Since January 2020 Elsevier has created a COVID-19 resource centre with free information in English and Mandarin on the novel coronavirus COVID-19. The COVID-19 resource centre is hosted on Elsevier Connect, the company's public news and information website.

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Diffuse Lung Disease

SESSION TITLE: Fellows Diffuse Lung Disease
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CHILDHOOD-ONSET INTERSTITIAL LUNG DISEASE: A RARE CASE OF IDIOPATHIC, NONSPECIFIC INTERSTITIAL PNEUMONIA IN A TEENAGER

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INTRODUCTION: Childhood interstitial lung disease (chILD) is remarkably rare with a reported prevalence from 0.13 per 100,000 children under 17 years to 16.2 per 100,000 children under 15 years of age (1). Here we present a case of a teenager with idiopathic nonspecific interstitial pneumonia (NSIP).

CASE PRESENTATION: A 15-year old previously healthy female presented with a three-week history of dry cough, fever, malaise and shortness of breath which progressed to dyspnea on exertion despite a course of outpatient antibiotics. She had no environmental exposures, history of smoking or vaping nor family history of pulmonary pathology. Initial vital signs were significant for hypoxemia to 80% and a chest x-ray and CT scan with bilateral consolidations and ground glass opacities. Initial laboratory tests showed elevated inflammatory markers, other tests including SARS-CoV-2 PCR were negative . Bronchoscopy, bronchoalveolar lavage, and transbronchial biopsies yielded no definitive diagnosis. Following bronchoalveolar lavage, she developed rapid clinical deterioration necessitating mechanical ventilation, paralytics, and inhaled nitric oxide. Given elevated inflammatory markers and after infection was excluded, intravenous steroids were initiated with partial improvement. After extubation, a protracted inability to wean off oxygen support, and spirometry showing ongoing, severe restrictive lung disease, lung biopsy via video-assisted thorascopic surgery was performed lending to the ultimate diagnosis NSIP. Hypersensitivity pneumonitis panel, ILD panel and genetic testing were unrevealing, as such her case was classified as idiopathic NSIP.

DISCUSSION: NSIP in adolescents is very rare. A meta-analysis (2) reports the average age of patients with NSIP to be 54.8 years. In the pediatric population, NSIP can be associated with systemic disease processes such as collagen vascular disease, including systemic lupus erythematosus, juvenile rheumatoid arthritis, and dermatomyositis associated with antibodies to melanoma differentiation-associated gene 5 (anti-MDA5), as well as a complication of blood stem cell transplants and surfactant dysfunction mutations, although most cases are idiopathic (3). Given that the etiology of NSIP is broad and in most cases idiopathic, response to corticosteroids is also variable. Our patient had a partial response to corticosteroids early on in her treatment, manifested by augmented gas exchange and improved symptomatology. The relatively slow response to treatment in the first several weeks is likely consistent with her biopsy findings which showed fibroblast proliferation and fibrosis.

CONCLUSIONS: ILD and particularly NSIP are rare conditions in adolescents but should be suspected in patients with hypoxemia and abnormal imaging in the absence of infectious or autoimmune etiologies. An extensive workup is recommended to determine association with systemic conditions, treatment and prognosis.

Reference #1: (1) Griese M, Haug M, Brasch F, et al. Incidence and classification of pediatric diffuse parenchymal lung diseases in Germany. Orphanet J Rare Dis. 2009;4:26. Published 2009 Dec 12. doi:10.1186/1750-1172-4-26

Reference #2: (2) Ebner L, Christodoulidis S, Stathopoulou T, et al. Meta-analysis of the radiological and clinical features of Usual Interstitial Pneumonia (UIP) and Nonspecific Interstitial Pneumonia (NSIP). PLoS One. 2020;15(1):e0226084. Published 2020 Jan 13. doi:10.1371/journal.pone.0226084

Reference #3: (3) Vece TJ, Fan LL. Interstitial Lung Disease in Children Older Than 2 Years. Pediatr Allergy Immunol Pulmonol. 2010;23(1):33-41. doi:10.1089/ped.2010.0008

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