Bronchial Leiomyoma: Importance of Preoperative Biopsy for Lung Preserving Surgery

Adil Hashim Al Kindi[1]*, Marwa Taher Al Ibrahim[2], Malyar Mohsin Al Marhoon[2], Faiza Abdullah Al Kindi[3], Abdulmageed Salem[1] and Radiya Al Ajmi[4]

[1]Cardiothoracic Surgery, Sultan Qaboos University Hospital, Muscat, Oman
[2]Oman Medical Speciality Board
[3]Radiology Department, Royal Hospital
[4]Pathology Department, Sultan Qaboos University Hospital

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***Corresponding author:** alkindi2001@gmail.com

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Abstract:

Tracheo-bronchial leiomyoma is a rare tumor of the airway. They arise from lower respiratory tract tissue of the brochi, trachea and lung tissue. Symptomatology is based on the degree of endoluminal bronchial obstruction and surgical resection is generally the mainstay of treatment. We are presenting a 33 years old gentleman who suffered from chronic cough and breathlessness for 2 years caused by large endobronchial leiomyoma diagnosed by pre-operative biopsy. The tumor was surgically resected through bronchotomy and complete preservation of the lung parenchyma. We stress the importance of definitive pre-operative diagnosis of this rare tumor so that lung preserving surgical techniques could be employed.

Introduction:

Leiomyoma of the tracheo-bronchial tree is a rare tumor of the respiratory tract accounting for <2% of benign tumors of the lung.[1] Symptoms depend on the degree of airway obstruction.[1,2,3,4] Bronchial leiomyoma has a benign course and local complete resection of the tumor is therapeutic.

We report a patient with symptomatic endobronchial leiomyoma that was resected employing lung parenchymapreserving techniques. In this report, we stress the importance of pre-operative diagnostic workup for successful lung preservation.

Case report:

A previously healthy, 33 years-old-gentleman, presented with two years history of cough and breathlessness. He was diagnosed with bronchial asthma and was treated accordingly but showed no improvement. In the last one year, he was having recurrent chest infections requiring frequent antibiotics.

On examination, he had decreased air entry in the right middle and lower lung zones. CT-scan of the Chest showed ovoid shaped endobronchial lesion in the right main bronchus, extending into the trachea causing total collapse of the right middle and lower lobes (Figure 1).

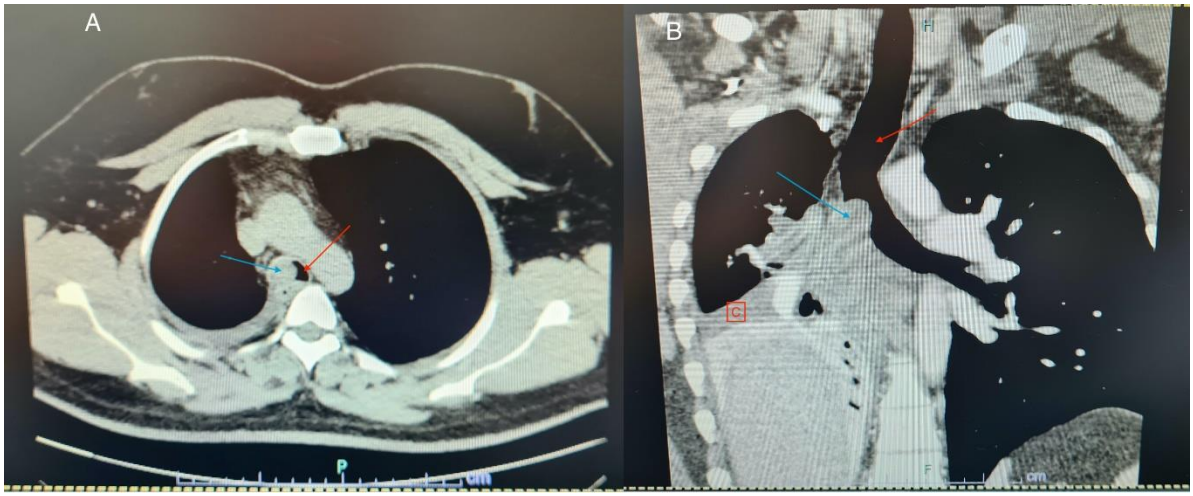

Figure 1: A) Axial views of chest CT-scan showing an ovoid mass (Blue arrow) obstructing the right main bronchus bulging into the carina and distal trachea (Red arrow) B) Coronal views showing collapse of the right middle and lower lobes (C) and mediastinal shift to the right

Bronchoscopy confirmed the presence of a smooth, lobulated oval mass bulging into the distal trachea from right main bronchus. It was impossible to pass the bronchoscope beyond the mass. The mass was pale, non-pulsating and appeared like an endobronchial carcinoid (Figure 2). Punch biopsies were taken. However, the mass did not bleed as is usually the case with endobronchial carcinoid. The biopsies revealed polypoid/nodular fragments of tissue lined

by benign bronchial epithelial cells. The cores showed spindle cells in edematous stroma. The spindle cells were positive for desmin, H. Caldesmon and SMA. The mass was diagnosed as endobronchial leiomyoma.

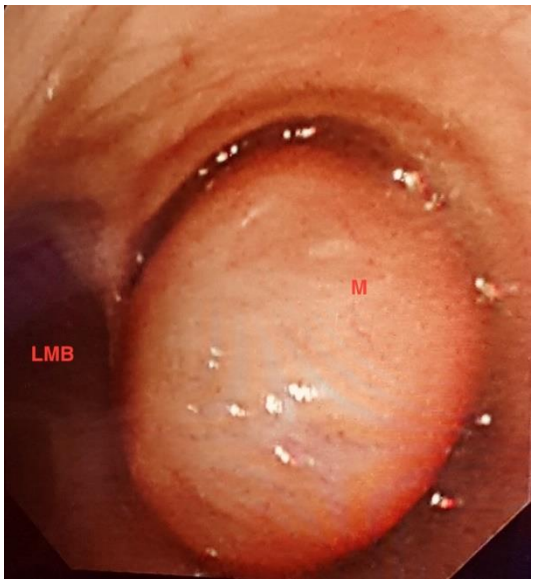

Figure 2: Bronchoscopic view of the mass (M) within the right main bronchus bulging into the trachea resulting in near total occlusion of the right main bronchial lumen. Left Main Bronchus (LMB)

Patient underwent mini-right posterio-lateral thoracotomy. The middle and lower lobes appeared normal despite repeated infections. However, it remained collapsed despite attempted positive ventilation. Bronchotomy of the right main bronchus extending to the bronchus intermedius was performed. The endobronchial mass bulged out of the bronchotomy (Figure 3A). It had a wide base that was attached to the bronchus intermedius. We resected the attachment together with a small margin of normal bronchial wall. The bronchial opening was closed with interrupted sutures to ensure that the bronchus was not narrowed. Suture line was reinforced with a parietal pleura flap. The right lung inflated fully under positive pressure ventilation.

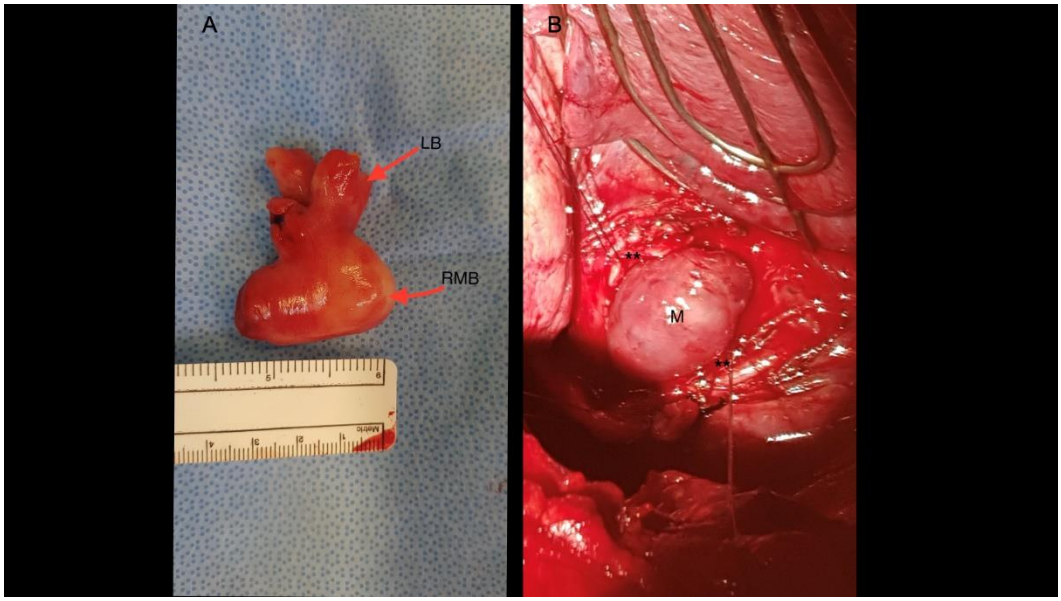

Figure 3: A) Intra-operative image showing the Right Main bronchus open with the edges retracted with stay sutures (**) and the mass appears to be bulging out of the bronchotomy (M) B) Gross appearance of the mass after surgical removal. The mass has taken the shape of the right main bronchus (RMB) and the lobar bronchus (LB).

Postoperatively, the patient did well and the right lung remained fully expanded. The mass was sent for histopathological examination. Macroscopic examination revealed a well circumscribed nodular mass measuring 4x3.5x2 cm with smooth outer surface (Figure 3B). Cut section showed homogenous, solid, white whorled surface. The microscopy showed a well circumscribed variably cellular spindle cell neoplasm partially lined by respiratory type epithelium. It is composed of fascicles of bland spindle cells having cigar shaped nuclei and abundant eosinophilic cytoplasm. The stroma shows variable hyalinization with thin and thick walled blood vessels and scattered mast cells. There is no atypia, increased mitosis or coagulative necrosis. Immunohistochemical stains were performed, the neoplastic spindle cells were strongly positive for SMA, Desmin and H-caldesmon (Figure 4A & B). They are negative for CK AE1/AE3, CD34, S100 and STAT 6. The final histopathological diagnosis was endobronchial leiomyoma.

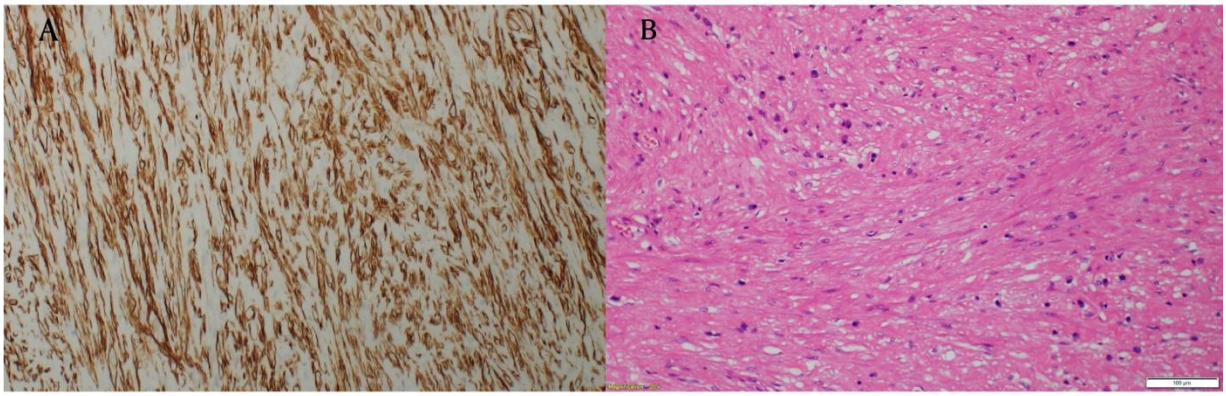

Figure 4: A) Microscopy of the neoplasm shows a cellular proliferation of intersecting fascicle of bland spindle cells. No nuclear atypia, necrosis or increased mitoses are identified. (Haematoxylin & Eosin stain at x20 magnification). **B:** These neoplastic cells are positive for H-caldesmon immunostain (at x10 magnification).

At 1 year follow up, his symptoms had completely resolved. Imaging showed fully expanded lung with no evidence of recurrence.

Discussion:

51% of pulmonary leiomyoma occur in the parenchyma, 33% in the bronchial tree and only 16% in the trachea . They present in adults with a slight female pre- ponderance and have been reported in patients with EBV, HIV/AIDS and cellular immunodeficiency. [4,5] Symptoms depend on the site and degree of airway compromise. They can present with cough, wheezing, shortness of breath and/or chest pain.[1,2,3] Parenchymal disease is asymptomatic and is usually an incidental finding.[1]

Diagnosis requires imaging, bronchoscopy and histopathological biopsy. There are no pathognomonic radiological features of leiomyoma. It may show atelectasis or hyperinflation of the lung due to airway obstruction. CT -scan has almost 100% sensitivity for detecting the mass but is not specific. The mass shows a homogeneous, smooth or lobulated appearance with diffuse enhancement and a well delineated margin. However, such features are shared with leiomyosarcoma, lipoma and neurogenic tumors.[3] Both carcinoid and leiomyoma appear lobulated, central and may have eccentric calcification. [6]MRI provides no clear diagnostic advantage over CT[5]Thus, imaging alone is not sufficient to differentiate benign from malignant endobronchial pathologies.

Hence it is important to perform bronchoscopy to visualize the location,shape of the base of the mass, get a biopsy for definitive histological diagnosis and plan definitive therapy.[1,3] The risks associated with endobronchial biopsy is very low. The risk of mortality is almost nil while the complications are mostly minor and self-limiting and do not exceed 10% in most cases.[7]

Cases of endobronchial resection of the tumor have been described. Methods used include Argon Plasma Cogulation (APC), Nd:YAG laser, biopsy forceps extraction, electrocautery and snare removal. [2] The tumor is first mechanically removed by forceps or snare followed by laser ablation of the stump. If the tumor is for ablation only, it can be vaporized with Nd:YAG laser using flexible bronchoscope.[8] These are less invasive techniques and contribute to faster and less eventful recovery. However, the use of such minimally invasive endoluminal methods is limited to technical and anatomical factors. Polypoid lesions and broad based lesions are not suitable as complete resection is not possible as they may extend outside the cartilage.[2] Fell et al, have reported tumor growth of these lesions with doubling in size of an endobronchial lesion within 18 months, potentially predisposing to hemorrhage from neovascularization and making long-term bronchoscopic follow-up of treated lesions imperative. [9]

Endoluminal resection therapies require repeated sessions, life time surveillance and have a high recurrence rate. [1] As such, they should be reserved for palliative cases not fit for surgery and for pediatric cases to delay surgical resection. [2]

Definitive treatment requires complete excision of the mass to prevent recurrence.[1] Many of the patients present with advanced lung destruction from recurrent infections requiring major lung resection such as lobectomy or even pneumonectomy.[2,3] This emphasizes the need for early intervention and removal of the tumor before airway obstruction results in repeated lung infections and destruction. Every effort should be made to preserve as much lung parenchyma as possible as this is a benign pathology and most patients are young with a median age of 46 years.[1] Lung preserving techniques such as wedge resection, segmentectomy, bronchoplasty and sleeve resection are important in young patients and those with limited lung reserve. Peri-operative and long term morbidity and mortality has been associated with more extensive lung resection.[10]Thus, limiting resection to that needed to treat the pathology is very important.

Conclusion:

Endobronchial leiomyoma is a rare benign pathology. It is important to make a definitive pre-operative diagnosis as similarly appearing pathologies require more aggressive intervention. Limited but complete excision of the mass should always be the goal.

In this case, the patient underwent excision of leiomyoma with complete preservation of the lung parenchyma that was only possible because of the definitive diagnosis on pre-operative biopsy.

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List of Abbreviations:

- SMA: Smooth Muscle Actin
- CK AE1/AE3: Cytokeratin AE1/AE3

- STAT-6: Signal transducer and activator of transcription **6**
- EBV: Epstein-Barr Virus
- HIV: Human Immunodeficiency Virus
- AIDS: Acquired ImmunoDeficiency Syndrome
- MRI: Magnetic Resonance Image
- Nd: YAG laser: Neodymium-Doped Yttrium Aluminum Garnet Laser