

Desquamative interstitial pneumonia revisited half a century later

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Dear Editor,

I read with special interest the recent case report about respiratory bronchiolitis-associated interstitial lung disease (RB-ILD) described by Toma *et al.* (2017) in this Journal [1]. This uncommon smoking-related inflammatory disorder involves changes of bronchiolitis and of interstitial lung disease, and may share some characteristics with idiopathic interstitial pneumonia, idiopathic pulmonary fibrosis, idiopathic nonspecific interstitial pneumonia, desquamative interstitial pneumonia (DIP), cryptogenic organizing pneumonia, and acute interstitial pneumonia [1]. The reported 39-year-old male, cigarette smoker, had contact with parrots and cats, and clinical, imaging and histopathological characteristics consistent with RB-ILD [1]. The work focused differential diagnosis of this entity with Langerhans cell histiocytosis, cryptogenic organizing pneumonia, hypersensitivity pneumonitis, and DIP [1]. The authors called attention to similar histopathological features of RB-ILD and DIP causing diagnostic pitfalls [1]. Tan-pigmented macrophages in respiratory bronchioles and more accentuated fibrosis are indicative of RB-ILD, whereas the pathological changes have more diffuse and uniform pattern of distribution in patients with DIP [1]. Establishment of definite diagnosis depends on clinical, imaging, respiratory function tests, and biopsy data [1–5]. Smoking cessation and corticosteroid therapy may improve the quality of life of people with these conditions [1–5]. Although not consensual, some authors believe that RB-ILD and DIP are different stages of the same disease [1, 5].

DIP was first described in 1965 with the strong belief that desquamation of epithelial cells played the major role but additional evidence has confirmed the significant importance of intra-alveolar macrophages and giant cells [1–5]. Currently, classical data of DIP include intra-alveolar macrophages, thickened septa, and type II pneumocytes [4]. Therefore, in all patients with suspicion of DIP, lung tissue sampling is required to establish definite diagnosis [1–5].

In this interesting scenery, comments are added about the first case of DIP reported in a Brazilian patient (1969), which was the 39th case study of literature [2]. Prior reports of DIP were of Bates & Christie (1964) (one case) [6], Liebow *et al.* (1965) (18 cases) [7], Gaensler *et al.* (1966) (12 cases) [8], Steinberg (1966) (three cases) [9], Klocke *et al.* (1967) (two cases) [10], Schneider *et al.* (1967) (one case) [11], and Ansari *et al.* (1968) (one case) [12]. Rather than a current or former male cigarette smoker, the patient was a 14-year-old female without active or passive smoking. She was previously healthy, but ever lived in close contact with a sister treated for tuberculosis. There was neither pathological antecedent, nor occupational exposure or close contact with pets in her environment. Four months before admission, she had dry cough, dyspnea, weight loss, followed by fever and expectoration. Physical examination did not show cyanosis, digital clubbing, or lymph node changes; but pulmonary auscultation revealed inspiratory crackles predominantly in the medium lung thirds, and the rest of evaluation was unremarkable. Initially, a plain radiograph showed bilateral and symmetric interstitial and alveolar infiltrates more conspicuous in the lower regions of the lung fields. Additional plain-film tomography disclosed images of hyperexpansion in upper lobes with sub pleural blebs, and bilateral reticular interstitial changes and honeycomb features in lower lobes. Except for arterial pH 7.22 and oxygen saturation (SaO₂) 57%, routine tests were unremarkable, and the search for mycobacteria and fungi was negative in sputum. The respiratory symptoms worsened on the second week of admission due to acute pneumonitis by *Klebsiella* spp. that did not improve with treatment. The blood gas analysis of control revealed pH 7.1, SaO₂ 70%, total CO₂ 53.4%, partial pressure of carbon dioxide (paCO₂) 71.7 mmHg, HCO₃⁻ 21.8 mM/L, and base excess – 9 mmol/L [2]. Despite of intensive care support, corticosteroid, antibiotic, and cardiac glycoside therapy, she had irreversible circulatory shock developing after a spontaneous pneumothorax. Necropsy study showed consistent findings of DIP characterized by cell and reticulin proliferation thickening septa; cells isolated or arranged in layers within alveoli with bulky nuclei, prominent nucleoli, and abundant Periodic acid–Schiff (PAS)-positive cytoplasm; giant cells with few bulky nuclei; and “tan macrophages” with cytoplasm negative for hemosiderin; and fibroblast proliferation permeated by mononuclear infiltrate. Worthy of note, some mitotic figures were observed among the intra-alveolar desquamative cells [2].

It should be stressed that the non-smoker (~10%) female (~1:2 vs. male) with DIP [5] had only 14 years old; and the lack of clinical response to intravenous glucocorticoid was due to the severity of her general condition [2]. The initial concern was about the hypothesis of pulmonary tuberculosis based on household contact with her sister; but etiologic search included other agents (cytomegalovirus, hepatitis C, and *Aspergillus* spp.), connective tissue diseases (lupus, rheumatoid arthritis, and systemic sclerosis), pneumoconiosis, use of marijuana, and drug reactions [1–5].

Comments are herein shortly done about RB-ILD and DIP, two uncommon lung disorders that may share clinical, imaging and pathological features propitiating diagnostic pitfalls mainly for primary health care workers.

Conflict of interests

The author had full freedom of manuscript preparation and there is no potential conflict of interests.

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