RARE CASE OF HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS SECONDARY TO TULAREMIA

Madison Bulger, M3
(Daniel Woolery, D.O.)
Pulmonology Critical Care

INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) is a rare, life-threatening syndrome of excessive immune activation that can be acquired genetically or sporadically. It is triggered by events that disrupt immune function and has a variable presentation that has a median survival of 2 months without prompt treatment. Tularemia is a highly infectious zoonotic illness caused by *Francisella tularensis* that most commonly results in ulcero-glandular disease. We have been unable to find previously documented cases of HLH secondary to tularemia.

CASE DESCRIPTION

We present a case of a 62-year-old female who originally presented to the emergency department with severe sepsis, leukopenia, acute kidney injury, and transaminitis. She was fluid resuscitated and started on broad spectrum antibiotics. An ulcerative lesion was noted in the left groin which was debrided. The patient's workup revealed pancytopenia, hypertriglyceridemia, ferritin >50,000 ng/mL, transaminitis, coagulopathy, and elevated soluble CD25 which met the diagnostic criteria for HLH. The patient was started on etoposide and dexamethasone per the current treatment guidelines of HLH, as no infectious source had been elicited. On day six, blood cultures from admission returned positive for *Francisella tularensis* and the patient was transferred to a tertiary care center due to its status as a potential bioterrorism agent. Following transfer, she was treated with gentamicin and other supportive care. She was discharged 31 days after initial admission.

DISCUSSION

This case illustrates what we believe to be the first reported case of HLH secondary to tularemia. In Missouri, tularemia should be considered early as a potential source of HLH.