Cirrhosis Associated Immune Dysfunction: Diffuse Curvulvaria Infection, CMV Viremia and PJP Pneumonia in an End-Stage Liver Disease Patient

Stavros A Doumas MD MSc[1], Albert C Shu MD[1], Pitchayut Nithagon MD[2], Veronica Nguyen MD[3], Amol Rangnekar MD[3]

1 Department of Medicine, MedStar Georgetown University Hospital, Washington DC, USA., 2 Department of Pathology, MedStar Georgetown University Hospital, Washington DC, USA., 3 Transplant Institute, MedStar Georgetown University Hospital, Washington DC, USA.

GEORGETOWN UNIVERSITY

MedStar Georgetown University Hospital

Background

- Infections are a major cause of morbidity and mortality in end-stage liver disease (ESLD)
- Cirrhosis-associated immune dysfunction (CAID) refers to the immune system dysregulation observed in the setting of ESLD
- Innate and adaptive immunity dysfunction is clinically evident by increased susceptibility to bacterial, fungal, and viral infections
- We present the case of a young patient with multiple concomitant opportunistic infections attributed to immune dysfunction secondary to ESLD

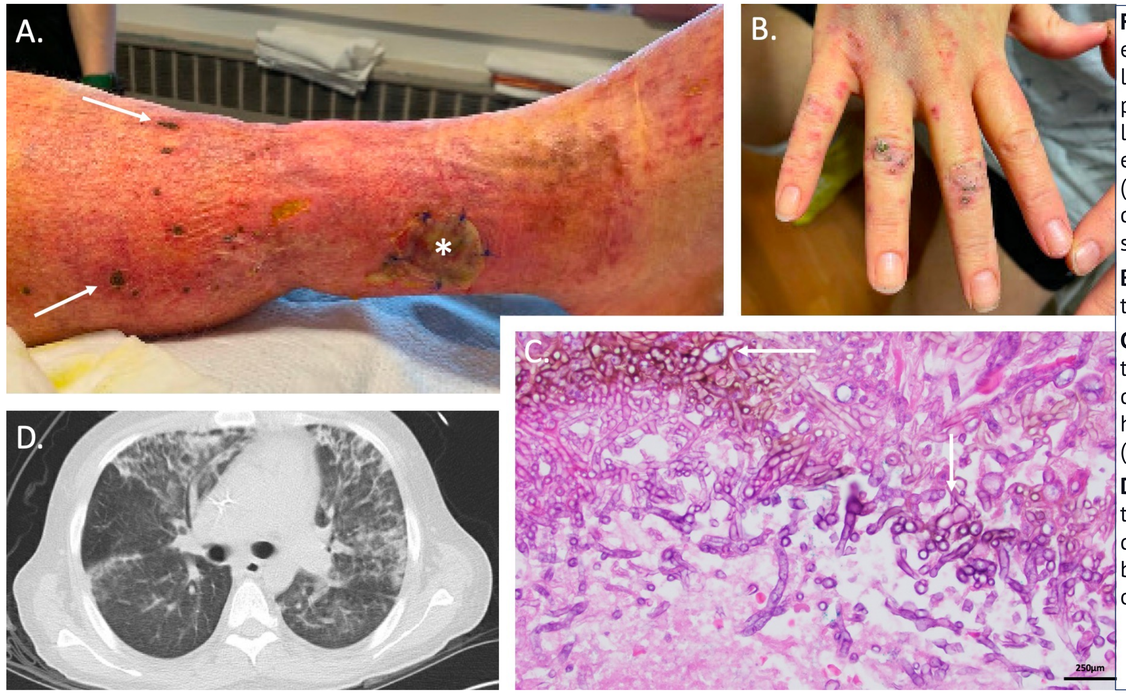

Figure 1 A. Picture of lower extremity lesions. Diffuse lower extremity confluent petechial rash with necrotic lesions covered by black eschar (white arrows). The (*) demonstrates the site of deep tissue biopsy sampling.

B. Representative lesions on the hand.

C. H & E stain of deep tissue biopsies demonstrating branching hyphae with angioinvasion (white arrows).

D. Chest computed tomography (lung window) demonstrating multifocal bilateral ground-glass opacities

Case Description

A 36-year-old male with a prior medical history of asthma and alcohol-related cirrhosis presented with abdominal distension, jaundice, and lower extremity swelling. Physical examination was notable for lower extremity scabbed black lesions. Model for End-Stage Liver Disease with sodium (MELD-Na) score was 34, and he was admitted for liver transplant evaluation. Admission serologies demonstrated high CMV IgG/IgM titers with a positive CMV PCR, and treatment was initiated. Over the first week, a coalescing petechial rash developed around the leg wounds and gradually spread to the upper extremities. Swab cultures grew Curvularia, a facultative pathogen mold found in decaying plants. Deep tissue sampling demonstrated branching hyphae with evidence of angioinvasion, and cultures confirmed Curvularia, prompting antifungal therapy initiation. Immunodeficiency work-up was negative for known conditions but was notable for low CD4, CD19, CD16+56+ counts, and low total IgG (normal on admission). The patient developed a productive cough, and chest imaging demonstrated multifocal pneumonia. Bronchoalveolar lavage was positive for PJP. The patient's clinical condition progressively deteriorated, requiring transfer to the intensive care unit for continuous renal replacement therapy and mechanical ventilation. Ultimately, after a two-month-long hospitalization, the patient passed away from sepsis-associated multi-organ failure.

Discussion

- We present a case of severe immune dysregulation attributed to ESLD
- The multiple concomitant opportunistic infections and observed immune abnormalities are evidence of global immune system dysfunction
- Notably, this is the first description of diffuse Curvulvaria infection in the setting of cirrhosis

Conclusions

- The spectrum of cirrhosis associated immune dysfunction (CAID) can be severe enough to mimic profound immunodeficiency states
- Clinicians should consider atypical infections when evaluating cirrhotic patients
- More research is called for to define the incidence of various opportunistic infections and elucidate the liver's role in immune homeostasis in both health and disease

References

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