The Rarest of All: Organizing Pneumonia in Adult-Onset Still's Disease


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Introduction: Adult-onset Still's disease (AOSD) is an idiopathic systemic inflammatory disorder, typically presenting with rash, leukocytosis, fever and arthralgia/arthritis. AOSD-associated pulmonary manifestations such as interstitial lung disease, pleuritis, pleural effusion, mediastinal lymphadenomegaly, pneumomediastinum, and organizing pneumonia (OP) have been poorly reported in the literature [1]. To our knowledge, described below is the 5th known case of biopsy-proven OP in AOSD [2-5]. Case: A 52-year-old woman with recently diagnosed AOSD, notable for multiple flare-ups coinciding with steroid taper, despite treatment with methotrexate and prednisone 60mg daily, presents with fever, pleuritic chest pain, dyspnea, and dry cough for 5 days. Upon arrival, she was hypotensive, and tachycardic. Chest CT revealed dependent ground-glass opacities, interlobular septal thickening, confluent consolidations within the right upper and bilateral lower lobes, and multiple subpleural nodular opacities. Despite fluid resuscitation and antibiotics, she required admission to the medical intensive care unit (MICU) for shock and acute hypoxic respiratory failure. Extensive infectious workup including multiple cultures, bronchoalveolar lavage from diagnostic bronchoscopy, and Pneumocystis jiroveci pneumonia yielded negative results. Given her immunocompromised state she was empirically started on doxycycline and cefepime for pneumonia, and methylprednisolone 60 mg every 6 hours for suspected AOSD flare-up. After an improvement in clinical status, it was changed prednisone 60mg daily and subsequently discontinued in 3 days. Soon after the discontinuation, she again became hypotensive with worsening hypoxia. She underwent video-assisted thoracoscopic surgery for lung biopsy. Pathology findings were consistent with OP. Treatment with methylprednisolone 125mg twice a day led to marked respiratory and hemodynamic improvement, then it was tapered slowly over 2 weeks to prednisone 60mg daily. She was discharged home in baseline respiratory status to be followed with outpatient steroid taper. Discussion: Typically a disease involving the skin and joints, AOSD is rarely associated with respiratory manifestations. Oftentimes, AOSD patients may have concurrent respiratory infections due to their immunocompromised state, which makes the diagnosis of OP challenging. A lack of improvement with antibiotics in acute respiratory failure in pneumonia of unknown infectious etiology should call for considerations of noninfectious work-up, such as an unusual presentation of OP secondary to AOSD. Failure to recognize the need of high-dose steroids accountable for clinical improvement can negatively impact the patients, especially those with steroid-dependent AOSD [6].

Figure 1.1 CXR with diffuse airspace opacities with increased vascular markings.

Figure 1.2 CT lung with bilateral posterior
with bilateral posterior consolidation, ground-glass opacities, and interlobular septal thickening.

Figure 1.3 The image shows proliferating fibroblasts (black arrows) inside an alveolar space. White arrows indicate alveolar lining cells, some of which are enlarged in a reactive type appearance. Mix