PJP as the Index Presentation of Dermatomyositis with RP-ILD

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Presentation

32-year-old woman presents with 3 days of nonproductive cough and exertional dyspnea.

ROS: +diffuse arthralgias, +oral ulcers, +rash.

PMH: Recent pregnancy loss at 22 weeks' gestation.
Meds: Ibuprofen.
FH: Mother with lupus; son with celiac disease.
SH: Grew up in Mexico

Initial Workup

Exam: SpO2 91%; oral ulcers; hyperpigmented papules on thighs; cracked skin on the sides fingers; hip flexor weakness; mild synovitis of symmetric small- and medium-sized joints.

Labs: Mild anemia, hepatocellular liver injury.
CT chest: Bilateral, patchy perihilar consolidations and mildly enlarged mediastinal lymph nodes.

Course

Treatment for community-acquired pneumonia.

Worsening hypoxemic respiratory failure.

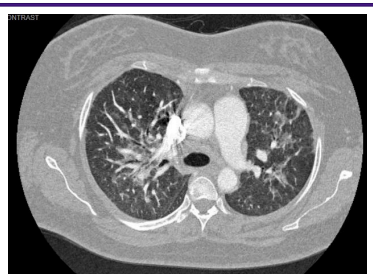

Figure:
Initial CT chest with contrast.

Additional Data

+Anti-MDA5 antibodies; worsening consolidations & groundglass on CT

DFA stain of broncho-alveolar lavage fluid +PJP.

Treatment

Trimethoprim-sulfamethoxazole IV -> primaquine + clindamycin.

Tacrolimus + prednisone.

Discharge home with family on day 21 after initial presentation.

Outcome

On day 35, presented with worsening hypoxemic respiratory failure requiring mechanical ventilation. Bronchoalveolar lavage studies were negative.

Treatment with pulse-dose steroids, rituximab, tofacitinib, continued tacrolimus.

Development of *E. faecalis* bacteremia.

Death due to septic shock on day 45 after initial presentation.

Background

> Rapidly progressive interstitial lung disease (RP-ILD) associated with anti-MDA5+ dermatomyositis (DM) carries a poor prognosis.

> Early, aggressive immunosuppression has been associated with improved survival (33% prior to initiation of a combined protocol vs. 89% after).[1]

> *Pneumocystis jirovecii* (PJP) pneumonia in patients with ANA-associated diseases is associated with high risk of mortality (for example, 45.7% in a single-center cohort).[2]

Discussion

> PJP pneumonia has previously been noted in patients with MDA5+ DM in the context of immunosuppression.[3,4]

> To our knowledge, this is the first report of anti-MDA5+ DM presenting with PJP pneumonia prior to initial DM diagnosis and immunosuppression.

> Consolidations and ground-glass on chest CT can be consistent with infection or RP-ILD. Respiratory failure was initially attributed to PJP pneumonia.

> Simultaneous autoimmune and infectious attack of the same organ raises diagnostic & management challenges.

Clinical Pearls

> Simultaneous infectious and autoimmune attack in the same organ can occur.

> PR-ILD is a feature of anti-MDA5+ inflammatory myositis and warrants early and aggressive immunosuppression.

> Consider a broad differential in patients presenting with clinical features of pneumonia and radiographic features of consolidations and ground-glass opacities.

References: 1. Tanizawa, et al, Arth Rheum, 2020. 2. Ward & Donald, Arth Rheum, 1999. 3. Quintana-Ortega, et al, Mod Rheum Case, 2021. 4. Seiller, et al, Annales de derm, 2020.

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