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Clinical Case Seminar **CCS1 (1-5)**

An elderly man with congenital hypogammaglobulinemia and persistent cough

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Abstract

We report here the case of a 71 year-old male, former smoker of 60 pack-years, with an history of congenital hypogammaglobulinemia associated to recurrent fever and persistent productive cough, treated with immunoglobulin G (IgG) replacement therapy. On the last 3 years he was followed by our outpatient clinic for chronic respiratory failure (treated with long-term oxygen therapy) secondary to diffuse bilateral bronchiectasis and pulmonary panacinar emphysema. The patient did not follow our influenza and pneumococcal vaccinations recommendations. The patient died after 3 years of follow-up.We report here an unusual case in an adult of diffuse and bilateral bronchiectasis secondary to congenital hypogammaglobulinemia.

Key Words: bronchiectasis, congenital hypogammaglobulinemia.

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Introduction

The bronchiectasis are permanent dilatation of bronchi (1). The cause of bronchiectasis is unknown up to 50% of cases, whereas immune defects, such as congenital hypogammaglobulinemia, are uncommon cause of bronchiectasis in the adult (2,3).

Case presentation

A 71 year-old male, past farmer, former smoker from 22 years of 60 pack-years, with an history of congenital hypogammaglobulinemia associated to recurrent fever and persistent productive cough, treated with immunoglobulin G (IgG) replacement therapy. On the last 3 years he was followed by our outpatient clinic for chronic respiratory failure (treated with long-term oxygen therapy) secondary to diffuse and bilateral bronchiectasis and pulmonary emphysema. His family history was negative for respiratory and immune system diseases. He was under regular oral treatment with lamotrigine (100 mg once/daily) for generalized

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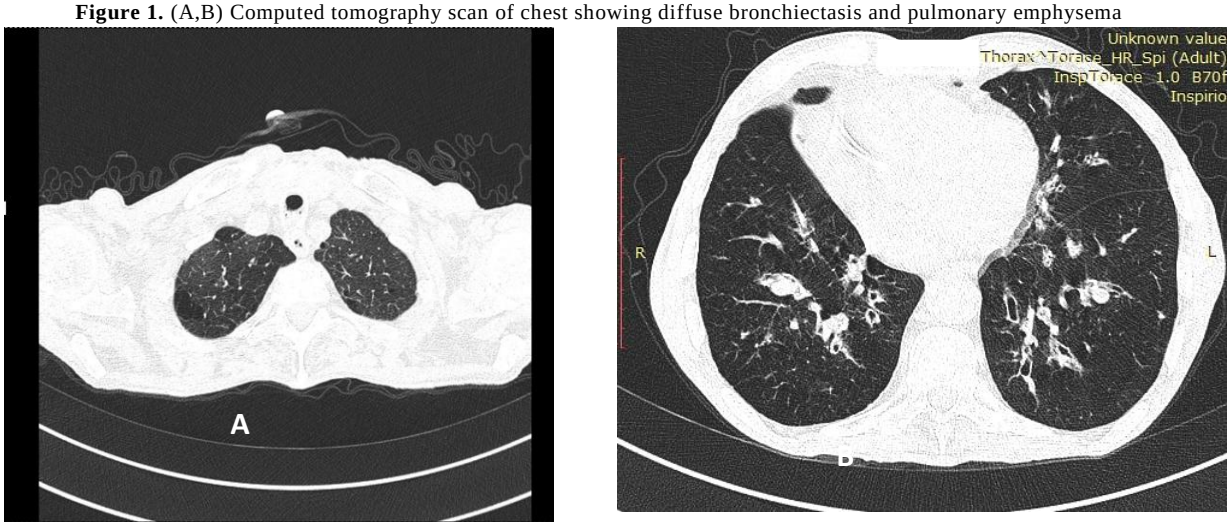
epilepsy, irbesartan (300 mg once/daily) for systemic arterial hypertension, esomeprazole (20 mg once/daily) for erosive gastritis and mesalazine (800 mg once/daily) for ulcerative colitis. At the last outpatient visit his vital signs were: systemic blood pressure 130/75 mmHg, pulse frequency 84/min rhythmic, body axillary temperature 36,5°C, oxygen saturation value of 90% (when breathing room air) with a respiratory rate of 16 breaths per minute. His height was 167 cm, weight of 42 Kg with a body mass index of 15 Kg/m^2. Physical examination of the chest revealed only the presence of a diffuse reduced intensity of the vescicular breath sound. Not other physical signs outside the chest were phatological. An arterial blood gas analysis (performed when the patient was breathing room air) showed: pH of 7.41, an arterial partial pressure of oxygen of 46 mmHg, an arterial partial pressure of carbon dioxide of 48 mmHg. Routine blood laboratory tests were normal. The last concentration of the serum immunoglobulin assay (measured one month after the last intravenous administration of IgG) is reported in table 1.

Table 1. Serum immunoglobulins concentration one month after last administration of immunoglobulin.

Serum immunoglobulins concentration	normal values	
Immunoglobulin A	70-400 mg/dL	5 mg/dL
Immunoglobulin G	700-1600 mg/dL	459 mg/dL
Immunoglobulin M	40-230 mg/dL	4 mg/dL

Figure 1. (A,B) Computed tomography scan of chest showing diffuse bronchiectasis and pulmonary emphysema



The last computed tomography scan of the chest showed the presence of diffuse and bilateral bronchiectasis together pulmonary panacinar emphysema (figures 1 and 2).
The temporal variation of the forced expiratory volume in the one second are showed in Figure 3.
The patient never followed our influenza and pneumococcal vaccination recommendations. During the 3 years of follow-up he had frequent (three in the last year) exacerbations of

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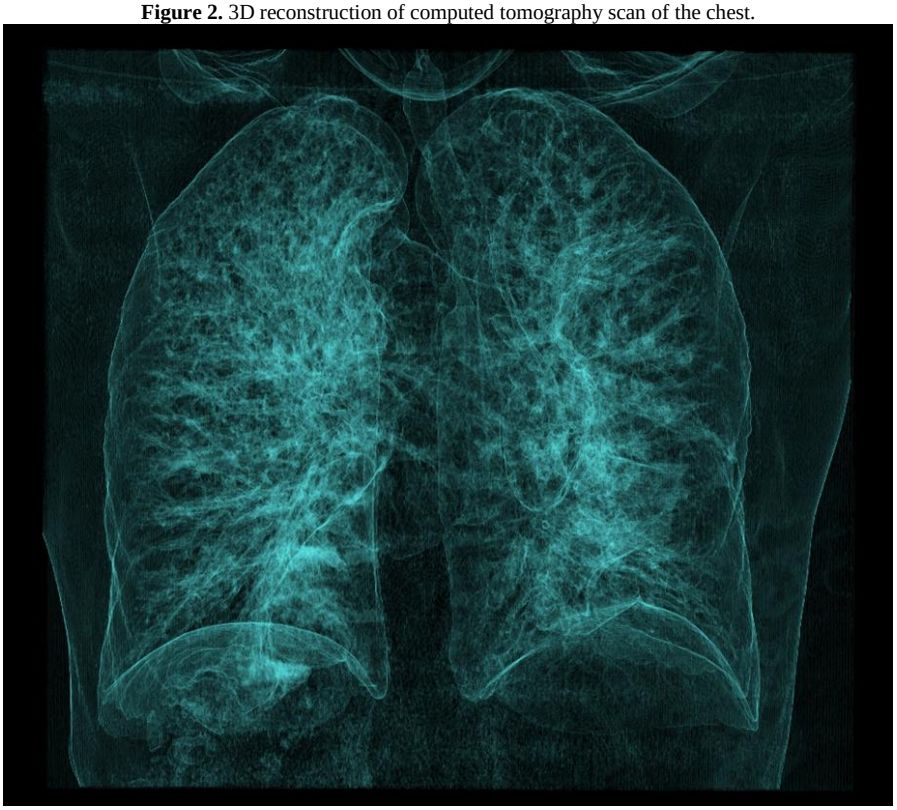
bronchiectasis with a progressive decline of the clinical condition until the death

Figure 2. 3D reconstruction of computed tomography scan of the chest.



Discussion

Primary hypogammaglobulinemias are disorders of heterogeneous etiology, characterized by low levels of serum immunoglobulins and impaired antibody production with increased susceptibility to pulmonary infections that often result in the onset of diffuse and bilateral bronchiectasis (3). The immunoglobulin G replacement therapy reduces the frequency of infectious episodes and prevents further destruction of the airways (1). The presence of bronchiectasis at the time of diagnosis is predictive of poor prognosis (4), and unfortunately the progression of the bronchiectasis occurs despite an adequate immunoglobulin replacement therapy (3). Indipendent predictors of mortality to the patients with bronchietasis include older age, low forced expiratory volume in the 1st second, low body mass index, previous hospitalizations, and > three exacerbations in the previous year (5).

The long-term management of patients with diffuse bronchiectasis is difficulty because outside of seasonal influenza and pneumococcal vaccination, there is little evidence that all the available drug treatment may significantly change the natural history (including prevention of exacerbations and hospital admissions) without causing significant side effects (6). The long-term oxygen therapy is often prescribed to these patients for it's effect on dyspnea but does not increase survival, at the opposite of what observed in chronic obstructive pulmonary disease (7). The long-term prognosis of the patients with bronchiectasis is poor and is influenced by the secondary cause (8).

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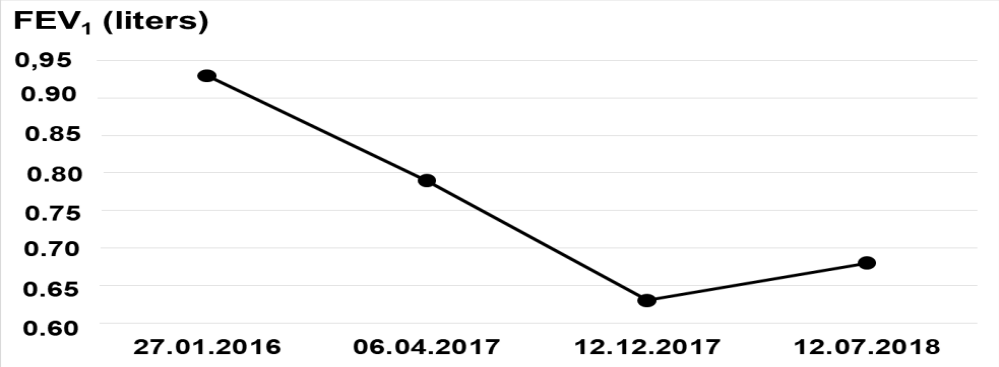
Figure 3. Variations of pre- bronchodilator forced expiratory volume in one second (FEV_1) over the years.



Conclusion

We report here an unusual case in an adult of diffuse and bilateral bronchiectasis secondary to congenital hypogammaglobulinemia.

Conflicts of Interest: There is no potential conflict of interest, and the authors have nothing to disclose. This work was not supported by any grant

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