

## Case Report

# A rare case of bronchial elastosis: An unusual presentation of an unexpandable lung

Raghav Gupta<sup>1,2</sup>, Hassan Patail<sup>1,3</sup>, Mohammad R Al-Ajam<sup>1,2</sup>

<sup>1</sup>SUNY Downstate Medical Center, Departments of <sup>2</sup>Pulmonary and Critical Care Medicine and <sup>3</sup>Internal Medicine, VA Hospital, Brooklyn, New York, USA

## ABSTRACT

Endobronchial obstruction is a known cause of an unexpandable lung. Endobronchial lesions are usually malignant, however benign cause like hemartoma, lipoma, amyloidosis and neuroendocrine tumors are known. We, hereby present the rare cause of bronchial elastosis presenting as right lower lobe lung collapse.

**KEY WORDS:** Collapsed lung, endobronchial obstruction, pleural effusion

**Address for correspondence:** Dr. Raghav Gupta, 706 Mindy Lane, Piscataway, NJ 08854, USA. E-mail: drraghav11@gmail.com

## INTRODUCTION

Elastosis is described as the focal excess of elastic fibers in the tissue. Elastosis has been commonly observed in the cardiology literature where it deposits as a fibroelastoma on cardiac valves associated with high risk of embolism and often needing surgery.<sup>[1,2]</sup> It has also been reported in the gastrointestinal (GI) tract often presenting as a mouth lesion or a colon polyp and on the skin as a tumorous growth.<sup>[3-5]</sup> Elastosis in the lungs is extremely rare. Till present, there has been only one case of bronchial elastosis published in a French paper.<sup>[6]</sup> Hereby, to the best of our knowledge, we report the second rare case of bronchial elastosis presenting as right lower lobe (RLL) lung collapse.

## CASE REPORT

An 89-year-old nonsmoker male with the past medical history of hypertension, dementia, and benign prostatic hyperplasia presented with progressive worsening of shortness of breath for 5 months and mild cough with clear white color sputum. He denied any fevers, chills,

weight loss, night sweats, or loss of appetite. On physical examination, he was alert and oriented, with normal vital signs. His SpO<sub>2</sub> was 92% while breathing ambient air. Bilateral diminished respiratory excursion and faint bibasilar wheezing were noted on chest examination, and cardiac auscultation found no murmurs or irregularities. The blood cell counts, electrolytes, and renal and liver function tests were unremarkable and QuantiFERON-Tuberculosis Gold was negative. An arterial blood gas analysis showed pH 7.45, PCO<sub>2</sub> 33.9 mmHg, and PO<sub>2</sub> 68.1 mmHg on room air. Chest radiography showed right-sided pleural effusion [Figure 1], and computed tomography of the chest showed bilateral pleural effusion right greater than left, with atelectasis of RLL [Figure 2]. The patient underwent diagnostic and therapeutic right-sided pigtail catheter placement which drained serous exudative pleural fluid on Light's criteria with lymphocytic predominance. Pleural cultures and cytology were negative. Postpleural fluid drainage and repeated chest imaging showed right-sided unexpandable lung with pneumothorax ex vacuo and possible endobronchial lesion in the right middle

Access this article online	
<b>Quick Response Code:</b> 	<b>Website:</b> <a href="http://www.lungindia.com">www.lungindia.com</a>
	<b>DOI:</b> 10.4103/lungindia.lungindia_31_17

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

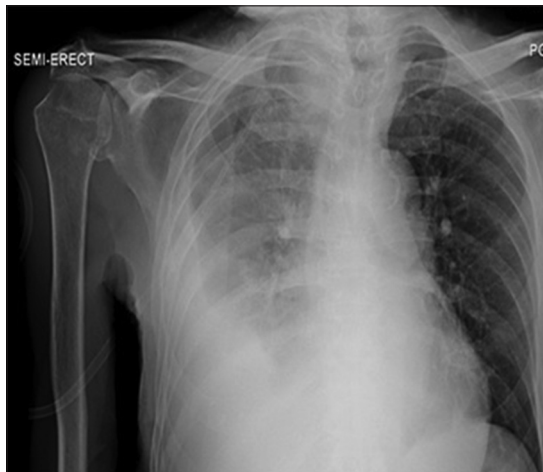
**For reprints contact:** [reprints@medknow.com](mailto:reprints@medknow.com)

**How to cite this article:** Gupta R, Patail H, Al-Ajam MR. A rare case of bronchial elastosis: An unusual presentation of an unexpandable lung. Lung India 2018;35:50-3.

lobe (RML) bronchus [Figures 3 and 4]. Subsequently, bronchoscopy was performed which showed narrowed RML bronchi with significant mucosal edema and a round smooth-bordered 3.0 cm endobronchial lesion at RLL bronchi not allowing the bronchoscope to pass beyond the lesion. Endobronchial biopsies, bronchoalveolar lavage, and cytological brushing were performed. RML biopsy showed chronic bronchitis and was negative for malignancy or granulomatous inflammation. RLL biopsy with Verhoeff's elastic staining showed plexus of elastic fibers parallel to basement membrane [Figure 5]. Immunoperoxidase staining on RLL showed the presence of elastic tissue as fibrillar amphophilic material which was centered around blood capillary vessels consistent with the diagnosis of bronchial elastosis [Figure 6]. The first cytology from pleural fluid was negative, and because of the patient's wishes, no further staging and intervention including bronchoscopy removal or surgical resection were performed. After physiological and clinical improvement, the patient was discharged in a stable condition to a long-term care facility.

## DISCUSSION

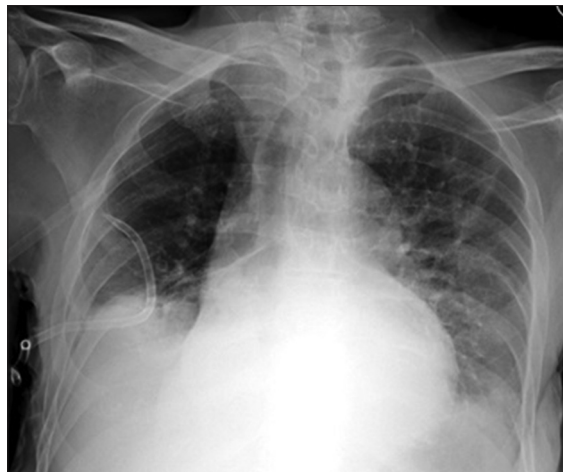
Endobronchial lesion can be malignant neoplasms, benign tumors, aspirated foreign bodies, postinflammatory, infectious, or traumatic strictures, and inflammatory polyps. Amyloidosis is one of the rare causes of endobronchial lesions. The most common are the primary malignancies including bronchogenic carcinoma and carcinoid tumors.<sup>[7]</sup> Benign endobronchial tumors can be divided into tumors of mesenchymal, submucosal gland, and surface epithelial origin. Of these, tumors of mesenchymal origin represent the great majority, with the most common being hamartoma.<sup>[8]</sup> Benign endobronchial neoplasms are a rare entity, and they may present with diverse pathological findings. Endobronchial tumors usually present with symptoms such as cough,



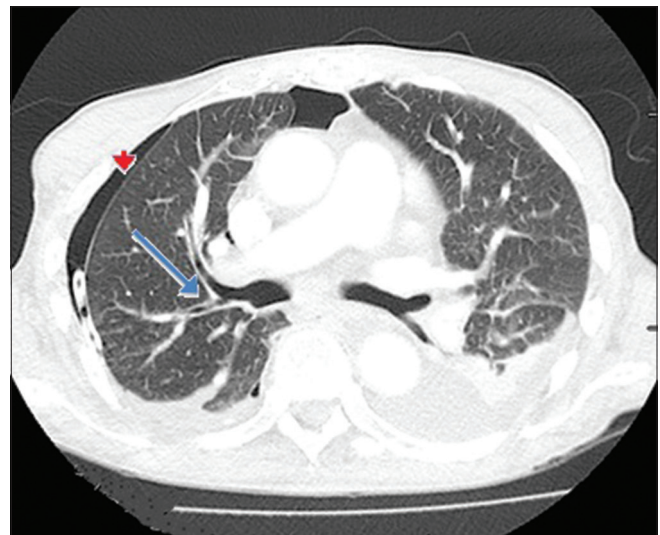
**Figure 1:** Chest radiography on admission showing right-sided pleural effusion



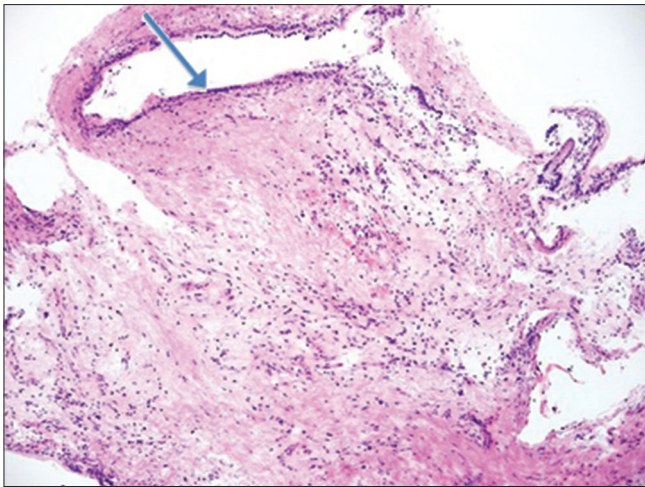
**Figure 2:** Computed tomography of chest on admission before thoracentesis confirming radiographic findings of right-sided pleural effusion



**Figure 3:** Chest radiography postpigtail catheter drainage showing incomplete expansion of right lung



**Figure 4:** Computed tomography of the chest postpigtail catheter drainage confirming right-sided unexpandable lung (red arrow) with possible endobronchial lesion in the right middle lobe bronchus (blue arrow)



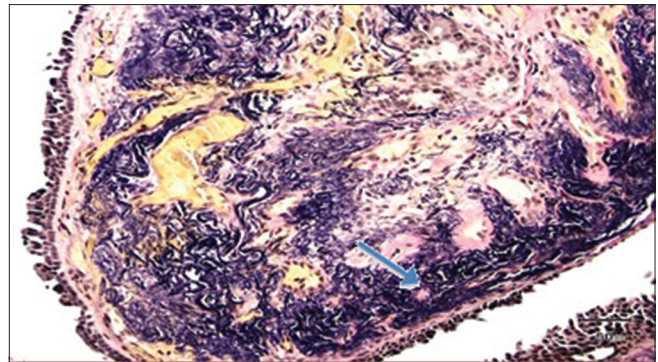
**Figure 5:** Right lower lobe lung biopsy with Verhoeff's elastic staining showing plexus of elastic fibers (blue arrow) parallel to basement membrane

hemoptysis, recurrent pneumonia, wheezing, and chest pain.<sup>[9]</sup> However, bronchial elastosis, a benign tumor of mesenchymal origin, has only been reported once in French literature thus far.<sup>[6]</sup>

The elastin content of lungs is quite variable, ranging from as low as 2% in rodents up to 28% in the cow and in humans. For this reason, most of the chemical and biological studies that have been conducted on elastin have used either the bovine neck ligament or the ascending aorta of several species.<sup>[10]</sup> Elastic fibers are ubiquitous in the lung, associated with collagen and proteoglycans. Elastin and collagen fibers from alveoli, bronchi, interlobular septae, and the pleura all appear to have connections with fibers of pulmonary arteries. Elastic fibers in respiratory bronchioles become more distinct distally, where the alveoli are also more developed.<sup>[10]</sup>

Arrigoni *et al.* studied 130 patients with benign tumors of the lung and only 6% of those were endobronchial in location.<sup>[11]</sup> Elastosis can occur when loss of function mutations arise to genes responsible for elastin production. This leads to overproduction of elastic tissue. Elastic tissue overproduction can be seen as small, uniform, symmetric papules that vary in anatomic location.<sup>[12-14]</sup> Histologic examination finds overproduction of elastic tissue in an interlaced or fragmented pattern. Elastosis is viewed as finely granular and/or fibrillar amphophilic material, which can sometimes include a fibrous component also known as elastofibromatous change. These changes appear centered around blood vessels as in our patient's pathology. The phenotypical appearance is often mistaken for amyloid, but Congo red stains remain negative whereas they stain strongly positive for elastin.<sup>[15]</sup>

Elastosis has been reported in cardiology, GI, and dermal literature. In cardiac literature, the origin of these tumors is still in question whether they are congenital or acquired.<sup>[1,2]</sup>



**Figure 6:** Immunoperoxidase staining demonstrating the presence of elastic tissue as fibrillar amphophilic material centered around capillary vessels consistent with the diagnosis of bronchial elastosis (elastic tissue shown by blue arrow)

Most are found on cardiac valves, and they are believed to be acquired by a microthrombi theory which entails small microthrombi coalescing on cardiac valves leading to endothelial damage and then forming elastomas.<sup>[16]</sup> These elastomas have a tendency to become embolic.<sup>[17]</sup> Histology from cardiac valve elastomas shows elastin and collagen fibers surrounded by endocardium.<sup>[18]</sup> In GI literature, most elastomas are found incidentally during colonoscopy and sometimes present as small intestinal obstruction, abdominal pain, and/or hematochezia.<sup>[3,4]</sup> The median age of presentation is in the seventh decade with equal male:female ratio. Pathology shows increased elastic fibers fine to fibrillary pattern and as bundles with angiocentricity. Etiology of these lesions in the GI system is unknown, but theories include reactive and degenerative processes including possible elastic degeneration of submucosal vessels and mucosal prolapse.<sup>[19]</sup>

Endobronchial lesions are usually malignant but when benign the differential diagnosis consists of lipoma, hamartoma, neuroendocrine tumors, amyloidosis, etc.<sup>[20]</sup> Amyloidosis is more heavily considered in the differential as pathology is difficult to differentiate based on hematoxylin and eosin stains alone.<sup>[19,20]</sup> Special stains for elastin are needed as performed in the case of our patient. Flexible bronchoscopy is necessary for diagnosis and evaluation of these endobronchial tumors. Bronchoscopy removal or surgical resection is the modality of treatment for these tumors. Surgery is considered for patients with having lesions with increased risk of recurrence, extrabronchial extension of the lesions, or destruction of the peripheral lung due to long-term atelectasis.<sup>[9]</sup>

In our case, the bronchial elastosis was found to be the rare cause of an endobronchial obstructive lesion resulting in an unexpandable lung. Patient's clinical symptoms improved temporarily by drainage of pleural effusion; however, the cause of the endobronchial lesion needs to be treated to prevent further episodes of deterioration.



### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

### REFERENCES

1. Abbasi AS, Da Costa M, Hennessy T, Kiernan TJ. Cardiac papillary fibroelastoma presenting as acute stroke. *BMJ Case Rep* 2013;2013. pii: Bcr2013010092.
2. Wang Y, Wang X, Xiao Y. Surgical treatment of primary cardiac valve tumor: Early and late results in eight patients. *J Cardiothorac Surg* 2016;11:31.
3. Märkl B, Kerwel TG, Langer E, Müller W, Probst A, Spatz H, *et al.* Elastosis of the colon and the ileum as polyp causing lesions: A study of six cases and review of the literature. *Pathol Res Pract* 2008;204:395-9.
4. Tosios KI, Economou I, Vasilopoulos NN, Koutlas IG. Elastofibromatous changes and hyperelastosis of the oral mucosa. *Head Neck Pathol* 2010;4:31-6.
5. Gambichler T, Altmeyer P, Stücker M. Cerebriform elastoma: An unusual presentation of actinic elastosis. *J Am Acad Dermatol* 2005;52:1106-8.
6. Paillas J, Dupuis J, Fabre M, Marais M, Vieillefond A. Bronchial elastoma. *Arch Anat Cytol Pathol* 1976;24:469-71.
7. Algin O, Gökalp G, Topal U. Signs in chest imaging. *Diagn Interv Radiol* 2011;17:18-29.
8. Wilson RW, Kirejczyk W. Pathological and radiological correlation of endobronchial neoplasms: Part I, benign tumors. *Ann Diagn Pathol* 1997;1:31-46.
9. Pandey K, Vaidya PJ, Kate AH, Chavhan VB, Jaybhaye P, Patole K, *et al.* Bronchoscopic and surgical management of rare endobronchial tumors. *J Cancer Res Ther* 2016;12:1093-7.
10. Starcher BC. Elastin and the lung. *Thorax* 1986;41:577-85.
11. Arrigoni MG, Woolner LB, Bernatz PE, Miller WE, Fontana RS. Benign tumors of the lung. A ten-year surgical experience. *J Thorac Cardiovasc Surg* 1970;60:589-99.
12. Fork HE, Sanchez RL, Wagner RF Jr., Raimer SS. A new type of connective tissue nevus: Isolated exophytic elastoma. *J Cutan Pathol* 1991;18:457-63.
13. Schaffenburg WC, Fernelius C, Arora NS. Buschke-Ollendorff syndrome presenting as a painful nodule. *JAAD Case Rep* 2015;1:77-9.
14. Lazar A, Wang W. Diseases of collagen and elastic tissue. *McKee's Pathology of the skin*. 4<sup>th</sup> ed. Elsevier, 2012. p. 935-66.
15. Gupta R, Naji W, Jindal A, Patel BS, Mittal G, Labelle A. A rare case of isolated pauci-immune pulmonary capillaritis. *Int J Crit Illn Inj Sci* 2014;4:319-20.
16. Gopaldas RR, Atluri PV, Blaustein AS, Bakaeen FG, Huh J, Chu D. Papillary fibroelastoma of the aortic valve: Operative approaches upon incidental discovery. *Tex Heart Inst J* 2009;36:160-3.
17. Gupta R, Robison LR. Actinomycosis with septic pulmonary emboli developing after dental abscess. *Mayo Clin Proc* 2013;88:e137.
18. Klarich KW, Enriquez-Sarano M, Gura GM, Edwards WD, Tajik AJ, Seward JB. Papillary fibroelastoma: Echocardiographic characteristics for diagnosis and pathologic correlation. *J Am Coll Cardiol* 1997;30:784-90.
19. Hobbs CM, Burch DM, Sobin LH. Elastosis and elastofibromatous change in the gastrointestinal tract: A clinicopathologic study of 13 cases and a review of the literature. *Am J Clin Pathol* 2004;122:232-7.
20. Borczuk AC. Benign tumors and tumorlike conditions of the lung. *Arch Pathol Lab Med* 2008;132:1133-48.