Intrathoracic Rosai-Dorfman Disease: Hemorrhage With Routine Diagnostic Procedure
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We present a case of a 33-year-old female with a slow growing, right peribronchial vascular mass and associated symptoms of progressive cough, dyspnea on exertion, and hemoptysis. On routine diagnostic flexible bronchoscopy with needle biopsy, the lesion hemorrhaged extensively requiring emergent thoracotomy, right lower and middle bilobectomy. The histopathology of the specimen was consistent with the rare and unusually located entity Rosai-Dorfman disease.


Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, typically presents as painless cervical lymphadenopathy, mostly reported in the Afro-Caribbean population during the first or second decade of life. Although extra nodal sites are reported, only rare reports exist about intrathoracic RDD, and clinical presentations have typically been benign and self-limited with mild hemoptysis. Advances in radiology and aggressive diagnosis will likely bring more of these patients to light, but the risks of biopsies and differentiation from cancer should be understood. The case presented describes these issues.

A 33-year-old female initially presented to the emergency department in 2009 for right lateral, pleuritic chest pain of 2 weeks duration and a productive cough. A contrast-enhanced computed tomography (CT) scan demonstrated an incidental finding of a vascular right middle and lower lobe peribronchial density narrowing the bronchi to the lateral segment of the right middle lobe, the right lower lobe, the right lower lobe lateral segment, and also the right hilar pulmonary arteries.

She subsequently presented to our clinic in January of 2013 after exacerbation of her chronic cough, dyspnea on exertion, and very mild intermittent hemoptysis. A repeat non-contrast CT (Fig 1B, 2B) revealed the right infrahilar mass to be slightly increased in size since 2009, occluding the right middle lobe subsegmental pulmonary arterial branch. A flexible bronchoscopy with biopsy and possible needle biopsy was scheduled.

Bronchoscopy revealed significant narrowing of the basilar segmental bronchi and the superior segmental bronchi to the right lower lobe. Upon aspiration of this area with a 21-gauge needle, profuse bleeding began and would not stop despite balloon tamponade, time, cold saline, and saline with epinephrine, finally responding to packing of the intermediate bronchus with SURGICEL (Ethicon, Somerville, NJ). Due to the ferocious blood loss, severe airway instability, an apparently growing mass, and otherwise poor options for permanent control with angiographic techniques, the decision was made to...

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proceed to a right posterolateral thoracotomy and ultimately right middle and lower bilobectomy.

A firm and seemingly calcified mass was found in the right lower bronchial area which involved the middle lobe along the fissure and in the hilum. Almost all these tissues bled easily and were markedly abnormal.

Histopathological examination showed an unencapsulated, loosely circumscribed, lobulated mass surrounding the main bronchi composed of diffuse histiocytic and plasmacytic infiltration of the lung parenchyma with prominent “emperipolesis” of plasma cells within the histiocytes (Fig 3, A, B). Emperipolesis was defined by Humble and colleagues as “the active penetration of one cell by another which remains intact,” thereby differentiating it from phagocytosis in that the engulfed cell remains viable. This was also seen within peribronchial lymph nodes, and extended around the pulmonary hilar vasculature. The histiocytes within the infiltrate were strongly positive for S100 and CD68-KP1, negative for CD163 and cytokeratin AE1/AE3, and with CD3 and CD20 marking T and B lymphocytes at the periphery. Immunoglobulin IgG and IgG4 stained the plasma cells extensively. Acid-fast bacillus and Grocott’s methenamine silver stains were negative for microorganisms. This staining pattern is consistent with Rosai-Dorfman disease.

**Comment**

Rosai-Dorfman disease (RDD), alternatively known as sinus histiocytosis with massive lymphadenopathy is a rare, benign disorder of unknown etiology that typically manifests with painless cervical lymphadenopathy and is diagnosed pathologically [7, 8]. It has since been shown that extranodal disease can occur in 25% to 40% of patients and can affect many different organ systems [1, 2]. In a single series by Cartin-Ceba and colleagues [1], intrathoracic involvement was documented in 43% of patients diagnosed with RDD, in whom the most common radiologic finding was mediastinal lymphadenopathy and the most frequent clinical symptoms were dyspnea and cough.

Given the benign nature of this disorder, either no therapy or conservative management with corticosteroids is the standard of care [2]. However, extranodal disease can cause life-threatening complications from extension into vital organs. In such cases surgical intervention may be warranted.

Cases requiring lobectomy and even complete pneumonectomy have been reported in the literature in patients with intrathoracic RDD [3–6]. All of these cases with the exception of 1 were elective secondary to associated symptoms of pleuritic chest pain, mass compression of the pulmonary artery, or pulmonary hypertension. The article by Scott and colleagues [6] describes an emergent case of a young patient with acute hemoptysis on presentation, whose source of bleeding could not be identified on bronchoscopy requiring an urgent total pneumonectomy.

We present a case of a 33-year-old female with unilateral, intrathoracic RDD who required emergent right bilobectomy for severe hemorrhage after routine diagnostic needle biopsy during bronchoscopy. We believe this to be the first such complication of a routine diagnostic procedure in a patient with intrathoracic RDD requiring conversion to thoracotomy.

Inspection of the resected specimen revealed a very friable, easily bleeding mass. The available CT studies did
Contralateral pneumothorax after pneumonectomy (CPAP) is a rare but potentially fatal condition. Therefore, when treating CPAP prevention of recurrence is very important. Despite a number of case reports about CPAP, its management is still controversial. We describe 4 cases of CPAP that were treated successfully by bullectomy and coverage with absorbable polyglactin mesh.

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We retrospectively investigated the surgical procedures and clinical courses of 4 patients with CPAP treated at our institution between January 2007 and October 2012. All of the patients were male and the average age was 66.5 years (range, 59 to 76 years). The reason for previous pneumonectomy was lung cancer in 3 patients and tuberculosis in 1. The periods between pneumonectomy and occurrence of pneumothorax were 15 days, 3 months, 1 year, and 3 years 4 months, respectively. Three of the patients experienced sudden severe dyspnea and presented at the emergency room of our institution; the remaining patient had suffered dyspnea for several days before admission. All of the patients were treated by conventional chest-tube drainage immediately after the diagnosis of CPAP. Chest computed tomography demonstrated emphysematous bullae, and all 4 patients were treated surgically under general anesthesia without any extracorporeal cardiopulmonary support. Three patients underwent surgery through an anterior axillary thoracotomy and 1 underwent video-assisted thoracoscopic surgery. All patients were treated by bullectomy and coverage with polyglactin mesh. In 1 case, selective double ventilation of the right upper and middle lobes using an intrabronchial blocker was performed to facilitate resection of the bulla on the mediastinal side of the right lower lobe (Fig 1, A, B). In this case, lung cancer at right S8 was also resected (Fig 2). In the case of video-assisted thoracoscopic surgery, intermittent ventilation using high concentration oxygen was required. The mean operation time was 74 minutes (25, 57, 65, and 150 minutes, respectively) and in all cases the bleeding volume was negligible. An extracorporeal membrane oxygenation system was placed on standby for 1 case, but it was not needed. None of the patients required extracorporeal cardiopulmonary support. Paroxysmal atrial fibrillation occurred in 1 case postoperatively, but no other postoperative complication was observed. The average drainage period and length of hospitalization after