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Nodular pulmonary light chain deposit disease: An unexpected finding on histology

Muazzam Tahir, Sean Galvin

ABSTRACT

Introduction: Nodular pulmonary light chain deposit disease is a rare histological diagnosis which can be associated with poor outcome. Case Report: A 73-year-old male was incidentally found to have two pulmonary nodules. Histology following resection confirmed nodular pulmonary light chain deposit disease, a rare condition. He was subsequently found to have multiple myeloma. Conclusion: Pulmonary light chain deposit disease has two forms namely diffuse and nodular. Diffuse form has poor prognosis and nodular is associated with lymphoproliferative and/or plasma cell dyscrasia in half of the patients, hence early diagnosis is crucial.

Keywords: Immunoglobulin light chain, Kappa, Lambda, Light chain deposition disease, Pulmonary nodule

How to cite this article

Tahir M, Galvin S, Nodular pulmonary light chain deposit disease: An unexpected finding on histology. Edorium J Respir Med 2016;2:1–3.

Muazzam Tahir[1], Sean Galvin[2]

Affiliations: [1]MBBS, Wellington Regional Hospital, registrar cardiothoracic surgery, cardiothoracic department, Wellington Regional Hospital, Wellington, New Zealand; [2]FRACS, Wellington Regional Hospital, Consultant surgeon cardiothoracic surgery, cardiothoracic department, Wellington Regional Hospital, Wellington, New Zealand.

Corresponding Author: Muazzam Tahir, Flat no 504, 20 Hanson Street, Mount Cook, Wellington, New Zealand, E-mail: drmuazzamtahir@gmail.com

Received: 16 March 2016
Accepted: 25 May 2016
Published: 20 July 2016

Article ID: 100002R05MT2016

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doi:10.5348/R05-2016-2-CR-1

INTRODUCTION

Pulmonary light chain deposit disease (LCDD) is a rare condition of uncertain etiology characterized by deposition of non-amyloid immunoglobulin light chains in multiple organs which do not show a fibrillar structure when examined ultra-structurally [1–3]. Differentiation of this condition from amyloidosis is important as latter may have better outcome.

CASE REPORT

A 73-year-old male was referred with two right sided pulmonary nodules, 18 and 6 mm in the right lower and middle lobes respectively. These were incidentally found on a Computed tomography (CT) scan of abdomen done in early 2015 for follows of hepatic artery aneurysm repair. Repair of hepatic artery was done in 2010 and since then he has been having two yearly follow up abdominal CT scans. A further CT chest showed no interval growth over three months' period. He was referred to cardiothoracic team at Capital and Coast District Health Board, Wellington for excision of these nodules. PET scan prior to procedure again did not show interval growth over five months' period. These nodules were moderately FDG positive on the PET scan.

The patient was otherwise healthy with past history of hepatic artery aneurysm repair, hypertension and cholecystectomy. It was decided to perform wedge resection of these nodules via video assisted thoracoscopic surgery (VATS). After initial VATS technique, open approach was taken and wedge resection of right middle lobe nodule with resection of interlobar

lymph nodes was done, however, a complete right lower lobectomy was done due to proximity of lower lobe nodule to inferior pulmonary vein. He was discharged home after an uncomplicated recovery.

Initial pathology report suggested a benign lesion consisting of amorphous material with foreign body reaction, focal calcification and a negative congo red staining. An external opinion was taken and the final histological diagnosis was nodular pulmonary light chain deposit disease. Histological features were nodular focus of eosinophilic extracellular material rimmed by multinucleated giant cells with intervening fibrosis containing a variable lymphoplasmacytic infiltrate. The extracellular material showed patchy salmon pink staining with congo red stains, no apple green birefringence under polarizing light, negative with PAS, PASD and Von Kossa stain. The material also showed deeper more diffuse brown staining with kappa light chain while little staining for lambda light chain.

Immunochemically plasma cells in the lesion were strongly CD138 and IgG positive. Scattered CD20+ B cells nodules were noted along with diffusely scatted CD3+ T cells. Diffuse sheets of B cells were not seen and majority of histiocytes stain with CD68. The kappa light chain predominance of the plasma cells is seen with kappa and lambda light chain immunostains.

Due to association of this condition with lymphoproliferative and/or plasma cell dyscrasia, our patient was followed with a CT scan of chest at sixth month interval which did not show disease recurrence but he is recently diagnosed with multiple myeloma and is on treatment for it.

DISCUSSION

Immunoglobulin light chains can deposit in tissues in two forms; AL amyloidosis and LCDD [4]. AL amyloid results from proteolytic degradation of monoclonal light chains into twisted beta pleated sheets with fibrillar appearance [5]. Amyloid has a characteristic tinctorial property of orangeophilia with congo red under light microscopy, apple green birefringent under polarization and metachromasia with crystal violet or methyl violet [6]. On the contrary, light chains in LCDD fail to form beta pleated sheets with granular deposits that lack the fibrillar pattern [6]. The deposits of LCDD also lack the tinctorial property of amyloid with the congo red dye, often are the variable region of the kappa chains and do not contain the p component [6]. Between 80% and 90% of patients with either AL amyloid or LCDD have a detectable monoclonal immunoglobulin or free light chain in either serum or urine [5].

Amyloid deposits in the lung have been classified as parenchymal (nodular or diffuse) and tracheobronchial types, most of which are AL type of amyloid [7, 8]. Diffuse parenchymal form of amyloid is frequently associated with plasma cell dyscrasia and has grim prognosis with median survival of two years after diagnosis [6]. Nodular parenchymal amyloidosis has better prognosis with only 10% have an associated plasma cell dyscrasia [6].

Light chain deposit disease was first described by Randall et al. [6, 9] in 1976 involving the kidneys with characteristic appearance as a nodular glomerulosclerosis [6], however, LCDD has been identified as a systemic disease with deposits involving many organs [6]. Pulmonary involvement is extremely rare and was first described by Kijner and Yousem in 1988 [10]. Due to histologic resemblance to amyloid by light microscopy, this entity is underdiagnosed [6]. There are two forms of pulmonary LCDD namely diffuse and nodular which were first categorized in 2007 by Bhargava et al. [6]. Chest X-ray usually shows pulmonary nodules which vary in size from a few millimeters to a few centimeters. Affected individuals may be asymptomatic or they can have shortness of breath or cough and develop severe respiratory failure [11].

The diffuse form on hematoxylin and eosin staining shows little to marked thickening of the basement membrane of alveolar, bronchiolar and vessel walls with preserved pulmonary architecture under light microscopy [6]. Diffuse form of both LCDD and amyloidosis share many features like similar mean ages of patients, male to female ratio and frequent association with plasma cell dyscrasia and multi organ involvement [6]. The clinical outcome for both diseases is poor with frequent renal failure and mortality [6]. The nodular form of LCDD and amyloidosis has similar mean ages of patients. Multi organ involvement and death are uncommon in both diseases [6].

CONCLUSION

In summary, diffuse form of pulmonary light chain deposit disease (LCDD) has poor prognosis and differentiating it from diffuse amyloid disease is not crucial as clinical outcome is poor for both diseases. The nodular pulmonary LCDD should be distinguished from nodular amyloidosis as 50% of patients with nodular LCDD have an associated lymphoproliferative and/or plasma cell dyscrasia and may develop renal failure as compared to nodular amyloidosis with better outcome. There is no consensus in literature about follow-up of these patients, however, we think that yearly follow-up with a chest CT scan and hematology review is appropriate for at least five years in patients with nodular pulmonary light chain deposition disease.

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Author Contributions

Muazzam Tahir – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sean Galvin – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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