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Case report

CT findings in extensive tracheobronchial amyloidosis

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Abstract. Primary pulmonary amyloidosis is a rare disorder that appears in three forms: tracheobronchial, nodular parenchymal, and diffuse parenchymal. We report the case of a 46-year-old women with extensive tracheobronchial amyloidosis which presented with a 2-year history of dyspnea and with signs of severe fixed obstruction in pulmonary function tests. Computed tomography of the thorax demonstrated marked thickening of the trachea and the central bronchial tree with substantial narrowing of the main, lobar, and segmental bronchi. Transbronchial specimen showed typical birefringence under polarizing microscope after staining with Congo Red. We did not find hints for systemic amyloidosis.

Key words: Amyloidosis – Radiography – Bronchial disease – Tracheal disease – CT

Introduction

Amyloidosis is a rare disease caused by extracellular deposition of amyloid, a chemically diverse insoluble glycoprotein in a beta-pleated sheet configuration. In histologic sections accumulation within organs shows characteristic green birefringence under polarizing microscope after staining with Congo Red [1–5]. Amyloid can infiltrate virtually all organ systems and may occur either idiopathically without underlying disease – termed primary amyloidosis – or as a part of a widespread systemic disease in association with inflammatory, hereditary (e.g., familial Mediterranean fever), or neoplastic conditions (especially of the myeloproliferative system) – termed secondary amyloidosis. Respiratory system is frequently involved in systemic amyloidosis, estimates being from 30 % to more than 90 % in the pathologic literature [6–9]. Clinical symptoms and radiographic demonstration of pulmonary involvement, however, are much less common [10]. Primary pulmonary amyloidosis, i.e., amyloid deposition restricted to the respiratory tract, is a very rare disease and may be divided into the following groups: tracheobronchial, nodular parenchymal, and diffuse parenchymal [11–14]. Tracheobronchial amyloidosis is the most frequent form of primary pulmonary amyloidosis. Although localized lesions do occur [15–19], tracheobronchial amyloidosis characteristically demonstrates diffuse involvement of the tracheobronchial tree with submucosal deposition of amyloid plaques and nodules that may cause atrophy of normal wall structures [3, 20–23]. Large lesions may infiltrate all layers of the tracheobronchial wall, erode cartilage, and lead to extensive airway stenosis. We report the radiographic and CT findings of a patient with diffuse tracheobronchial amyloidosis with extensive amyloid deposition in the central airways.

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Case report

In January 1995 a 46-year-old woman was referred to us with a 2-year history of intermittent wheeze that was nonresponsive to anti-asthmatic treatment. She was on 25 mg oral prednisolone, theophylline, and inhalative albuterol.

She presented in good health with mild resting dyspnea without worsening on exertion. On physical examination she was obese (178 cm, 85 kg) and had a predominant inspiratory stridor. No obvious wheezing was detected. Laryngeal examination did not reveal any reason for the stridor.

The lung function test showed a dramatic limitation of the peak in- and expiratory flow rates. Flow volume curve and body plethysmography were consistent with central airway stenosis. Contrary to chronic obstructive lung disease or asthma, the obstruction was fixed in every phase of in-and expiration and nonresponsive to bronchodilatators (i.e., inhalation of 400 ug Fenoterol). A chest radiograph showed distinct bronchial cuffing.

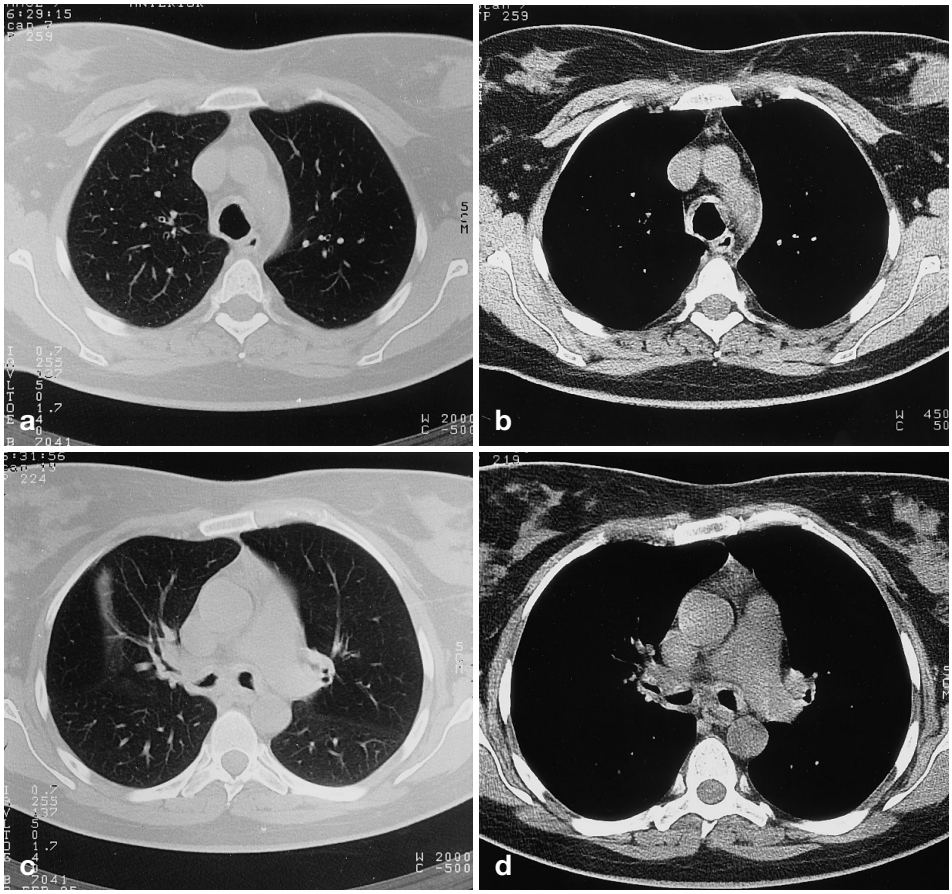

Fig. 1 a–d. Computed tomography in transverse section (slice thickness 5 mm, 135 kV, 180 mAs, high resolution). **a, c** Lung and **b, d** mediastinal window setting: CT scan shows a circumferential thickening of both tracheal and bronchial walls. Narrowing of the right upper lobe bronchus is clearly depicted **(c, d)**. Lung window settings did not show any parenchymal abnormalities

A CT examination of the lung (Somatom Plus S, Siemens, Erlangen, Germany; 180 mAs, 125 kV, 5-mm slice thickness, ultra-high-resolution algorithm, 10-mm table feed) showed circumferential thickening of the trachea (beginning from the subglottic level) and of main, lobar, segmental, and subsegmental bronchi with narrowing of their lumen (see Fig. 1). Mural nodulation of the tracheobronchial wall was visible. There was neither evidence of nodular or diffuse parenchymal involvement nor postobstructive parenchymal abnormalities.

Fiberoptic bronchoscopy showed a completely irregular surface of the bronchial mucosa with prominent reddish plaques, accompanied by whitish membranes and ulcerative changes with bleeding tendency, the so-called rock garden appearance. These changes extended 6 cm distal from the cricoid cartilage through the whole of the trachea into the ostia of the segmental bronchi. Concentric irregular narrowing of both main bronchi was seen with marked expiratory collapse involving the whole visible bronchial tree. The lumen of the right upper lobe bronchus was extremely narrow.

Biopsy specimen of the epithelium showed deposits of fibrin, moderate lymphocyte infiltration, and ample eosinophilic homogeneous mass in the connective tissue. After staining with Congo Red this material showed the typical birefringence under polarizing microscope as found in amyloid.

There was no clinical or laboratory evidence of systemic amyloidosis. No deposition of amyloid was found in pathologic specimens obtained from a rectum biospy. Due to the poor inflammatory activity seen in the specimens, no immunosuppression was implemented. In the state of acute bronchial infection high-dose prednisolone and antibiotic treatment were performed; moreover, nasal continuous airway pressure helped to maintain the airway's patency for several days.

The involvement of the peripheral bronchial tree forbids tracheal transplantation, and because of complete tracheal involvement, now including the larynx as well, lung transplantation is impossible. Therefore, repeated YAG laser coagulation in the trachea and bronchial tree and CO_2 laser coagulation of the larynx seemed to be the only therapeutic option. It was performed repeatedly between March and December 1996. Stabilization and improvement of the clinical situation and of lung function parameters was seen and could be maintained from December 1996.

Discussion

Although diffuse tracheobronchial amyloidosis is the most common appearance of primary pulmonary amyloidosis, it is still a rare disease. Thompson and Citron [24] reviewed the international literature in 1983 totaling 67 cases of tracheobronchial amyloidosis. Hui et al. [16] described 10 cases of diffuse tracheobronchial amyloidosis in a retrospective clinicopathologic study of 48

cases from the files of the U.S. Armed Forces Institute of Pathology. Males are more often affected by the disease than females [3].

Patients with tracheobronchial amyloidosis show commonly poor clinical signs. Clinical features are cough, stridor, or wheezing and hemoptysis; therefore, diagnosis is often late.

There are only a few reports on imaging findings in tracheobronchial amyloidosis [7, 10, 17, 20, 25, 26]. In the chest radiograph bronchial wall thickening and irregular narrowing may be seen. With CT the submucosal deposits of amyloid are of soft tissue density. The lumen of trachea and bronchi appears narrowed because of the thickening of the wall [27, 28].

Thickening of the tracheal and bronchial walls can be clearly depicted with CT; also bronchial lumen narrowing as well as bronchiectasis, atelectasis, or poststenotic pneumonia can be demonstrated with CT. As was the case in our patient, the CT findings in severe tracheobronchial amyloidosis are highly suggestive of this disease; the final diagnosis was obtained by bronchoscopic biopsy.

Radiologically, there are only few differential diagnoses that have to be considered; one is tracheobronchiopathia osteoplastica, but it is not certain whether or not it was only a late sequela of diffuse tracheobronchial amyloidosis, as Thompson and Citron [24] suggest. Computed tomography shows thickening of the tracheal wall with irregular calcifications protruding into the lumen from the anterior and lateral walls (cartilaginous portions) which was considered as a pathognomonic finding [27, 28]. Another differential diagnosis seems to be severe chronic tracheobronchitis as associated with colitis ulcerosa showing circumferential thickening of tracheobronchial walls [27–29]. Thickening of tracheobronchial wall as seen in relapsing polychondritis caused by recurrent episodes of inflammation [30] seems to be another differential diagnosis.

Besides its role in the diagnosis of tracheobronchial amyloidosis, CT clearly depicts the extent of the disease and its potential postobstructive pulmonary complications. As repeated laser coagulation therapy currently is the therapy of choice in patients with extensive tracheobronchial amyloidosis, CT of the central airways can be used as a guiding tool before bronchoscopic interventions, and may be used for follow-up after laser coagulation therapy.

References

1. Böhmer R, Missmahl HP (1981) Amyloidose. Dtsch Ärztebl 78: 2173–2178
2. Gertz MA, Kyle RA (1989) Primary systemic amyloidosis: a diagnostic primer. Mayo Clin Proc 64: 1505–1519
3. Schäfer H, Burkhardt A (1983) Stoffwechselstörungen C.II. Amyloidose. In: Blümcke S, Burkhardt A, Doerr W, Fasske E, Gebbers JO, Hartung W, Herbst R, Könn G, Mittermayer C, Morgenroth K, Müller KM, Oellig WP, Pfannkuch F, Schäfer H, Schejbal V, Vogel M (eds) Pathologie der Lunge Spezielle pathologische Anatomie; Bd 16/II. Springer, Berlin Heidelberg New York
4. Scott PP, Scott WW Jr, Siegelman SS (1986) Amyloidosis: an overview. Semin Roentgenol 21: 103–112
5. Utz JP, Swensen SJ, Gertz MA (1996) Pulmonary amyloidosis. The Mayo Clinic Experience from 1980 to 1993. Ann Intern Med 124: 407–413
6. Brauner GJ, Bazza FA, Mihm MC (1974) Acquired bullous disease of the skin and solitary amyloidosis of the lung. Am J Med 57: 978–986
7. Breuer S, Simpson GT, Rubinow A, Skinner M, Cohen AS (1985) Tracheobronchial amyloidosis: treatment by carbon dioxide laser photoresection. Thorax 40: 870–871
8. Colby TV, Carrinton CG (1988) Infiltrative lung disease. In: Thurlbeck WM (ed) Pathology of the lung. Thieme, New York, pp 489–493
9. Desai RA, Mahajan VK, Benjamin S, Vanordstrand HS, Cordasco EM (1979) Pulmonary amyloidoma and hilar adenopathy: rare manifestation of primary amyloidosis. Chest 76: 170–173
10. Urban BA et al. (1993) CT evaluation of amyloidosis: spectrum of disease. Radiographics 13: 1295–1308
11. Ayuso MC, Gilabert R, Bombi JA, Salvador A (1987) CT appearance of localized pulmonary amyloidosis. J Comput Assist Tomogr 11: 197–199
12. Campbell RE, Barone CA, Makris AN, Miller DA, Mohuchy T, Putnam SG, Schroeder KG, Standiford KN, Stewart DW (1993) Image interpretation session. Primary pulmonary amyloidosis. Radiographics 14: 204–205
13. Graham CM, Stern EJ, Finkbeiner WE, Webb WR (1992) High-resolution CT appearance of diffuse alveolar septal amyloidosis. AJR 158: 265–267
14. Wilson SR, Sanders DE, Delarue NC (1976) Intrathoracic manifestations of amyloid disease. Radiology 120: 283–289
15. Cotton RE, Jackson JW (1964) Localized amyloid "tumors" of the lung simulating malignant neoplasms. Thorax 19: 97–103
16. Hui AN, Koss MN, Hochholzer L, Wehunt WD (1986) Amyloidosis presenting the lower respiratory tract: clinicopathologic, radiologic, immunohistochemical, and histochemical studies on 48 cases. Arch Pathol Lab Med 110: 212–218
17. M'Rad S, Le Thi Huong D, Wechsler B, Monsigny M, Buthiau D, Colchen A, Godeau P (1988) Amylose tracheo-bronchique localisee. Une nouvelle observation avec etude tomodensitometrique et en resonance magentique nucleaire. Revue de la literature. Rev Pneumol Clin 44: 260–265
18. Petermann W, Barth J, Schlüter E (1987) Localised amyloidosis of central airways. Eur J Respir Dis 71: 210–212
19. Rubinow A, Celli BR, Cohen AS, Rigden BG, Brody JS (1978) Localised amyloidosis of the lower respiratory tract. Am Rev Respir Dis 118: 603–611
20. Cook AJ, Weinstein M, Powell RD (1973) Diffuse amyloidosis of the tracheobronchial tree. Radiology 107: 303–304
21. Cordler JF, Loire R, Brune J (1986) Amyloidosis of the lower respiratory tract. Clinical and pathologic features in a series of 21 patients. Chest 90: 827–831
22. Gottlieb LS, Gold WM (1972) Primary tracheobronchial amyloidosis. Am Rev Respir Dis 105: 425–429
23. Kamberg S, Loitman BS, Holtz S (1962) Amyloidosis of the tracheobronchial tract. N Engl J Med 266: 587–591
24. Thompson PJ, Citron KM (1983) Amyloid and the lower respiratory tract. Thorax 38: 84–87
25. Biggemann M, Kappes R, Hilweg D (1990) Mediastinale und bronchovaskuläre Amyloidose – ein ungewöhnlicher röntgenmorphologischer Befund. Radiologe 30: 344–346
26. Foer B de, Hermans R, Feenstra L, Baert AL (1993) CT and MRI of laryngeal amyloidosis. Fortschr Röntgenstr 159: 492–494
27. Kwong JS, Müller NL, Miller RR (1992) Diseases of the trachea and main-stem-bronchi: correlations of CT with pathologic findings. Radiographics 12: 645–657
28. Kwong JS, Adler BD, Padley SP, Müller NL (1993) Diagnosis of diseases of the trachea and main bronchi: chest radiography vs CT. Am J Roentgenol 161: 519–522
29. Ohkubo Y, Narimatsu A, Higuchi M, Suzuki K, Kohno A, Shigeta T, Takizawa Y, Nitta S (1990) CT findings of the benign tracheobronchial lesions with calcification. Rinsho Hoshasen 35: 839–846
30. Im JG, Chung JW, Han SK, Han MC, Kim CW (1988) CT manifestations of tracheobronchial involvement in relapsing polychondritis. J Comput Assist Tomogr 12: 388–391