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Case report

Rash, disseminated intravascular coagulation and legionella: Episode 10 and a rewind into the past

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A B S T R A C T

Legionella pneumophila is the most common cause of legionellosis and is one of the organisms causing atypical pneumonia. We report the presentation of disseminated intravascular coagulation (DIC) and skin rash in a single case of severe *Legionella pneumonia*. The unique clinical presentation of a diffuse rash diagnosed as purpura fulminans and the unpredictable variations encountered during the diagnostic work-up of the case make this write-up crucial. This article synthesizes all reported cases of *L. pneumonia* associated with cutaneous manifestations as well as cases presenting with DIC. Furthermore, this manuscript illustrates the correlation between cutaneous and coagulopathic manifestations, and morbidity and mortality from *L. pneumonia*.

Introduction

Cutaneous manifestations in Legionellosis are very uncommon. They may present as maculopapular, erythematous, or petechial skin lesions [1]. About 9 cases thus far have been reported describing a rash associated with Legionella infection. A clear mechanism for the rash was not evident in the majority of them. Another uncommon, but well-described phenomenon associated with *Legionella pneumophila* and *Legionella longbeachae*, is disseminated intravascular coagulation (DIC) [2]. Legionella urine antigen testing is the main diagnostic test utilized to detect *L. pneumophila*. However, it has about a 25–30% false negative rate [3]; awareness about this is lacking and a failure to cover for organisms like legionella early during the illness may lead to uncontrolled endotoxin-related phenomena such as DIC. We hereby elucidate a complicated case of *L. pneumophila* accompanied by a clinically visible rash as well as coagulopathy culminating in respiratory failure and shock. We have also extracted data from various reference sources including PUBMED, EMBASE, MEDLINE and Ovid, to provide a consolidated view of all reported cases of legionella associated with cutaneous manifestations and DIC.

Case presentation

A 44-year-old male with a past medical history of hyperlipidemia was brought to the ER with a one-week history of cough, body aches, fever, fatigue and a red maculopapular rash on the inner thighs. At presentation, he was in respiratory failure, underwent emergent endotracheal intubation and mechanical ventilation and was admitted to the medical ICU. Initial chest X-ray and CT scan revealed right middle and lower lobe pneumonia (See Figs. 1 and 2). Labs tests showed leukopenia (WBC 1.6x10E+09/L) and thrombocytopenia (platelet count 94,000). He was started on broad-spectrum antibiotics including vancomycin, ciprofloxacin, metronidazole, and doxycycline. Twenty-four hours into hospitalization, the patient's rash became more confluent, with dark necrotic-appearing areas (see Fig. 3), and spread to involve the arms, legs, trunk, tip of the nose and left ear along with acral cyanosis. Due to concern for infective endocarditis, a trans-thoracic echocardiogram was performed that was reported as negative for any vegetation. Serological testing including viral studies, Lyme antibody (Ab), and Rickettsia Ab were negative. Routine blood, urine, and sputum cultures were also negative. Although the initial Legionella urinary antigen testing was reported negative, subsequent repeat analysis in the ICU was positive. In the ICU, the initial serology titers for legionella serogroup 1 were positive at 1:64 and subsequently 1:1024 during the first week. His antibiotics were then adjusted to include ceftriaxone, doxycycline, and moxifloxacin. Simultaneously, work-up for his rash was undertaken and a biopsy was

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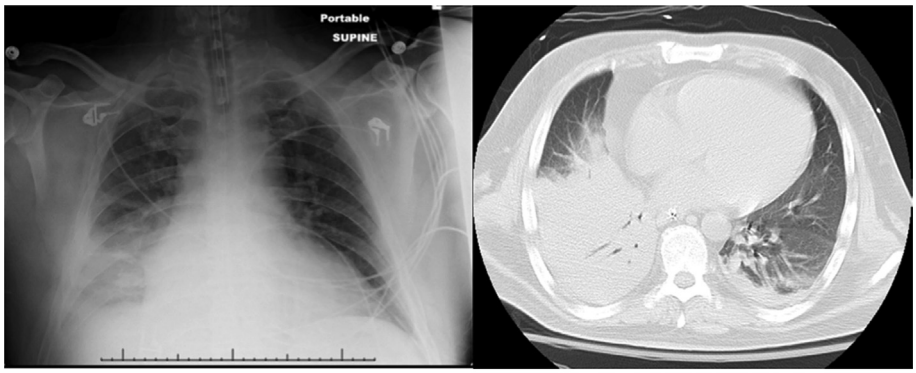

Fig. 1. ICU admission chest X-ray: consolidation at right base; perihilar pulmonary congestion. CT chest: right middle lobe and right lower lobe pneumonia.

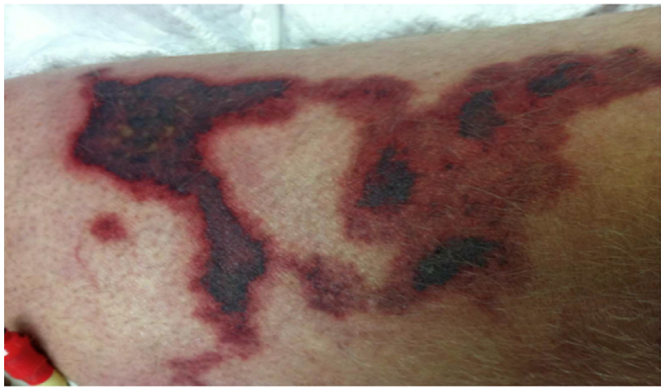

Fig. 2. Image of skin findings: diffuse maculopapular rash with dark necrotic-appearing areas.

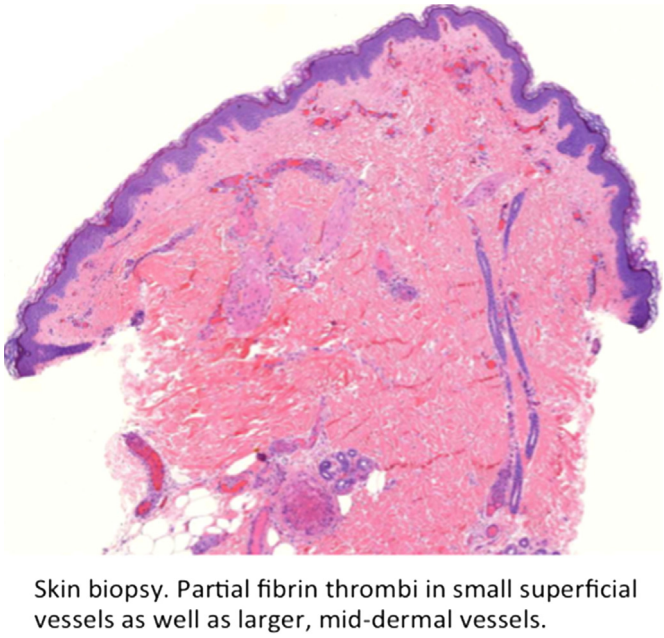

Fig. 3. Skin biopsy histopathology: partial fibrin thrombi in small, superficial vessels as wells as larger mid-dermal vessels.

obtained from his right thigh. Histopathology revealed partial fibrin thrombi in small, superficial vessels as well as larger mid-dermal vessels and fibrinoid degeneration of the vessel walls, overall consistent with a coagulopathy (see Fig 4). The differential diagnosis included DIC, thrombotic thrombocytopenic purpura (TTP), heparin-induced thrombocytopenia (HIT), and anti-phospholipid antibody syndrome. Subsequent work-up revealed a negative HIT panel. Thrombocytopenia with a normal creatinine was not consistent with TTP. He was finally diagnosed with DIC secondary to *Legionella pneumonia* based on elevated fibrin split products (FDP) and decreased fibrinogen levels. Under appropriate antibiotic coverage, his clinical status improved. The diffuse rash cleared, and the leukopenia as well as thrombocytopenia resolved. He was extubated later during the ICU course and was discharged home within 2 weeks.

Discussion

Legionnaire's disease was discovered in 1976 after an outbreak of pneumonia at an American Legion convention in Philadelphia. The causative organism was later isolated as *L. pneumophila*, an aerobic gram-negative rod. Legionellosis comprises of two syndromes, Legionnaire's disease and Pontiac fever. Legionnaire's disease refers to severe pneumonia that can be associated with multi-system disease [4]. Pontiac fever is an acute, self-limited, febrile illness sparing the lungs. About 64 serogroups of *L. pneumophila* have been identified but serogroup 1 is responsible for 70–90 percent of cases. It is transmitted by the aspiration of water contaminated with Legionella and not by person-to-person contact. This can originate from humidifiers, air conditioning, showers, respiratory therapy equipment, etc. Normally, mucociliary action helps clear Legionella in the upper respiratory tract. Organisms that reach the alveoli are consumed by macrophages, multiply within these cells until rupture, and then infect other macrophages. Legionella causes an acute fibropurulent pneumonia with alveolitis and bronchiolitis [5]. It can later affect other organs of the body like the kidneys, liver, brain, and spleen. Symptoms are non-specific including fever, fatigue, headache, confusion, and lethargy [6]. The causative organism, clinic-radiological dissociation, absence of lobar pneumonia in the early phase and paucity of symptoms seen in bacterial pneumonias make it definable as an atypical pneumonia. The mortality rate in Legionnaire's disease is 5–80% depending on certain risk factors like age, underlying