Case Report

Bronchiectasis in rare pulmonary diseases: A case series

Ines Maria Grazia Piroddi, Edmundo Rosales[1], Catia Cilloniz[1], Adrian Ceccato[1], Antonello Nicolini

Respiratory Diseases Unit, Hospital of Sestri Levante, Sestri Levante, Italy, [1]Department of Pneumology, Hospital Clínic, CIBERES, IDIBAPS, Barcelona, Spain

ABSTRACT

Bronchiectasis, defined as permanent dilatation of the airways, usually causes chronic cough and sputum production with intermittent bacterial exacerbations. Bronchiectasis can have a severe impact on quality of life for many patients due to these symptoms. Establishing the cause of bronchiectasis may be difficult. Even with exhaustive clinical, laboratory, and pathologic testing, up to 50–80% of cases of bronchiectasis may still be idiopathic. Congenital bronchiectasis is much rarer than previously considered. A variety of respiratory and systemic diseases such as autoimmune or rheumatologic diseases may be complicated by pathological bronchial dilatation, and therefore various medical specialists will be dealing with the condition in one-way or another. Some bowel diseases are associated with a variety of systemic manifestations including large and small airway involvement: One of the most commonly associated airway diseases is bronchiectasis. On this regard, we report five patients with bronchiectasis and rare diseases admitted to our hospitals since 2012. Patients were recruited over a period of 36 months after opening a bronchiectasis outpatient clinic.

Key words: Autoimmune diseases, bronchiectasis, bronchiectasis outpatient clinic, rare diseases

INTRODUCTION

Bronchiectasis, defined as permanent dilatation of the airways, usually causes chronic cough and sputum production with intermittent bacterial exacerbations. Bronchiectasis can have a severe impact on quality of life for many patients due to these symptoms.[1,2] Table 1 summarizes the conditions associated with bronchiectasis.

Establishing the cause of bronchiectasis may be difficult. Even with exhaustive clinical, laboratory, and pathologic testing, up to 50–80% of cases of bronchiectasis may still be idiopathic.[3] Childhood infections, including pertussis, were thought to have caused 11% of the bronchiectasis cases and 10% of cases were related to the prior granulomatous disease.[4]

Several respiratory infections can cause not only bronchiectasis including measles, pertussis, and tuberculosis but also viruses (HIV, paramyxovirus, adenovirus, and influenza), Gram-negative bacteria (*Pseudomonas aeruginosa* and *Haemophilus influenzae*), and other atypical mycobacteria.[5]

Address for correspondence:
Dr. Antonello Nicolini, Respiratory Diseases Unit, Hospital of Sestri Levante, Sestri Levante, Italy.
E-mail: antonellonicolini@gmail.com

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Table 1: Conditions associated with bronchiectasis

Mucociliary clearence defects
Kartagener's syndrome,primary ciliary dyskinesia,Young's syndrome
Post-infectious complications
Bacteria, Mycobacterial infections (Tuberculosis and Mycobacterium Avium Complex),whooping cough,viral (measles, adenovirus, Influenza virus)
Mechanical bronchial obstruction
Foreign body,stenosis,tumour,lymphonode,
Immune disorders
Hypogammaglobulinemia,immunoglobulin G subclass deficiency, HIV,allergic bronchopulmonary aspergillosis, post-lung transplant
Sequelae of aspiration or toxic inhalation
Rheumatic or chronic inflammation condition
Rheumatoid arthritis, Sjogren's syndrome,inflammatory bowel disease
Chronic obstructive pulmonary Disease (COPD)
Others diseases
Cartilage deficiency ,alfa-1 antitrypsin deficiency,Yellow nail syndrome

Non-cystic fibrosis (CF) bronchiectasis secondary to *Mycobacterium avium* complex infection is a typical feature of the Lady Windermere syndrome.[5]

Bronchiectasis is found typically in the middle lobe and lingula, older patients, and immunocompetent women with no smoking history or pulmonary disease.[5]

Congenital causes of bronchiectasis include CF, primary ciliary dyskinesia (PCD), and Xl-alfa 1 antitrypsin (AAT) deficiency.[3] Congenital bronchiectasis is much rarer than previously considered. Specific causes include Williams–Campbell syndrome (bronchial cartilage deficiency), tracheobronchomegaly (Mounier-Kuhn syndrome), Marfan's syndrome, late presenting H-type tracheobronchial, and esophagobronchial fistula.

Bronchiectasis has been reported in congenital lung malformations such as sequestration or rarely with a rib malformation.[6] In children, aspiration of foreign bodies into the lower respiratory tract is the most common and most important obstructing lesion causing bronchiectasis with the incidence peaking in the 2nd year of life.

Adults may also develop bronchiectasis secondary to aspiration of foreign material and due to endobronchial tumors although this is a rare cause of bronchiectasis.[6]

A variety of respiratory and systemic diseases may be complicated by pathological bronchial dilatation, and therefore various medical specialists will be dealing with the condition in one way or another.[7-9]

Bronchiectasis has been demonstrated on high-resolution computed tomography (HRCT) in about 30% of cases of rheumatoid arthritis although it may be clinically silent and it may precede or follow the development of rheumatoid arthritis. Various hypotheses exist regarding the association between bronchiectasis and rheumatoid arthritis including chronic suppurative infections leading to bronchiectasis, treatment with disease-modifying antirheumatic drugs, or alternatively that chronic infection in a bronchiectasis patient provide additional antigenic stimuli that then triggers rheumatoid arthritis. It is also hypothesized that rheumatoid arthritis and bronchiectasis share a genetic predisposition.[10]

Bronchiectasis is seen in 7–25% of the patients with asthma or chronic obstructive pulmonary disease (COPD) and interstitial lung disease. In these conditions, bronchiectasis is usually found in the context of severe disease and is then not considered the primary disease.[3,9] Coexisting bronchiectasis and COPD may represent a more severe phenotype as also indicated by colonization or infection with potentially pathogenic organisms such as *P. Aeruginosa* and atypical mycobacteria.[11]

Allergic bronchopulmonary aspergillosis has as one of its key features bronchiectasis, which tends to be central in its distribution.[8]

Osteoporosis and osteopenia have a high prevalence in patients with lung diseases. The prevalence of osteopenia and osteoporosis is high in patients with non-CF bronchiectasis.[12] Crohn's disease is an inflammatory bowel disease associated with a variety of systemic manifestations including large and small airway involvement. The most commonly reported airway disease is bronchiectasis.[13]

We report five patients with no CF-bronchiectasis and rare diseases admitted to our hospitals since 2012. Patients were recruited over a period of 36 months after opening a bronchiectasis dedicated outpatient clinic. Out of one patient died all of them are still in follow-up in our clinics.

CASE REPORTS

Case 1 (Kartagener's syndrome)

A 67-year-old female presented to the outpatient clinic in May 2010 with chief complaints of recurrent episodes of sneezing, cough with expectoration, shortness of breathing, and episodes of exacerbations of bronchitis. Chest X-ray showed dextrocardia and diffuse interstitial opacities [Figure 1a].

On auscultation, bilateral wheeze and bilateral crackles were audible. Chest CT revealed dextrocardia and diffuse bronchiectasis [Figure 1b]. CT paranasal sinuses showed nasal polyposis, chronic sinusitis, and hypoplastic frontal sinuses. Pulmonary functional evaluation showed severe restriction (total lung capacity [TLC] - 59%, residual volume [RV] - 66%, forced vital capacity [FVC] - 44%, and forced expiratory volume 1 s [FEV_1] - 39%). Two years later, she performed a nasal brushing with determination of 100%

cilia lacking of inner dynein arm associated with axonemal disorganization in 60% of the cilia. In the follow-up visits, the patient showed a progressive impairment of respiratory conditions. Two years later, arterial blood gasses (ABGs) analysis showed hypoxia (PaO_2 52 mmHg). The patient was treated with oxygen therapy for 2 years. She died in 2014 after intensive care admission because of an episode of severe respiratory failure.

Case 2 (alpha 1 antitrypsin deficiency)

A 78-year-old male, former smoker referred to other hospital for severe COPD, emphysema, and alpaa-1 antitrypsin (AAT) deficiency (PiMZ genotype), was admitted to our hospital with diagnosis of severe hypercapnic respiratory failure (pH 7.25, PaO_2 60 mmHg on FiO_2 30%, $PaCO_2$ 80 mmHg PaO_2/FiO_2 ratio 200). He had been treated with substitution treatment with AAT once a week since 2012. The AAT concentration at admission was of 78.6 mg/dL (reference value: 90–200 mg/dL). He was treated with noninvasive ventilation for 5 days until normalization of ABG. Pulmonary function testing showed a very severe airflow obstruction (FVC 38% FEV_1 18% FEV_1/FVC 0.46) and hyperinflation (TLC 227%, RV 288%, motley 185%) and diffusing lung carbon monoxide (DLCO) 29%. A high-resolution CT of the thorax showed diffuse panacinar emphysema with thickening of bronchial walls and bronchiectasis in both lower pulmonary lobes [Figure 2]. Abdominal ultrasound showed no signs of chronic liver damage.

Sputum analysis yielded *P. Aeruginosa* and methicillin-sensitive *Staphylococcus aureus* (MSSA). Laboratory tests showed neutrophil-dominated inflammation (leukocytes 12.8 × 10^3/μL, neutrophils 86.2%, and lymphocytes 10.4%) and C-reactive protein 14.5 mg/dL (n.v. 0.50 mg/dL). The patient was treated with ertapenem 1 g/day and dicloxacillin 1 g/6 h for 10 days, bronchodilators, corticosteroids, and chest physiotherapy with the rapid improvement of clinical conditions. He was discharged 2 weeks after the admission.

Case 3 (Sjogren's syndrome)

A 68-year-old female, no smoker, referred to the outpatient clinic because of pulmonary involvement of Sjogren's syndrome since 2006. The patient has been treated with prednisone 50 mg/day and azathioprine 50 mg/day for 7 years. Two months before, she complained chest pain, breathlessness, cough, and fever, but no sputum. Laboratory evaluations showed neutrophil-dominated inflammation (leukocytes 14.21 × 10^3/μL, neutrophils 89.4%, and lymphocytes 13.2%) and C-reactive protein 8.48 (n.v. 0.50). Chest high-resolution CT demonstrated several ground-glass areas associated with bilateral bronchiectasis [Figure 3]. Pulmonary function is as follows: FVC 66%, FEV_1 69%, TLC 72%, RV 74%, and DLCO 58%. ABG pH 7.45, $paCO_2$ 35 mmHg, and paO_2 66 mm on room air. The patient underwent fiberbronchoscopy (owing to

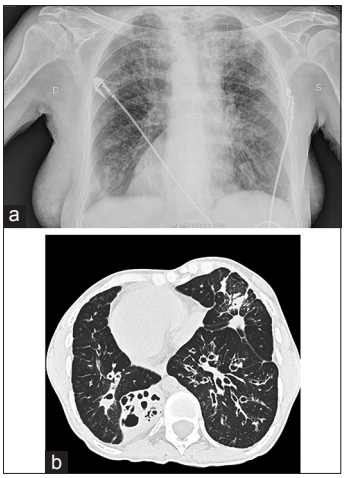

Figure 1: Chest X-ray and computed tomography. Dextrocardia and interstitial lung pattern (a). Chest CT diffuse bilateral bronchiectasis at lower lobes associated with bronchial wall thickening and atelectasis (b)

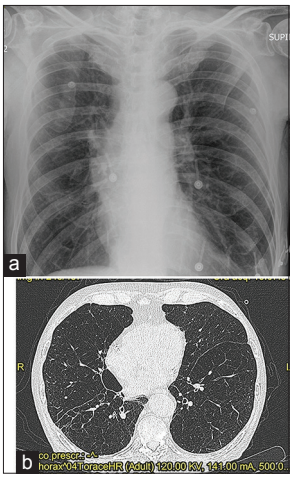

Figure 2: (a and b) Chest X-ray: Hyperinflation of the lungs due to emphysema. Computed tomography: Diffuse panacinar emphysema associated with bulla and bronchiectasis

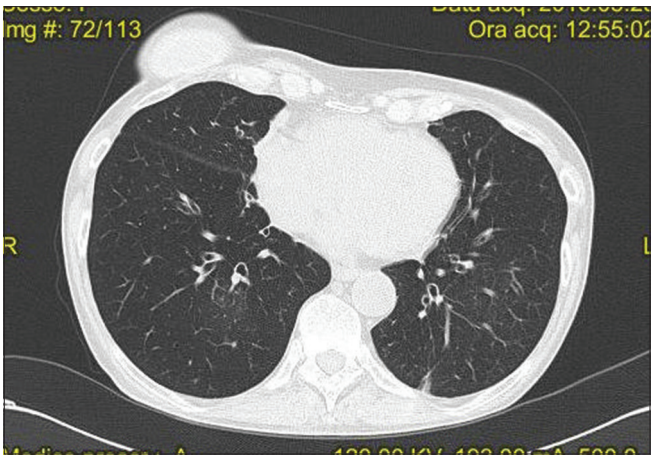

Figure 3: Chest computed tomography bronchiectasis at lower lobes

she does not produce expectoration): MSSA and *Aspergillus* spp. were isolated from bronchoalveolar lavage. She was treated with dicloxacillin 1 g every 6 h for 2 weeks and itraconazol 200 mg twice a day for 4 weeks. After treatment, a normalization of biochemical parameters (leucocytes and C-reactive protein) was observed.

Case 4 (Churg-Strauss syndrome)

A 50-year-old male, no smoker, no toxic habits, no known allergies, with difficult control severe asthma was referred to our hospital for evaluation. He has a clinical history of hepatitis C treated with interferon and ribavirin but was suspended after 4 weeks for side effects with the appearance of Raynaud syndrome with digital ischemia, the latter treated with amlodipine and pentoxifylline.

Asthma was diagnosed 25 years ago after a bronchospasm crisis. Initially, poor control of asthma with many visits to the emergency room, but since 10 years ago, better control of symptoms. He used many combinations of inhaled bronchodilators and corticosteroids, even omalizumab for 6 months with no response and no reduction of corticosteroids.

In the last year, he had many exacerbations with the use of corticosteroids almost permanently and antibiotics sometimes, with little time symptoms free between exacerbations. In the last year, his treatment included inhaled formoterol/beclometasona 6 mcg/100 mcg two inhalations twice daily, inhaled tiotropium bromide 18 mcg once daily, montelukast 10 mg once daily orally, and inhaled salbutamol as needed.

Habitual symptoms were bronchorrhea with coughing, wheezing, and feeling breathless, especially at night. No symptoms of chronic rhinosinusitis or gastroesophageal reflux. Inspiratory wheezing and rhonchi on physical examination.

The skin prick test was positive to dust mite and grass pollen, negative to fungus. Complete blood count showed peripheral blood eosinophilia (1000 eosinophils/μL, 11.4%) with higher level of total IgE (1046 kU/L), but lower level of IgE specific to *Aspergillus*. Levels of IgM, IgA, and IgG were normal. Antineutrophil cytoplasmic antibodies (ANCAs) were negative. FVC was 4.32 L (80%), FEV_1 was 2.24 L (55%), FEV_1/FVC was 0.52, and postbronchodilator test was negative. Sputum culture was negative for bacteria and mycobacteria. Chest CT showed bronchial wall thickening that affects all lung lobes, segmental atelectasis in the middle lobe with mucous plugs, and mucous plugs and bronchial dilatation in both lower lobes [Figure 4]. Skin biopsy performed 2 years ago of purpuric lesions at knees and ankles showed acute inflammation with eosinophilic infiltration and fibrinoid necrosis of vessel walls.

Considering clinical history, laboratory test, and biopsy findings, eosinophilic granulomatosis with polyangiitis (previously called Churg-Strauss syndrome) was suspected. The patient was evaluated by the liver unit and tests showed that hepatitis C was cured and hepatic function was normal. Chronic oral corticosteroids (prednisone 1 mg/kg) were initiated with the improvement of symptoms. Methotrexate was initiated and well tolerated; the dose was increased until 15 mg weekly and used for maintenance. Subsequently, the dose of oral corticosteroids was decreased until 7.5 mg of prednisone daily maintaining a good control of symptoms.

During following check-ups, *S. pneumoniae* and *H. influenzae* were isolated from sputum at different times in the context of exacerbations and were treated with oral antibiotics and increase of oral corticosteroid. For this reason, long-term oral doxycycline was initiated with a concomitant decreased of a number of exacerbations and control of symptoms.

Last tests showed 6% of eosinophilia, FVC 4.05 L (76%), FEV_1 2.19 (53%), FEV_1/FVC 0.54, and a positive postbronchodilator test.

Case 5 (Swyer-James-MacLeod syndrome)

A 55-year-old female with dyspnea on exertion and is admitted to our hospital. Allergic to sulfonamide, no smoker, and no other toxic habit. She has a clinical history of iron deficiency anemia following controls at hematology unit, community-acquired pneumonia 5-year ago treated as outpatient. First-degree atrioventricular blockage diagnosed as a consequence of an episode of dizziness, no specific treatment. She has a past surgical history of amygdalectomy, appendectomy, and excision of benign fibrous tumor in both breasts. Furthermore, she has a history of recurrent respiratory infections during childhood. The usual treatment was only oral iron supplemental.

The initial physical examination found patient was eupneic, auscultation with crackles at the base of right lung, no wheezing, and no Ronchi. The respiratory rate was 20/min with oxygen saturation of 98% at room air and pulse rate of 83/min. No signs of cardiac congestion, no cardiac murmurs, and no signs of deep vein thrombosis.

Blood analysis showed a normal blood count, hemoglobin of 13.8 g/dL, and hematocrit of 40% with normal mean corpuscular volume. Renal and hepatic function was normal. ABG at room air showed a pH of 7.42, a partial pressure of oxygen of 85 mmHg, and a partial pressure of carbon dioxide of 36 mmHg. D-dimer was negative.

FVC was 2.58 L (90%), FEV_1 was 1.81 L (81%), FEV_1/FVC was 0.70, and postbronchodilator test was positive for FVC (+390 mL, +15%), but no FEV_1 (+60 mL, +3%). Plethysmography showed a VC of 2.67 L (89%), RV of 2.29 L (128%), TLC of 4.96 L (102%), and RV/TLC% of 46%. DLCO was 99%.

The ventilation/perfusion lung scintigraphy and pulmonary angiography rule out pulmonary embolism but showed a marked hypoperfusion in the right lung predominantly in the right upper lobe (RUL) and right lower lobe (RLL). Chest CT showed large areas of oligohemia with bronchiectasis that affects RUL and RLL at inspiration CT [Figure 5a] and air trapping at expiration CT [Figure 5b] suggestive of constrictive bronchiolitis. These findings suggest a chronic lung involvement secondary to bronchiolitis.

All these findings are consistent with Swyer-James syndrome (also called Swyer-James-MacLeod's syndrome) probably in relation to the history of recurrent infections during childhood. Inhaled bronchodilators were initiated because of positive bronchodilator test and presence of air trapping at pulmonary function test and chest CT. The patient was discharged to home; she continues follow-up at pneumology unit.

All the alive patients have given the consent to publish material regarding them.

DISCUSSION

Bronchiectasis can result from many diseases, which makes etiological investigation a complex process demanding special resources and experiences although it has been proved that etiological diagnosis is useful for therapeutic approach.[14] In a recent study by Amorim *et al*. who reviewed 202 cases of bronchiectasis, the most commonly identified cause was postinfectious (30.3%), mostly tuberculosis (27.2%), but 57.4% no definitive etiological diagnosis was established.[14] Among them, they found one case of PCD, three cases of autoimmune disease, and one case of intestinal inflammatory diseases between rare diseases. In a review including 1577 patients, Gao *et al*.[15] concluded that intensive investigations of the etiologies of bronchiectasis might change patient's management and therefore should be incorporated into routine clinical practice. Moreover, while the pathophysiology of bronchiectasis is well defined, the etiologies are varied.[16] Throughout the literature, patients without an identified etiology are reported as idiopathic disease. A focused medical history and focused laboratory investigation should reveal the etiology of non-CF-bronchiectasis in many cases.[16]

We have reported five cases of bronchiectasis-associated with rare diseases with low prevalence.

In the first case, we have presented a case of Kartagener's syndrome. In this syndrome, the main defect is a ciliary dyskinesia. PCD is a rare autosomal recessive disease, caused by specific primary structural and/or functional abnormalities of the motile cilia, which contribute to retaining of secretions and microbial pathogens. Approximately, half of the patients with PCD have Kartagener's syndrome (bronchiectasis, sinusitis, and situs inversus).[8,9] Young's syndrome is characterized by obstructive azoospermia and the defect in mucociliary clearance appears to arise from tenacious, poorly cleared mucous.[8,17] None specific treatments are available, the physician mainly should focus in the airway clearance and the vaccine immunization. In the second, a case of AAT deficit is presented. With regard to AAT deficiency, emphysema is the most commonly associated pulmonary abnormality.[3] However, Parr *et al*.[7] have demonstrated that 27% of 74 AAT-deficient patients had HRCT scan evidence of bronchiectasis. An association of AAT deficiency in the etiology of bronchiectasis was postulated after a number of case reports linked severe (Pi ZZ phenotype) AAT deficiency to bronchiectasis in individual or small numbers of cases. Many of these reports mention other possible

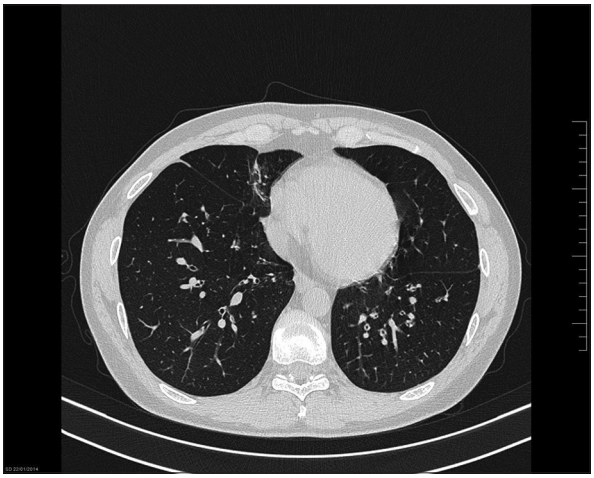

Figure 4: Chest computed tomography bronchial wall thickening that affects all lung lobes, segmental atelectasis in the middle lobe with mucous plugs, and bronchial dilatation in both lower lobes

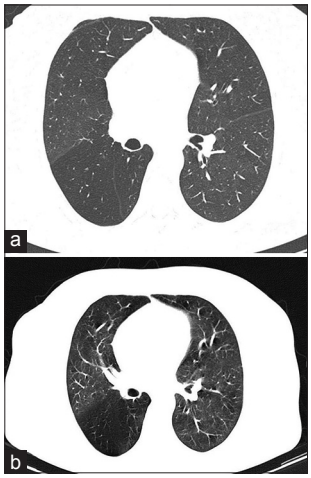

Figure 5: (a and b) Chest computed tomography large areas of oligohemia with bronchiectasis that affects right upper lobe and right low lobe at inspiration and air trapping at expiration computed tomography

causes of bronchiectasis in patients' histories, and exclusion of specific conditions such as immune deficiency and CF is variable.[6] The treatment with AAT has shown a reduction in the progress of emphysematous disease and mortality.[18]

In the third case, we have discussed Sjogren's disease an autoimmune disease characterized by exocrine glands affectation although the lung complications are common in the disease bronchiectasis may be present, the most common form is the interstitial compromise such as nonspecific interstitial pneumonia, usual interstitial pneumonia, or less common form as lymphoid interstitial pneumonia. There is not a specific treatment and immunosuppressive treatments or corticosteroids usually are the most prescribed.[19]

Among the pulmonary compromised by ANCA-vasculitis related the asthma symptoms, pulmonary hemorrhage, or granulomatous infiltrate are a common feature. The bronchiectasis is reported mainly in microscopic polyangiitis, however, may be present in eosinophilic granulomatosis with polyangiitis disease.

With regard the last case, Swyer-James-MacLeod syndrome is considered to be a relatively uncommon and complex disease characterized by unilateral hyperlucency of the lung in the chest X-ray. This disease is characterized by bronchiolitis and bronchiectasis.[20]

CONCLUSIONS

Our case series shows that bronchiectasis is part of broad spectrum of systemic diseases, and etiologic diagnosis was achieved after focused clinical investigation.

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Conflicts of interest
There are no conflicts of interest.

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