Case report

Case report: Pulmonary amyloidosis in two patients with chronic obstructive lung disease (COPD)

Haya Mardahayev, Sara Mohammed, Khalid Moddathir Mohmmed*, Hala Rizalla, James Hayes
Cavan and Monaghan Hospital, Ireland RCSI Group

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ABSTRACT

We present two cases of patients with chronic obstructive lung disease (COPD) who developed different forms of pulmonary amyloidosis. In both cases malignancy was considered as primary diagnosis. Transthoracic biopsy confirmed pulmonary amyloidosis.

In the first case the patient presented with progressive dyspnea over a two years period. Initial assessment was consistent with a diagnosis of COPD but progressive changes in symptoms and lung functions and subsequently CT Thorax revealed possible airway obstruction. Bronchoscopy confirmed an obstructive lesion initially considered to be malignant but was found to be due to tracheobronchial amyloid (TBA).

Our second case presented with symptoms and signs consistent with COPD. Follow up chest X-rays revealed a pulmonary nodule which on CT examination was considered to be malignant. Transthoracic biopsy confirmed pulmonary amyloidosis.

Although it is a rare condition amyloid disease should to consider as the part of the differential diagnosis in COPD patients who present with signs and symptoms consistent with pulmonary malignancy.

1. Introduction

Pulmonary amyloid may present as a nodular form, a diffuse interstitial disease or localized to transbronchial tree [1&2]. It may be a relatively benign process or form a part from a system disorder [3]. Clinical presentation can be varied and is dependent on the site and extend of tissue involvement. Diagnosis is dependent on appropriate histology and workup should include careful assessment for systemic disorders.

Patients may require localized or systemic treatment with lifelong surveillance. We present two patients with COPD who were found to have lesion suspicious for pulmonary malignancy but in whom pulmonary amyloidosis was the final diagnosis. While unusual pulmonary amyloid disease remains part of the differential diagnosis for COPD patients with lesions suggesting underlying malignancy.

2. Case presentation

2.1. Case report 1

A sixty four year old male, with a history of hypertension and sleep apnoea who presented with progressive dyspnea, copious sputum with background of 20 pack years smoking. Initial pulmonary function test (FVC values 61%, FEV1 45% and DLCO 84% of predicted. FEV1/FVC ratio measured at 0.58) [Fig. 4].

And chest X-ray was consistent with COPD. However over a 12 month period his symptoms deteriorated greater than expected.

Subsequent CT Thorax revealed mild emphysematous changes, lung parenchymal cysts, and minor traction bronchiectasis consistent with obstructive airway in addition to bronchial wall thickening suspicious for malignancy [Fig. 1]. Bronchoscopy confirmed narrowing in the right main bronchus [Fig. 2]. Biopsy was uncomplicated confirmed tracheobronchial amyloidosis [Fig. 3].

2.2. Case report 2

A sixty sex year old male has a long history of COPD and asthma with frequent exacerbations. He has been lifelong smoker and worked as a journalist. He was referred by his GP to the respiratory clinic due to worsening symptoms.

He was admitted for symptoms management and further workup. Chest X-Ray showed only background changes of COPD. However over a 12 month period his symptoms deteriorated greater than expected.

Subsequent CT Thorax revealed mild emphysematous changes, lung parenchymal cysts, and minor traction bronchiectasis consistent with obstructive airway in addition to bronchial wall thickening suspicious for malignancy [Fig. 1]. Bronchoscopy confirmed narrowing in the right main bronchus [Fig. 2]. Biopsy was uncomplicated confirmed tracheobronchial amyloidosis [Fig. 3].
Fig. 1. HRCT of case 1 showed right main bronchi wall thickening, mimicking bronchial malignancy and mucous plugs.

Fig. 2. Bronchoscopy was done and reported as irregular proximal RMB, early changes in LMB highly suspicious of malignancy, normal cords and trachea.

Fig. 3. (A) A low power view showed morphous material in keeping with amyloid deposition in the submucosa (B) Congo red staining on transbronchial right middle bronchus biopsy demonstrated apple green birefringence.
bilateral. The largest is subpleural indeterminate nodule/plaque of right upper zone measuring 18 × 9mm. There was some bronchial basal wall thickening and moderate Centro-lobular emphysematous changes noted. The possibility of malignancy with lung metastasis was raised.

The PFT’s were performed and demonstrated FEV1 22%, FVC of 45% and DLCO 105% of predicted. FEV1 and FVC ratio was noted at 0.38 which is consistent with a severe obstructive pattern [Fig. 7].

Bronchoscopy samples were taken from RML and histology reported as benign respiratory cytology. Furthermore, a CT guided biopsy was done and histology showed benign fibrosis with multinucleated giant cells of foreign body and hyaline changes. No malignant cells were seen. Further staining with Congo red and ISH for kappa and lambda took place. The amorphous eosinophilic material stained positively with Congo red and apple green birefringence confirming a diagnosis of amyloidosis, and debunking malignancy [Fig. 6]. However no monoclonality of light chains was seen with ISH.

3. Discussion

Amyloidosis may be considered a generic term for an heterogenous group of diseases where the end point is the deposition of abnormal fibrils of proteinaceous material. These proteins can vary in size and may form part of a systemic or local disease process [3,4]. The cornerstone of diagnosis remains obtaining biopsy material stained positive by Congo red dye using a polarized microscope which demonstrated a typical green birefringence pattern [5,6].

Pulmonary amyloidosis may present a diffuse parenchymal process which is usually part of a systemic disease. It is often a severe disease which may progress rapidly. It can be associated with pulmonary hypertension either by direct involvement of pulmonary interstitial or by associated vascular plaques. Conversely tracheobronchial amyloid and nodular pulmonary amyloidosis are more benign in presentation and prognosis [7]. The two cases presented here are typical of these two forms of amyloidosis. Here the precursor protein is thought to be synthesized and produced by local cell types such as macrophages or monocytes. In both these cases there was an association with COPD. To date we have found no association with systemic illness, blood dyscrasias or lymphomatous disease. Long term follow up will however be necessary for both these patients.

4. Conclusion

We present two different cases of local pulmonary amyloidosis. Both cases were associated with chronic obstructive pulmonary disease with signs initially considered to be due to malignancy. Although extremely rare amyloidosis may need to be considered in patient presenting with signs and symptoms of thoracic malignancy.
Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethics approval and consent to participate

Not applicable.

Conflicts of interest

The authors declare that they have no competing interests regarding the publication of this paper.

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Fig. 6. (A) Sheets of esinophilic material in keeping with amyloid deposits, admixed with multinucleated osteoclast -like giant cells lymphocytes and mucosa cells. (B) Congo red staining positivity with apple green birefringence.

Fig. 7. Case 2 PFT’s, FEV1 of 0.64, FVC of 1.68 and DLCO of 105% predictive value. FEV1 and FVC ratio was noted at 0.38 which is consistent with a severe obstructive pattern.
Availability of data and materials

Datasets used and/or analyzed for this study have been included in this published article.

Authors’ contributions

Study concept and design: KM and SA. Acquisition of data: JH, KM, SA, HR, HM Analysis and interpretation of data: KM, JH, and SA. Drafting of the manuscript: KM, and SA. Critical revision of the manuscript for important intellectual content: All authors. All authors read and approved the final manuscript.

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Abbreviations

TBA TracheoBronchial Amyloidosis
HRCT High Resolution CT scan
COPD Chronic Obstructive Pulmonary Disease
RMB Right Main Bronchus
LMB Left Main Bronchus
DLCO Diffusing capacity of the Lungs for Carbon monoxide

FEV1 Forced Expiratory Volume-one second
FVC Forced Vital Capacity
PFT Pulmonary Function Test
ISH In-Situ Hybridisation techniques

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2019.100897.

References