Lung (2015) 193:875–883
DOI 10.1007/s00408-015-9791-x

STATE OF THE ART REVIEW

Respiratory Tract Amyloidosis. State-of-the-Art Review with a Focus on Pulmonary Involvement

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Received: 26 June 2015 / Accepted: 19 August 2015 / Published online: 27 August 2015

Abstract Amyloidosis is a constellation of disease entities characterized by abnormal extracellular deposition and accumulation of protein and protein derivatives, which show apple-green birefringence when stained with Congo red and viewed under polarized light. Amyloid can infiltrate virtually all organ systems and can display multiple and diverse imaging findings. Pathologically, respiratory involvement occurs in 50 % of patients with amyloidosis, and its clinical signs and symptoms vary depending on whether the disease is systemic or localized. The four main patterns of respiratory tract involvement are tracheobronchial, nodular parenchymal, diffuse alveolar septal, and lymphatic. Imaging findings of amyloidosis are nonspecific and vary in each pattern; knowledge about the disease impairment type is thus very important, and amyloidosis should be considered in the differential diagnosis of other very common diseases, such as infectious diseases, neoplasms, and vasculitis. This literature review describes the main clinical and imaging manifestations of amyloidosis, focusing on respiratory tract involvement and differential diagnosis.

Keywords Amyloidosis · Imaging · Computed tomography · Lung diseases

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Introduction

Amyloidosis is a constellation of disease entities characterized by abnormal extracellular deposition and accumulation of protein and protein derivatives. It shows apple-green birefringence when stained with Congo red and viewed under polarized light [1]. Virchow coined the term "amyloid," which means "starch-like" or "cellulose-like," in 1853 [2]. This name was derived from the ability of glycosaminoglycans of amyloid to react with iodine and sulfuric acid, which at the time was a marker for starch [3]. The disease becomes clinically significant when the diffuse form affects organ function by replacing the normal cell structure, or when the rarer focal form (amyloidoma) has a mass effect [1].

Amyloidosis can be hereditary or acquired, localized or systemic, and potentially lethal or merely an incidental finding [4]. It is found in 0.2–0.5 % of routine postmortem examinations, 10–15 % of patients with multiple myeloma, 20–25 % of patients with rheumatoid arthritis, 23.6 % of paraplegics, 26–40 % of individuals with familial Mediterranean fever, and up to 50 % of individuals with chronic tuberculosis and leprosy [5]. Cohen and Wills [5] found amyloid deposits in 16 % of 100 consecutive postmortem examinations of patients whose average age was 80 (range 56–96) years, which corroborates data from

similar studies and confirms the increased incidence of amyloidosis in elderly individuals.

Amyloid can infiltrate virtually all organ systems and can display multiple and diverse imaging findings. Pathologically, respiratory involvement occurs in 50 % of patients with amyloidosis [6], and its clinical signs and symptoms vary depending on whether the disease is systemic or localized [6]. The four main patterns of pulmonary involvement are lymphatic, diffuse alveolar septal, nodularparenchymal, and tracheobronchial, with the former two patterns seen more commonly in patients with systemic involvement and the latter two occurring more commonly in localized disease [7]. The aim of this review is to describe the main clinical and imaging manifestations of amyloidosis, focusing on respiratory tract involvement and differential diagnosis of each impairment pattern.

Pathogenesis

An extracellular amyloid deposit is invariably made up of three components. The fibrillar protein is made of fibrils with thicknesses of 7–10 μm that are arranged primarily in a secondary β-pleated sheet structure. The other two components serum amyloid P (SAP) and charged glycosaminoglycans, which are ubiquitous in all types of amyloidosis and likely contribute to the formation of the β-pleated sheet structure [1]. This twisted β-pleated sheet configuration of an amyloid deposit results in its characteristic appearance when stained with Congo red and accounts for its relative resistance to normal proteolytic digestion, which promotes accumulation [8].

Amyloid is deposited within parenchymatous and mesenchymatous tissues and around blood vessels, where it does not evoke an inflammatory response, but compresses and replaces normal tissue and interferes with cellular and organ physiology [9]. Its fibrils may also be cytotoxic, possibly by promoting apoptosis [10], which could account for their damaging consequences in Alzheimer disease (in which few deposits develop) [4].

The cause of amyloidosis is unknown. Some researchers have postulated that it represents a derangement of immunoregulation after a protracted antigen challenge [8]. The protein of amyloid fibrils may be composed of light chains synthesized by increased plasma cells and secreted extracellularly, cleaved by lysosomal enzymes in tissue macrophages, polymerized into immunoglobulin light-chain amyloid fibrils, and then deposited [11]. Amyloid is unquestionably related in some basic manner to abnormalities in immune mechanisms, and many conditions known to predispose individuals to amyloidosis are accompanied by plasmacytosis and hypergammaglobulinemia [12].

Although the aminoacid sequence underlies the potential for a protein to form amyloid, and a sustained supply of the respective fibril precursor is an essential prerequisite for amyloid development, little is known about the genetic or environmental factors that determine individual susceptibility to amyloidosis, or those that govern its anatomical distribution and clinical effects [4].

Classifications

In the World Health Organization (WHO) classification, amyloidosis types are defined by the variable fibrillar protein constituents [13]. In primary amyloidosis, amyloid light-chain (AL) protein, which is equivalent to the variable portion of an immunoglobulin light chain, is deposited [8]. This type of amyloidosis occurs in the absence of preexisting disease, with the exception of dyscrasias (e.g., multiple myeloma and Waldenström macroglobulinemia) [6], and eventually progresses to multiple myeloma in at least 30 % of patients [8]. More males than females are affected, and the mean age at presentation is 55–60 years [8], with median survival of 1.5 years [14]. Virtually any organ system except the brain can be involved, and almost any combination of systems involved is possible [4], but many patients present with renal insufficiency. Renal failure and congestive heart failure are common causes of death in affected individuals [8].

Secondary amyloidosis involves the amyloid A (AA) protein, which represents a normal serum alpha globulin that is secreted by the liver as a result of chronic inflammatory states and "mishandled" by the macrophage [8]. Secondary amyloidosis, the most common form of the disease [6], has been reported as a result of Crohn disease, adult or juvenile rheumatoid arthritis, Reiter syndrome, ankylosing spondylitis, familial Mediterranean fever, Sjögren syndrome, dermatomyositis, vasculitis, chronic osteomyelitis, tuberculosis, syphilis, pyelonephritis, bronchiectasis, cystic fibrosis, systemic lupus erythematosus, and parasitic infection [1]. Neoplasms such as those in Hodgkin disease and renal cell carcinoma are also associated with secondary amyloidosis [8]. It has a median survival period of 4.5 years [15] and its incidence has increased gradually, presumably due to the longer life expectancy of patients with the underlying chronic diseases [1]. A third, hereditary or senile systemic type of amyloidosis (ATTR) is derived from mutant and wild-type transthyretin [16]. Familial amyloidosis typically presents as peripheral sensorimotor neuropathy in the second or third decade of life [6]. The kidneys and heart can also be involved [6].

The WHO classification also includes diseases related to other fibrillar protein types, such as Alzheimer disease,

type II diabetes, and transmissible spongiform encephalopathies [4]. Amyloidosis can also be classified as systemic (80–90 % of patients) or localized (10–20 % of patients) [3, 8]. Systemic amyloidosis is frequently distinguished from the localized form by the demonstration of protein in subcutaneous fat, rectal mucosa, bone marrow, urine, and/or serum. Localized amyloidosis, in contrast, is characterized by discrete focal amyloid deposits [3] and is restricted to a single organ [17]. Less frequently, amyloidosis is classified as typical, denoting the involvement of parenchymal organs, or atypical, indicating that manifestations are within organs other than the kidney, liver, spleen, or adrenals [12].

Diagnosis

The diagnostic gold standard for amyloidosis is histological confirmation by Congo red staining, which produces red-green birefringence under crossed polarized light [18]. Positive histological results for amyloid must be followed up with immunohistochemical analysis to determine the fibril type. Suitable antibodies are widely available, but AL deposits often do not stain, presumably because these fibrils are derived from variable-domain monoclonal light-chain fragments that are unique in each case [18]. However, immunohistochemical exclusion of AA and ATTR amyloidosis renders the diagnosis of the AL type overwhelmingly likely [4].

Anatomical and functional evaluation of the tissues/organs affected by amyloid deposits must be tailored to each case. In amyloidosis of the respiratory tract, examinations are likely to include plain radiography, high-resolution computed tomography (HRCT), endoscopy, and comprehensive respiratory function tests. Evidence of systemic disease should be sought clinically, including by urinalysis and hematological and biochemical profiling. Immunoglobulin gene rearrangement studies may identify subtle clones in bone marrow or, in localized AL amyloidosis, within the amyloidotic tissue [4].

SAP is a pentameric nonfibrillar protein component of all amyloid deposits [19]. SAP scintigraphy [20] is used to identify and monitor visceral amyloid deposition in patients with systemic amyloidosis with an acceptable radiation dose. This technique is most sensitive for solid viscera, such as the liver, kidney, and spleen; although it is poor for lung imaging, it remains useful to determine whether amyloid is present in other organ systems [18]. Functional imaging with fluorodeoxyglucose-positron emission tomography (FDG PET-CT) and amyloid scintigraphy with technetium-aprotinin have been shown to highlight areas of amyloid deposition, although the exact role of these techniques remains uncertain [6, 21]. In some cases, dual-phase FDG PET-CT imaging can be used to differentiate between malignancy and amyloid deposits. In early phases, FDG metabolic activity can be seen but delayed images show reduced activity that would not be seen with malignancy [18].

Respiratory Complications

Amyloid is localized to the respiratory tract in 50 % of patients with amyloidosis, and is more common in the systemic form of the disease [6]. Most cases of respiratory amyloidosis are of the AL type, although the presence of chronic inflammatory disease, family history, or extreme old age should signal other possibilities [4]. Three major patterns of involvement have been described: tracheobronchial, nodular parenchymal, and diffuse alveolar septal. Lymphatic, laryngeal, and pleural impairment have also been described [6, 16].

Tracheobronchial Pattern

A tracheobronchial pattern of involvement is the most common form of respiratory amyloidosis. Although histological involvement of the airways is probably common in systemic AL amyloidosis, symptomatic tracheobronchial amyloidosis is usually localized [4]. Tracheobronchial amyloidosis typically occurs in the fifth to sixth decade of life. Patients are often asymptomatic, but can demonstrate hemoptysis, stridor, cough, hoarseness, dyspnea or wheezing, and recurrent pneumonia [8, 11]. Histologically, amyloid is found in the subepithelial interstitial tissue and often surrounds tracheobronchial ducts and acini, some of which may show secondary atrophy. Calcification and foreign-body giant cell reaction may be present [17].

HRCT demonstrates localized tumor-like nodules, multiple discrete plaques, or circumferential thickening with or without luminal narrowing of the trachea, main bronchus, and lobar or segmental bronchi [17] (Fig. 1). Mural nodulation and calcification of the thickened tracheobronchial wall are commonly seen. Luminal narrowing at various levels of the tracheobronchial tree can cause associated abnormalities, such as atelectasis, recurrent infection, bronchiectasis, and post-obstructive air trapping [17].

A significant advantage of HRCT in the evaluation of tracheobronchial amyloidosis is the ability to accurately assess the degree of luminal narrowing. Volumetric acquisition and multiplanar reconstruction enable the accurate evaluation of airway luminal diameters and areas and extent of wall thickening. HRCT can also be used to guide bronchoscopic sampling and identify sites for treatment [6].

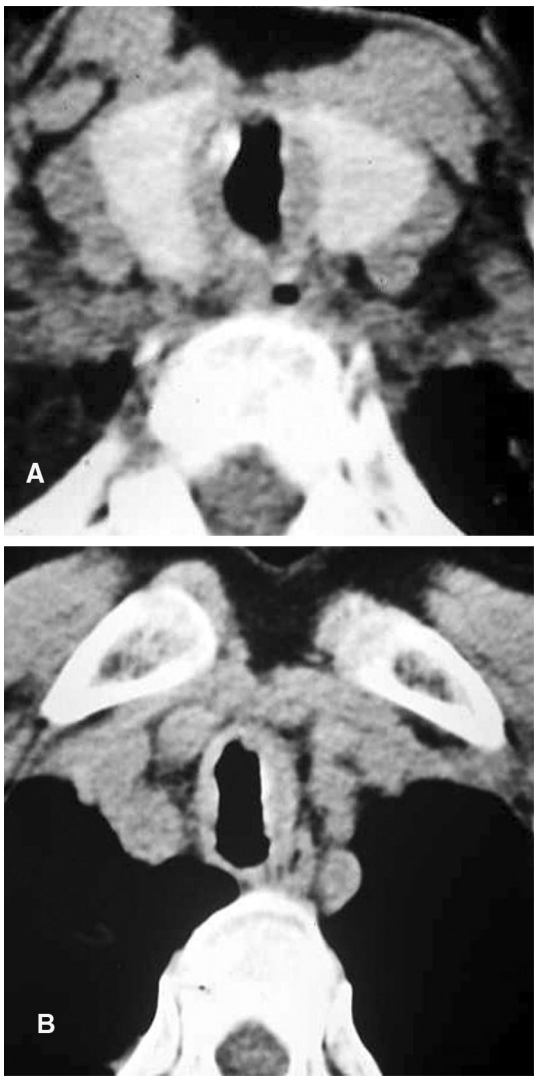

Fig. 1 A 57-year-old woman reporting dry cough, shortness of breath, dyspnea, and hoarseness. Axial CT images with mediastinal window settings (**a**–**b**) show irregular tracheal lumen, determined by multiple parietal nodular lesions. Discrete calcifications are also present in some nodules

The differential diagnosis is limited and includes diffuse tracheal diseases, such as tracheobronchopathia osteochondroplastica (TBO), relapsing polychondritis [22], Wegener granulomatosis, sarcoidosis, inflammatory bowel tracheobronchitis, tracheobronchial paracoccidioidomycosis [23], and tuberculosis [24] (Table 1). In contrast to other diffuse tracheal diseases, relapsing polychondritis and TBO characteristically spare the posterior tracheal and bronchial membranes, whereas amyloidosis involves them [25]. TBO is an uncommon condition characterized by the formation of osseous and/or cartilaginous in the large airways, with no amyloid deposition [11, 26]. Like amyloidosis, granulomatous disease and inflammatory bowel tracheobronchitis can present as circular mucosal thickening and irregularity; differential diagnosis can be achieved by the analysis of associated findings, but tissue biopsy is essential for a definitive diagnosis. Wegener granulomatosis can lead to ulceration in the tracheobronchial tree, which is uncommon in amyloidosis [25].

Nodular Parenchymal Pattern

Nodular parenchymal amyloidosis is a rare form and may be manifested by solitary or multiple nodules [17]. This form is usually of the localized AL type [4] and patients are often asymptomatic, rarely presenting with cough, shortness of breath, or hemoptysis [8, 17]. Histologically, amyloid is often present only in the alveolar interstitium at nodule peripheries. Normal parenchymal structure is usually absent in the central regions, as the compact amyloid mass contains numerous multinucleated giant cells, lymphocytes, and plasma cells [17].

The nodules can demonstrate five characteristic features on HRCT: (1) peripheral or subpleural location, more frequently in the lower lobes, which may be bilateral [4]; (2) well-defined, sharply, or lobulated marginated appearance; (3) calcification, often centrally or in an irregular pattern within the nodule (seen in about 50 % of cases); (4) multiple shapes and sizes ranging from 0.5 to 15 cm; and (5) slow growth, often over years, with no regression [16, 27]. Cavitation may also occur [17] (Fig. 2).

Multinodular parenchymal amyloidosis can be seen in patients with Sjögren syndrome and/or lymphoproliferative diseases [28], such as mucosa-associated lymphoid tissue lymphoma and lymphoid interstitial pneumonia (LIP) [17]. In these conditions, amyloidosis can be specific to the lung and associated with bullae or thin-walled cysts [17] (Fig. 3). The association between pulmonary amyloidosis and cyst formation is not clear; three mechanisms may be involved: airway narrowing with inflammatory cell infiltration, leading to the check valve mechanism; increased fragility and disruption of the alveolar walls as a result of amyloid deposits; and ischemia leading to destruction of alveolar walls as a result of amyloid deposition around capillaries and within the interstitial tissue, ultimately obliterating alveolar capillaries [28].

Cystic lung disease occurs rarely in the absence of Sjögren syndrome or lymphoproliferative disease [6, 29]. However, several studies have shown that the association of amyloid deposits with pulmonary cysts is related to the presence of LIP, which manifests on HRCT as ground-glass opacity, interlobular septal thickening, centrolobular nodules, airspace consolidation, and lung cysts. LIP has been associated with various conditions characterized by abnormal immune function, such as Sjögren syndrome, acquired immunodeficiency syndrome, and systemic lupus erythematosus [30].

Table 1 Respiratory tract amyloidosis patterns—main differential diagnosis

Tracheobronchial	Nodular parenchymal	Diffuse parenchymal
Tracheobronchopathia osteochondroplastica	a. Nodules with calcification	Pneumonia
Relapsing polychondritis	Granulomatous diseases	Interstitial fibrosis
Wegener granulomatosis	(tuberculosis, histoplasmosis; coccidioidomycosis; blastomycosis)	Rheumatoid lung
Sarcoidosis	Hyalinizing granulomas	Sclerodermia
Inflammatory bowel tracheobronchitis	Chondromas	Lymphangitic carcinomatosis
Tracheobronchial paracoccidioidomycosis	Calcified metastasis	Metastatic calcification
Tracheobronchial tuberculosis	b. Nodules with cysts	Alveolar microlithiasis
	Langerhans cell hystiocitosis	
	Lymphocytic insterstitial pneumonia	
	Lymphangioleiomyomatosis and micronodular pneumocyte hyperplasia	

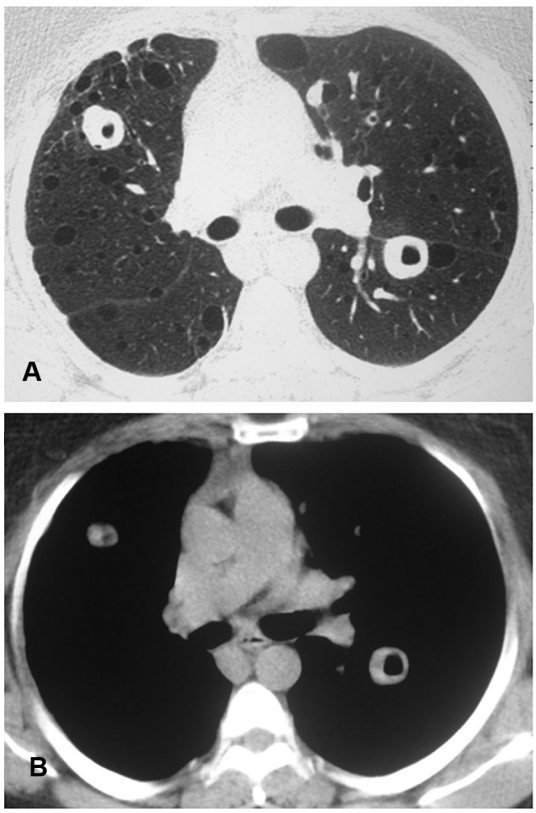

Fig. 2 A 52-year-old woman with Sjögren syndrome. Axial CT images with lung (**a**) and mediastinal (**b**) window settings showed sparse pulmonary cysts, compatible with lymphocytic interstitial pneumonia. Nodules, most cavitated, are also present. Note in (**a**) the presence of a nodule abutting the cyst wall on the lingula. Lung biopsy confirmed the diagnosis of amyloidosis

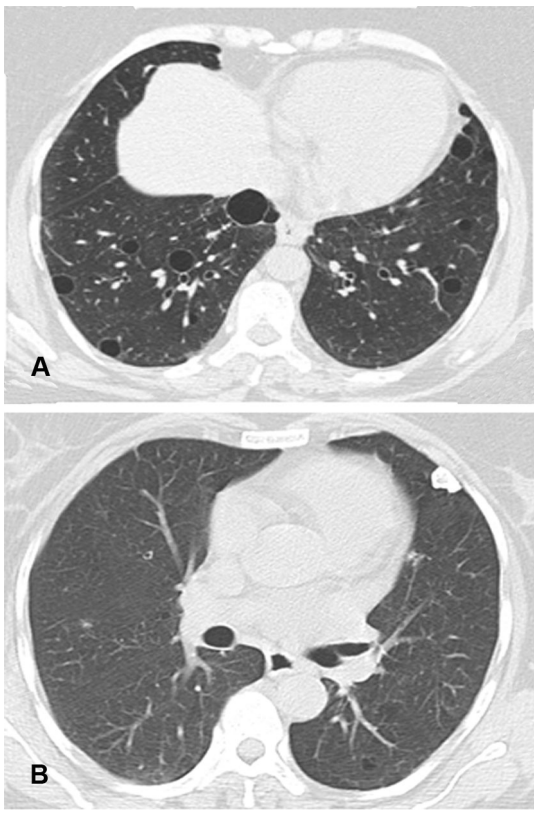

Fig. 3 A 53-year-old woman with lymphocytic interstitial pneumonia. Axial CT images with lung window settings at different levels (**a**–**b**) show multiple cyst lesions and some parenchymal nodules. Note in (**b**) a peripheral calcified nodule in the left lung. Biopsy confirmed the diagnosis of associated amyloidosis

The identification of multiple nodules and cysts on HRCT in patients with Sjögren syndrome clearly suggests a diagnosis of pulmonary amyloidosis and lymphoproliferative disease [30], especially when nodules abut cyst walls [31] (Fig. 4). In these cases, histopathologic specimens show amyloid deposition and lymphoid cell infiltration in the cyst walls and narrowed corresponding bronchioli [30, 31]. The most important factor to consider is that malignant lymphoma may develop with the CT

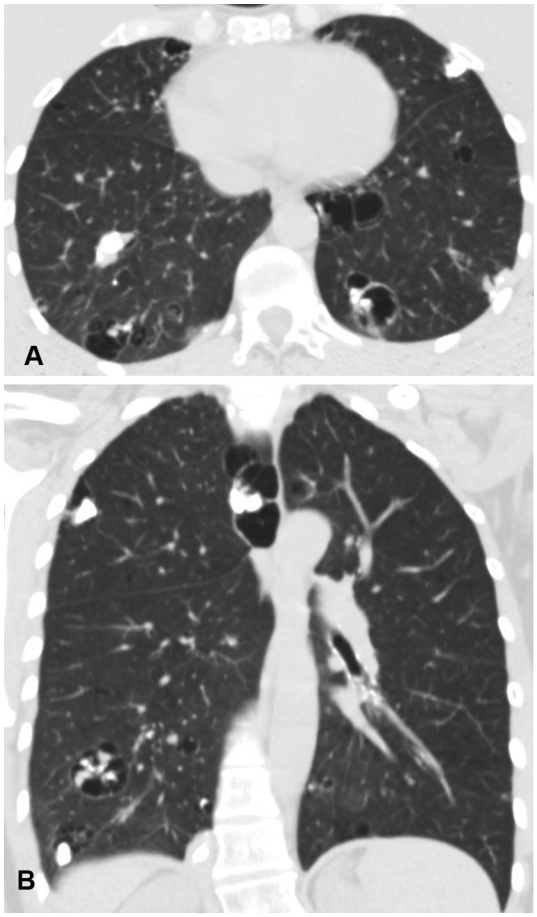

Fig. 4 A 56-year-old woman with biopsy-confirmed amyloidosis. Coronal (**a**) and axial (**b**) CT images with lung window settings show multiple cysts and nodules. Note that some nodules are calcified and abut cyst walls, which is highly suggestive of an association with lymphoproliferative disease, particularly lymphocytic interstitial pneumonia and amyloidosis

appearance of these features, requiring differentiation by lung biopsy [28–31].

The differential diagnosis of multiple nodules and masses is broad and includes primary or metastatic neoplasm and granulomatous disease [6, 8, 17]. Multifocal calcified nodules or masses are seen most commonly as the sequelae of disseminated infections, such as tuberculosis and histoplasmosis, and are associated less commonly with other infectious causes, including coccidioidomycosis and blastomycosis, or other diseases such as hyalinizing granulomas and chondromas [32]. Multifocal calcifications are unusual in metastatic diseases, except for osteogenic sarcoma and chondrosarcoma [33]. Pulmonary calcification in metastatic mucin-producing adenocarcinomas (including those of the breast, colon, thyroid gland, and ovary) is rare, even when the primary tumor contains calcifications. Successfully treated metastases can also calcify [33].

The differential diagnosis of multiple cysts and nodules includes mainly Langerhans cell histiocytosis (LCH), LIP, and lymphangioleiomyomatosis associated with micronodular pneumocyte hyperplasia in women with tuberous sclerosis [31]. In LCH, the nodules usually have diameters <1 cm and centrolobular location, and cyst lesions always save the regions of costophrenic angles; these features differ from the varying sizes and cyst wall abutment of nodules associated with randomly distributed cysts observed in amyloidosis [30, 31]. Nodules in LIP are also located in centrilobular regions, and are usually <5 mm in diameter with poorly defined margins. Nodules of lymphangioleiomyomatosis associated with micronodular pneumocyte hyperplasia in women with tuberous sclerosis are generally <1 cm in diameter, and their distribution shows upper or peripheral predominance [31].

Diffuse Alveolar Septal Pattern

A diffuse parenchymal pattern of involvement is probably the least common form of respiratory amyloidosis [8]. Most affected patients have systemic amyloidosis [6] and present clinically with recurrent pneumonia, likely due to interstitial involvement of diffuse alveolar septal amyloid [1, 16]. Histologically, amyloid is present in the media of small blood vessels and in the parenchymal interstitium. In the interstitium, amyloid is present in relation to endothelial and epithelial basement membranes, and can appear in a uniform linear pattern or as multiple small nodules [6].

Vascular involvement has been described in 88–90 % of patients with AL amyloidosis [9]. As mentioned previously, deposition of amyloid in the pulmonary vasculature is a common histological finding, but clinical expression secondary to amyloid vasculopathy is infrequent [9]. Amyloid deposition in blood vessel walls can result in endothelial dysfunction and eventually lead to pulmonary arterial hypertension [9].

The HRCT findings of diffuse parenchymal amyloidosis consist of abnormal reticular opacities, interlobular septal thickening, ground-glass opacities [21], multiple small well-defined nodules measuring 2–4 mm in diameter, and confluent consolidation in the subpleural region [17, 32] (Fig. 5). Calcifications can be seen in some nodules and areas of consolidation [17, 32]. Interlobular septal thickening may be nodular, and this aspect can also be seen along peribronchovascular structures and fissures [34, 35].

The main differential diagnosis includes pneumonia, many interstitial lung diseases (e.g., interstitial fibrosis, rheumatoid lung, and scleroderma), and neoplastic diseases such as lymphangitic carcinomatosis [6, 8]. Consolidations with calcifications can also be seen in metastatic calcification and alveolar microlithiasis [36]. The former is found in hypercalcemic states and in patients who have

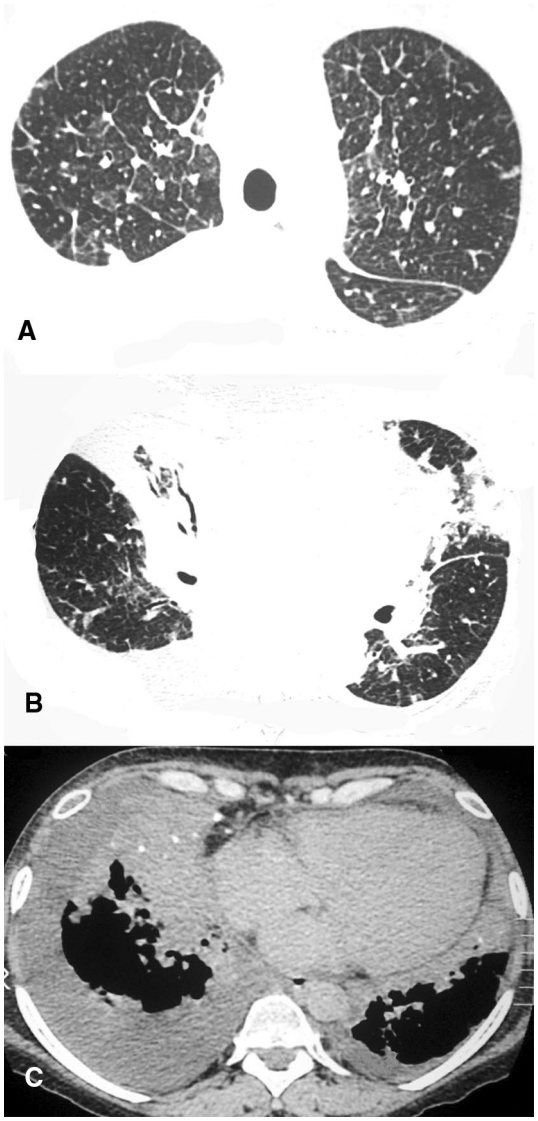

Fig. 5 A 50-year-old woman who reported dyspnea for 4 months. Axial CT image with the lung window setting at the level of the upper lung (**a**) shows diffuse, smooth interlobular septal thickening with nodulation. Axial CT images with lung (**b**) and mediastinal (**c**) window settings at the level of the lower lung show parenchymal consolidations with air bronchogram and small punctate foci of calcification. Bilateral pleural effusion, more extensive on the right side, is also present

undergone kidney transplantation or are being treated with intravenous calcium [33]. On HRCT, it usually presents as centrilobular, nodular ground-glass opacities with numerous fluffy and poorly defined nodules, but in contrast to amyloidosis, no interlobular septal thickening is present [37]. Alveolar microlithiasis, a rare disease of unknown origin, is characterized by intra-alveolar accumulation of spherical calcified concretions [33, 38]. HRCT findings include diffuse ground-glass attenuation, subpleural linear calcifications, small parenchymal nodules, nodular fissures, subpleural nodules, crazy-paving pattern, dense consolidations, and subpleural cysts [39]. The apparent calcification of septa on CT is due to the accumulation of calcospherites in alveoli adjacent to the septa [32].

Other Thoracic Findings

Lymph node involvement is seen pathologically in about 20 % of amyloidosis cases [8]. Hilar or mediastinal adenopathy is rarely associated with localized pulmonary amyloidosis [4, 18], and its discovery should prompt the search for a systemic etiology [5]. Although it is usually asymptomatic, occasional cases of tracheal compression and superior vena cava obstruction have been reported [4, 18]. HRCT findings of amyloid lymphadenopathy are variable; calcification is not uncommon and can be speckled [8, 18] (Fig. 6). Low-density areas within lymph nodes have also been described [18]. The differential

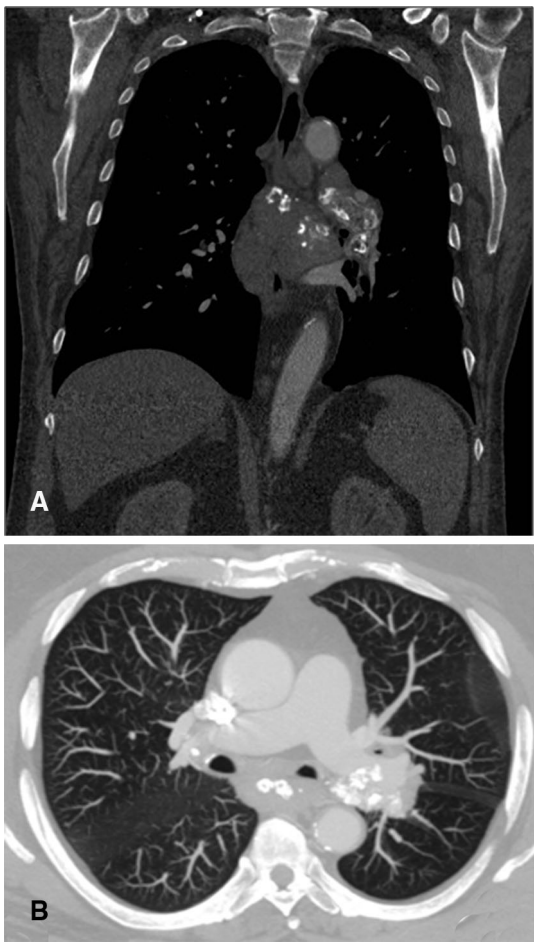

Fig. 6 A 53-year-old woman with systemic amyloidosis. Coronal contrast CT image with the mediastinal window setting (**a**) shows multiple calcified mediastinal lymph nodes. Axial reformatted mini–maximum intensity projection image at the infracarinal level (**b**) shows multiple calcified infracarinal and bilateral hilar lymph nodes

diagnosis includes sarcoidosis, silicosis, coal worker's pneumoconiosis, treated lymphoma, and granulomatous diseases [33]. Pleural involvement has also been reported, manifesting as pleural thickening and persistent pleural effusions [6, 40].

The larynx is the most common site of localized amyloidosis in the head and neck region [11]. Amyloid appears in 0.5–1 % of benign laryngeal disease; it usually appears during the fifth or sixth decade of life [41], with the incidence increasing with age [4], and the male to female ratio is 3:1 [11]. Symptomatic laryngeal amyloidosis is usually localized, but is rarely a manifestation of the systemic AL type [42]. In order of frequency, laryngeal lesions are localized in the ventricle, vestibular folds, vocal folds, epiglottis, aryepiglottic folds, and subglottis [11]. Amyloid deposits occur most commonly in the supraglottic larynx and usually present with hoarseness or stridor, but can cause a sensation of "fullness" in the throat, choking, and dyspnea on exertion [42]. The appearance of the laryngeal amyloidosis on HRCT is nonspecific. The lesion presents as a relatively well-defined, submucosal, homogenous soft-tissue mass with high density or calcification. Contrast enhancement is absent or minimal [11].

The differential diagnosis includes hyalinized myomatous polyps, malignancy, and granulomatous conditions such as tuberculosis and scleroma of the larynx; the main differentiation must be made with chondrosarcoma, which is the most frequent cause of a calcified mass in the larynx [43]. The signal characteristics of amyloid on magnetic resonance imaging (MRI) resemble those of skeletal muscles. This feature may be an important differentiating point because muscle is an easy reference frame, and chondrosarcoma does not appear in this manner on MRI [43].

Treatment and Prognosis

Average survival in patients with systemic amyloidosis and pulmonary involvement is 16 months [44]. Although amyloidosis is a benign lesion, it can be fatal as a result of airway obstruction or respiratory failure [11]. Serial SAP scintigraphy in more than 1000 patients with various types of systemic amyloidosis has confirmed that amyloid deposits do regress, albeit relatively slowly, in the majority of cases when the supply of the respective fibril precursor protein is reduced [11]. Regression has been demonstrated systematically in patients with AA amyloidosis following intensive anti-inflammatory therapy, in patients with AL amyloidosis following chemotherapy, and in patients with familial amyloid polyneuropathy and dialysis amyloidosis following liver and renal transplantation [11]. The key to effective monitoring and treatment of amyloidosis is relatively frequent (approximately 6 monthly) repetition of these investigations, bearing in mind that organ function may not closely reflect amyloid load [4]. No curative treatment has been identified to date, especially for systemic amyloidosis, and conservative surgical excision with functional preservation remains the treatment of choice [11].

Conclusion

HRCT findings of amyloidosis are nonspecific and vary in each involvement pattern. Knowledge about the disease impairment type is very important, and amyloidosis should be in the differential diagnosis of other very common diseases, such as infectious diseases, neoplasms, and vasculitis. Tissue biopsy is always required for definitive diagnosis.

Compliance with Ethical Standards

Conflict of Interest None.

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