
Granulomatous-lymphocytic interstitial lung disease at the emergency department: Think about it!

Sir,

Acute dyspnea is one of the most common causes of admission to the emergency department (ED), and sometimes, it can be ascribed to rare parenchymal cause, like in this clinical case of granulomatous-lymphocytic interstitial lung disease (GLILD).^[1,2]

We report a case of a 20-year-old woman presented at the ED with a nonproductive cough and progressive dyspnea, not responding to antibiotics. The patient had a history of noninvestigated recurrent pulmonary and sinus infections, undiagnosed common variable immunodeficiency (CVID). Blood test revealed low D-dimers levels (0.8 µg/mL) and negative electrocardiography for acute myocardial

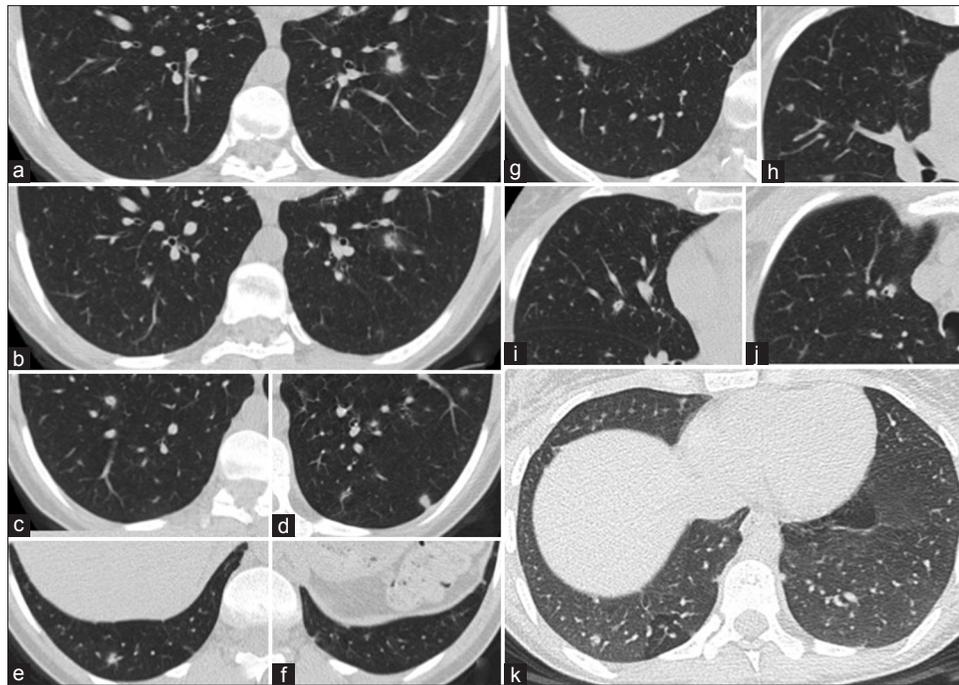


Figure 1: Axial high-resolution computed tomography showing multiple bilateral bronchocentric nodules and micronodules with prevalence in lower lobe zone (a-g) and in upper lobes lower segments (h-j) associated to follicular bronchiolitis (d and h). Expiratory acquisition demonstrates presence of lobular air-trapping (k)

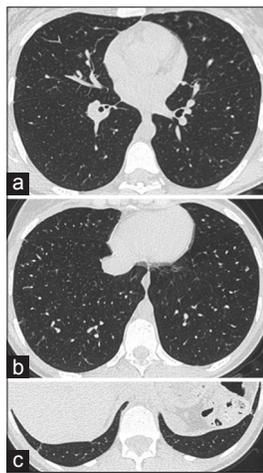


Figure 2: Posttreatment high-resolution computed tomography after 4 months shows complete disappearance of nodules and micronodules (a and b) without air-trapping in the expiratory examination (c)

infarction. C-reactive protein and erythrocyte sedimentation rate were slightly increased. Chest radiography was negative for the presence of pulmonary pneumonia or thoracic abnormalities. Abdominal ultrasound was also performed to exclude possible acute abdominal emergencies, and it revealed splenomegaly. Clinical examination showed no lymphadenopathies and/or chest or abdominal pain. Cough and dyspnea started 2 months before, and there was no fever or sweat. She was not a smoker. Her family history highlighted that her mother suffers from thyroiditis, celiac disease, and rheumatoid arthritis. After that, the patient underwent a computed tomography angiography (CTA) to

exclude the presence of pulmonary embolism. CTA resulted negative for pulmonary embolism but showed the presence of bronchial wall thickening and some nodules. Pulmonary function test showed reduced diffusing capacity of the lung for carbon monoxide with moderate restriction. For these reasons and to better evaluate the parenchymal findings, 2 days later, the patient underwent a high-resolution computed tomography (HRCT) of the chest that highlighted the presence of multiple nodules and micronodules of the lung parenchyma with lower lung zone predominance and prevalence in the peribronchial space, bronchial wall thickening, and air trapping in the expiratory scans, with no lymphadenomegalies [Figure 1]. Chest findings suggested the presence of infections or pulmonary inflammatory diseases. Bronchoscopy with bronchoalveolar lavage showed airway secretions negative for bacterial or fungal infections. The immune evaluation showed elevated CD4+ : CD8+ T-cell ratio and serum IgM and low levels of serum IgG and IgA. She had also undergone a splenectomy with the granulomatous disease of the spleen that confirmed the hypothesis of CVID. The patient started therapy with intravenous immunoglobulin and corticosteroids and after 4 months reached clinical improvement and disappearance of radiological signs [Figure 2].

GLILD can be observed as the pulmonary manifestation of common CVID, the most prevalent primary immunodeficiency.^[3] CVID has a prevalence of 1:50.000–1:200.000, characterized by low levels of serum IgG, IgA, or/and IgM and by susceptibility to recurrent infections, especially of the upper respiratory tract. GLILD can affect approximately 10%–30% of all CVID patients and can be associated with multisystemic

lymphoproliferative involvement (spleen, kidneys, and lymph nodes) with increased morbidity and mortality.^[4,5] HRCT thoracic findings include bronchial wall thickening, with air trapping in expiratory examination, ground-glass opacities, consolidations, nodules, fibrosis, bronchiectasis, emphysema, follicular bronchiolitis, lymphocytic interstitial pneumonitis, nodular lymphoid hyperplasia, and organizing pneumonia.^[4] All these signs might occur combined or separately; however, the pathognomonic characteristic element is given by the presence of granulomas with lymphoproliferative infiltrates.

In our experience, the most common parenchymal finding was the presence of nodules and micronodules with greater distribution in the peribronchovascular space, lower lobe predominance, and also basal segments of upper lobes. Mild follicular bronchiolitis and air-trapping were also present; they disappeared after medical treatment.

Different pathologies can mimic nodular infiltrates such as other granulomatous disease (histiocytosis sarcoidosis), pulmonary infections, or other primary immunodeficiencies.^[5,6] Lung biopsy should be performed when possible and reveal in most cases the presence of nodular peribronchiolar inflammation with lymphocytes. In our patient, the lack of the lung biopsy was replaced by splenectomy with proven granulomatous disease. Pulmonary infections were excluded after bronchoalveolar lavage such as sarcoidosis, in association with the poor upper lobe and scissural involvement in contrast to our case. From a technical point of view, the lack of HRCT scan of the lung at the first CT examination made it necessary for the young patient to undergo two different CT examinations. Considering that acute dyspnea could be ascribed to both parenchymal and vascular causes, it should be advisable to perform an HRCT scan of the lung (with a slice thickness not >1.00mm) before contrast medium administration in patients referred to the ED with acute dyspnea. In the near future, relatively new techniques such as fluorodeoxyglucose positron emission tomography-CT scan could be used to assess and monitor GLILD because of their capability to mesh functional and anatomical findings with a multi-systemic metabolically high activity approach.^[7]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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