A 73 year-old woman, a lifetime non-smoker, presented to the pulmonary clinic with chronic dyspnea on exertion and cough. Physical exam was unremarkable and pulmonary function testing showed normal spirometry. A chest radiograph revealed calcified mediastinal adenopathy and increased density in the right middle lobe region (Figure 1). A computed tomography scan of the chest revealed significant narrowing of the right middle lobe bronchus with partial atelectasis and prominent calcified
mediastinal lymphadenopathy (Figure 2). Bronchoscopy showed no endobronchial
lesions but there was evidence of extrinsic compression surrounding the right middle
lobe orifice. An endobronchial biopsy revealed noncaseating granulomas.
Bronchoscopy cultures and cytology were negative and this was presumed to be from a
previous infection with histoplasmosis given the patient's long-term residence in an
endemic area. Given chronic narrowing of right middle lobe bronchus with persistent
atelectasis of the right middle lobe, the patient was diagnosed with right middle lobe
syndrome. She was started on combination therapy with a long-acting beta agonist and
inhaled corticosteroid with complete resolution of her symptoms.

Right middle lobe syndrome (RMLS) is defined as recurrent or chronic atelectasis of the
right middle lobe. Although more commonly described in children, it is becoming more
prevalent in adults with a predilection for women. There are two distinct types of
pathophysiology- obstructive and non-obstructive. Obstructive pathophysiology is
defined when there is an endobronchial lesion or extrinsic compression of the middle
lobe bronchus by lymphadenopathy (as in our case) or a tumor. Non-obstructive
pathophysiology occurs when there is recurrent infection or inflammation leading to
bronchiectasis and scarring. Certain anatomical characteristics, including the acute
take-off angle of the right middle lobe bronchus create poor conditions for drainage and
collateral ventilation (1).

Symptoms of RMLS include chronic or recurrent cough, dyspnea, wheezing and
recurrent infections. High resolution computed tomography of the chest is the gold
standard for imaging, as this will show narrowing of the right middle lobe orifice along
with etiologies of extrinsic compression (Figure 2). Patients suspected of having RMLS
warrant a bronchoscopy to evaluate for patency of right middle lobe bronchus, to
exclude malignancy and for evaluation of infectious etiologies (1). The treatment of
RMLS includes bronchodilator therapy along with mucolytics, chest physiotherapy and
antibiotics if bronchiectasis is problematic. Lobectomy may be warranted if malignancy
is diagnosed, aggressive medical management fails or hemoptyisis occurs (2).

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References

1. Gudbjartsson T, Gudmundsson G. Middle lobe syndrome: a review of
[CrossRef] [PubMed]

2. Einarsson JT, Einarsson JG, Isaksson H, Gudbjartsson T, Gudmundsson G. Middle
lobe syndrome: a nationwide study on clinicopathological features and surgical
treatment. Clin Respir J. 2009 Apr;3(2):77-81. [CrossRef] [PubMed]