclerc F, Leteurte S, Brichet A, Pouessel G, Dorkenoo A, Fourier C, Cremer R. Plastic bronchitis mimicking foreign body aspiration that needs a specific diagnostic procedure. *Intensive Care Med.* 2003;29:329-31.
10. Costello JM, Steinhorn D, McColley S, Gerber ME, Kumar SP. Treatment of plastic bronchitis in a Fontan patient with tissue plasminogen activator: a case report and review of the literature. *Pediatrics.* 2002;109:e67.
11. Durward A, Forte V, Shemie SD. Resolution of mucous plugging and atelectasis after intratracheal rhDNase therapy in a mechanically ventilated child with refractory status asthmaticus. *Crit Care Med.* 2000;28:560-2.
12. Manna SS, Shaw J, Tibby SM, Durward A. Treatment of plastic bronchitis in acute chest syndrome of sickle cell disease. *Arch Dis Child.* 2003;88:626-7.

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