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Case report

Plastic bronchitis: A case report

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ABSTRACT

Plastic bronchitis is a rare and underdiagnosed disease characterized by the formation and expectoration of bronchial casts of amorphous material, which can be potentially fatal. It is more frequent in pediatric population. Symptoms can range from chronic cough and dyspnea to respiratory failure depending on the area of the compromised airway. Casts are classified as type I when constituted by inflammatory cells and accompany diseases such as asthma and pneumonias; and type II when acellular and are associated with congenital heart diseases following procedures such as Fontan.

We report the case of a male schoolchild with a history of complex congenital heart disease, treated with palliative surgery, evaluated in the emergency department for cough and respiratory distress. The mother referred expulsion of gelatinous material after coughing. During clinical evaluation, expulsion of bronchial casts was evidenced, suggesting a plastic bronchitis. He underwent a diagnostic and therapeutic bronchoscopy and received initial treatment with respiratory therapy, nebulized hypertonic saline solution, mucolytics, dornase alpha and nebulized heparin. The hospitalization revealed a stenosis of the right pulmonary artery, which was corrected with stent. The patient progressed satisfactorily with improvement of cough and expectoration. He was discharged with combined treatment, nebulized medications and those concerning his underlying disease.

1. Introduction

Plastic bronchitis (PB) is a rare disease characterized by the expectoration of amorphous, rubbery, cohesive and branched bronchial casts that obstruct the airway partially or totally [1]. It occurs at any age but is more frequent in the pediatric population. The first known description of PB dates from the second century BC by Galen, a Greco-Roman physician, who described patients that expectorated soft and viscous casts that he interpreted as veins and arteries. 1500 years later, Morgagni identified the material as mucus casts coming from the airway rather than pulmonary veins [2]. To date, less than 600 cases of PB have been reported, its incidence is unknown, and it is probably an underdiagnosed disease [3].

PB manifests clinically with cough, wheezing, dyspnea, hypoxemia, chest or pleuritic pain and occasionally fever. Its characteristic feature is the formation of bronchial casts that can vary from rubbery structures that fill the small airways to casts formed of amorphous material that

occupy the entire bronchial tree [3]. When obstruction of the central airway occurs, respiratory failure presents especially in those individuals with poor ability to manage secretions. Physical examination can reveal wheezing, decreased breath sounds and dullness to percussion. Chest X-ray and CT scan typically show partial atelectasis of the segments involved with compensatory hyperinflation of the unaffected segments. Diagnosis is confirmed by rigid and/or flexible bronchoscopy or by repeated observation of the expectorated structures [3].

PB occurs accompanying various respiratory, cardiac and systemic diseases. It is currently classified according to the composition of the bronchial casts described by Seear et al. [4] as type I, characterized by casts containing inflammatory cells with eosinophilic infiltrate with a center formed by Charcot-Leyden crystals surrounded by respiratory epithelium. This case is associated with situations characterized by severe bronchial inflammation such as severe asthma, pneumonia, allergic bronchopulmonary aspergillosis, cystic fibrosis, bronchiectasis, acute thoracic syndrome secondary to sickle cell disease, smoke

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Fig. 1. A. Chest X-ray with perihilar congestion and bilateral basal interstitial infiltrate with signs of sternotomy closure by previous surgery. B, C, D. Chest CT scan showing zones of left partial atelectasis and mucus casts obstructing the left source bronchus, as well as a patchy interstitial infiltrate.

inhalation among others. Type II is characterized by acellular amorphous material composed of fibrin and mucin without inflammatory cells that accumulates after surgical repair procedures in congenital cyanotic heart disease, such as Fontan procedure [5,6]. In these cases, the intra thoracic hemodynamic balance is altered because the systemic blood flow is derived to the pulmonary circulation, with lymphatic drainage imbalance, increase of the pulmonary venous pressure and induction of the formation of bronchial casts [1]. In addition, PB sometimes presents without a known risk factor, in these cases the casts are acellular suggesting an undetected underlying abnormality of the lymphatic drainage system [7].

We describe the case of a male school patient with a history of complex congenital heart disease, treated with palliative surgery, who presents a clinical picture compatible with PB. The clinical, radiological and pathological findings, as well as the treatment and outcomes are described.

2. Case report

Patient of 4 years and 5 months old, with a history of complex congenital heart disease suspected from the prenatal period by fetal echocardiography that showed a pulmonary atresia without ventricular septal defect treated with Glenn surgery at 8 months old and subsequently fenestrated Fontan procedure at 3 years of age. Chronically managed with carvedilol 1.12 mg every 8 hours, sildenafil 6 mg every 8 hours, bosentan 20 mg every 12 hours, enalapril 2.5 mg every 12 hours, furosemide 8 mg every 12 hours, acetylsalicylic acid 70 mg daily, warfarin with adjustment according to INR and spironolactone 20 mg every day. Without previous respiratory symptoms, consulted for 5 days of dry cough and respiratory distress, followed by expectoration of bronchial casts of whitish amorphous material accompanied by spots of

blood. On physical examination, heart auscultation showed no gallop or murmurs, rhonchi were present in both lung fields, abdomen without masses and the extremities showed no abnormalities. Chest X-rays revealed reticular opacities predominantly bilateral at the bases. Due to the characteristics of the expectorated material, a diagnosis of PB was considered and the patient was hospitalized for management. Laboratory exams at admission showed leukocytes 7160, neutrophils 59.6%, lymphocytes 30.7%, Hb14.4, Hct 43.4%, platelets 109.000, Creactive protein 1.11, PT 30.3 seconds, PTT 40.4 seconds, INR 2.11, Alpha 1 Antitrypsin 1.5 (0.9-2), BUN 18, creatinine 0.4, sodium 136, potassium 3.52, chlorine 99.4, calcium 8.64, arterial gases with pH 7.406, PO 236.9, PCO 245.7, HCO 328.1 and negative blood cultures at 48 hours. Chest CT scan showed a post-surgical condition with permeable Fontan circulation, moderate stenosis of the preanastomotic right pulmonary artery, a decrease in the caliber of the distal branches without evidence of acute pulmonary thromboembolism with a pattern compatible with chronic thromboembolism, mosaic attenuation, bronchial occupation by long mucus plugs with secondary atelectasis, thin aortopulmonary collateral bronchial arteries (< 1.5mm), tricuspid atresia, hypoplastic right ventricle and IAC without VSD. Studies for hypercoagulability evaluation in addition to normal coagulation factors VIII, V, VII, X, XI and II were normal (see Fig. 1).

Treatment was initiated with oxygen therapy, micro-nebulizations with bronchodilator salbutamol prior to the nebulization of 3% hypertonic saline solution every 12 hours, in addition to nebulizations with dornase alpha 2.5 mg daily and nebulized heparin 5000 units every 8 hours. The patient persisted with cough, mobilization of pulmonary secretions, moderate dyspnea and expectoration of casts of gelatinous material for six days. Bronchoscopy was performed, the mucosa was found with edema and moderate erythema with remains of the thick mucus material on the carina and main bronchi as well as

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Fig. 2. A. Hematoxylin and eosin staining represents acellular mucoid material. B. Expectorated mucoid material molding the bronchial tree. C. Movat pentachromic staining, the acellular mucoid material is represented in red. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

casts which were removed leaving the bronchial tree clean. Culture of the tracheal secretion reported a mixed flora. The macroscopic analysis of the expectorated sample evidenced a mucoid material that shaped the bronchial architecture. Microscopically, with hematoxylin and eosin staining and Movat staining, acellular mucoid material was observed which confirms bronchial cast type II (Fig. 2). During hospitalization a cardiac catheterization was performed, showing a patent fenestrated Fontan, with low pulmonary vascular resistances and pulmonary pressure with Fio2 100%, and stenosis of the pulmonary artery for which stent placement was performed without complications. It was considered that the obstruction at the level of the pulmonary valve could be the trigger of PB. The patient continued with combined treatment and respiratory therapy achieving a saturation improvement and progressive reduction of the expectoration of casts, with discharge for outpatient management where, according to the evolution, the relevance of lymphatic embolization for long-term management of PB will be defined.

3. Discussion

PB is characterized by the production of bronchial casts of protease content, which behave as a foreign body along the bronchial tree [8]. It is a potentially fatal pathology that commonly occurs in patients with a history of Fontan palliative cardiovascular surgery. Fontan procedure was designed for children with complex congenital heart defects with univentricular hearts. It consists of establishing passive blood flow through the lungs by deviating both the inferior and superior vena cava circulation directly to the pulmonary arteries, which generates an important alteration of the pulmonary vascular dynamics. As the procedure becomes safer and the survival rates increase, PB occurs more frequently, where the formation of bronchial casts occlude the airway without producing stenosis. There is no evidence of decreased tracheobronchial lumen despite multiple airway cleaning interventions. Occasionally Fontan procedure malfunctions due to stenosis of the derivation improving the pulmonary symptoms but at the same time altering the systemic circulation. Sometimes the procedure must be reverted to improve or decrease an almost fatal PB, which is always a complex situation due to the cardiovascular implications [9].

The general prevalence is unknown [10], in Fontan it is reported between 4 and 14% [1], with a mortality described in 5–60% of cases [5,10].

The increase in venous pressure with dysfunction and loss of integrity in the bronchial mucosa membrane and the increase in the pressure of the lymphatic vessels of the thorax associated with broncholymphatic fistulas with production of bronchial casts are involved [9]. The symptoms are nonspecific and should be suspected in patients with chronic cough, wheezing and progressive dyspnea since early diagnosis and treatment influence long-term prognosis [1,9].

The treatment of PB is a challenge; no intervention has been proven effective in all patients, regardless of the etiology and physical properties of the bronchial casts (solid or elastic). Rigid and/or flexible bronchoscopy and the intensive medical support remain as the primordial management for this type of patients [3]. Prevention of cast formation is the main objective. Treatment has been established by previous case reports and includes bronchial cleaning with bronchoscopy, management of increased venous pressure and aggressive pulmonary vasodilation with sildenafil and bosentan [1], as well as inhaled and systemic steroids due to their muco-regulatory properties, especially in PB type I [11], use of mucolytics (dornase alpha, N-acetylcysteine, hypertonic saline solution) [1,12], intravenous or inhaled heparin similar to its use in cases of injury by smoke inhalation, (intravenously improves oxygenation, decreases barotrauma and airway edema; the mechanism of action is unclear [13]), followed by fibrinolytics such as urokinase and inhaled tissue plasminogen activator [14]. In patients with poor response to treatment, it is necessary to remove the Fontan or perform a heart transplant [1,9] and in adults, lymphatic percutaneous embolization is described to attenuate the loss of lymph, with partial or complete resolution of the symptoms [15]. Despite the efforts, mortality in this type of patients is still very high.

4. Conclusion

PB is a rare, variable and potentially fatal disease. In the case described, the cause was associated with the palliative management of complex congenital heart disease and its treatment required multidisciplinary intervention with bronchoscopy, multiple inhaled therapy and adequate control of the underlying disease, with stabilization of the expectoration of bronchial casts in the long term.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2019.100876.

References

- [1] C.M. Avitabile, D.J. Goldberg, K. Dodds, Y. Dori, C. Ravishankar, J. Rychik, A multifaceted approach to the management of plastic bronchitis after cavopulmonary palliation, Ann. Thorac. Surg. 98 (2014) 634–640.
- [2] R.S. Johnson, E.G. Sita-Lumsden, Plastic bronchitis, Thorax 15 (1960) 325-332.
- [3] T.V. Borgan, L.S. Finn, D.J. Pyskay Jr., G.J. Redding, D. Ricker, A. Inglis,

R.L. Gibson, Plastic bronchitis in children: a case series and review of the medical literature, Pediatr. Pulmonol. 34 (2002) 482–487.

- [4] M. Seear, H. Hui, F. Magee, D. Bohn, E. Cutz, Bronchial casts in children: a proposed classification based on nine cases and a review of the literature, Am. J. Respir. Crit. Care Med. 155 (1) (1997) 364–370.
- [5] M.H. Eberlein, M.B. Drummond, E.F. Haponik, Plastic bronchitis: a management challenge, Am. J. Med. Sci. 335 (2008) 163–169.
- [6] K.R. Schumacher, T.P. Singh, J. Kuebler, K. Aprile, M. O'Brien, E.D. Blume, Risk factors and outcome of Fontan-associated plastic bronchitis: a case-control study, J. Am. Heart Assoc. 3 (2014) e000865.
- [7] B.K. Rubin, Plastic bronchitis, Clin. Chest Med. 37 (3) (2016 Sep) 405–408.
- [8] P. Madsen, S.A. Shah, B.K. Rubin, Plastic bronchitis: new insights and a classification scheme, Paediatr. Respir. Rev. 6 (4) (2005 Dec) 292–300.
- [9] G. Grutter, D. Di Carlo, F. Gandolfo, R. Adorisio, S. Alfieri, G. Michielon, et al., Plastic bronchitis after extracardiac Fontan operation, Ann. Thorac. Surg. 94 (3) (2012 Sep) 860–864.
- [10] R.L. Caruthers, M. Kempa, A. Loo, E. Gulbransen, E. Kelly, S.R. Erickson, et al.,

Demographic characteristics and estimated prevalence of Fontan-associated plastic bronchitis, Pediatr. Cardiol. 34 (2) (2013 Feb) 256–261.

- [11] Y. Onoue, Y. Adachi, F. Ichida, T. Miyawaki, Effective use of corticosteroid in a child with life-threatening plastic bronchitis after Fontan operation, Pediatr. Int. 45 (1) (2003) 107–109.
- [12] A. Kumar, K.R. Jat, M. Srinivas, R. Lodha, Nebulized N-Acetylcysteine for management of plastic bronchitis, Indian Pediatr. 55 (8) (2008 Aug 15) 701–703.
- [13] P.A. Walker, S.K. Shah, P.A. Letourneau, N.D. Allison, C.S. Cox, Treatment of plastic bronchitis using serial flexible bronchoscopy and aerosolized heparin therapy, Eur. J. Pediatr. Surg. 23 (2) (2013 Apr) 157–160.
- [14] A. Mendoza Soto, L. Galletti, P. Gómez de Quero, M.V. Ramos Casado, J.M. Velasco Bayón, Plastic bronchitis. A case report and review of the literature, An Pediatr. (Barc) 62 (1) (2005) 72–75.
- [15] M.G. Itkin, F.X. McCormack, Y. Dori, Diagnosis and treatment of linphatic plastic bronchitis in adults using advanced lymphatic imaging and percutaneous embolization, Ann. Am .Thorac. Soc. 13 (10) (2016 Oct) 1689–1696.