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# Acute Q fever posing a diagnostic dilemma: a case report

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**ABSTRACT:** Q fever is a worldwide zoonosis, caused by *Coxiella burnetii*, that can be easily diagnosed in its usual clinical presentation. However, Q fever may mimic systemic autoimmune and inflammatory disorders making its recognition very challenging. We present a case of acute Q fever presenting in a patient with latent undiagnosed. Systemic Lupus Erythematosus (SLE): diagnostic challenges, potential pitfalls and therapeutic issues encountered in the management of this patient were presented and discussed.

— **Key words:** Q fever, *Coxiella burnetii*, Systemic Lupus Erythematosus, Diagnosis.

## INTRODUCTION

Q fever is a self-remitting zoonosis caused by *Coxiella burnetii* (*C. burnetii*), a gram-negative intracellular bacteria. Acute presentation of disease is characterized by atypical pneumonia, hepatitis, endocarditis/pericardial effusion along with arthralgia and myalgia<sup>1</sup>. Diagnosis is based on the presence of typical clinical findings along with a specific serological response: an indirect immunofluorescence IgM ratio of 1:50 with a 1:200 IgG ratio has been proposed to be as high as 100% specific for acute Q fever<sup>2</sup>.

However the presence of some clinical features of acute Q fever (such as arthritis, rash, pleural and pericardial effusion) side by side with the finding of positivity for antinuclear, anti-smooth muscle and antiphospholipid antibodies<sup>3</sup> may make very difficult the differential diagnosis with autoinflammatory and autoimmune disease especially in low endemic areas.

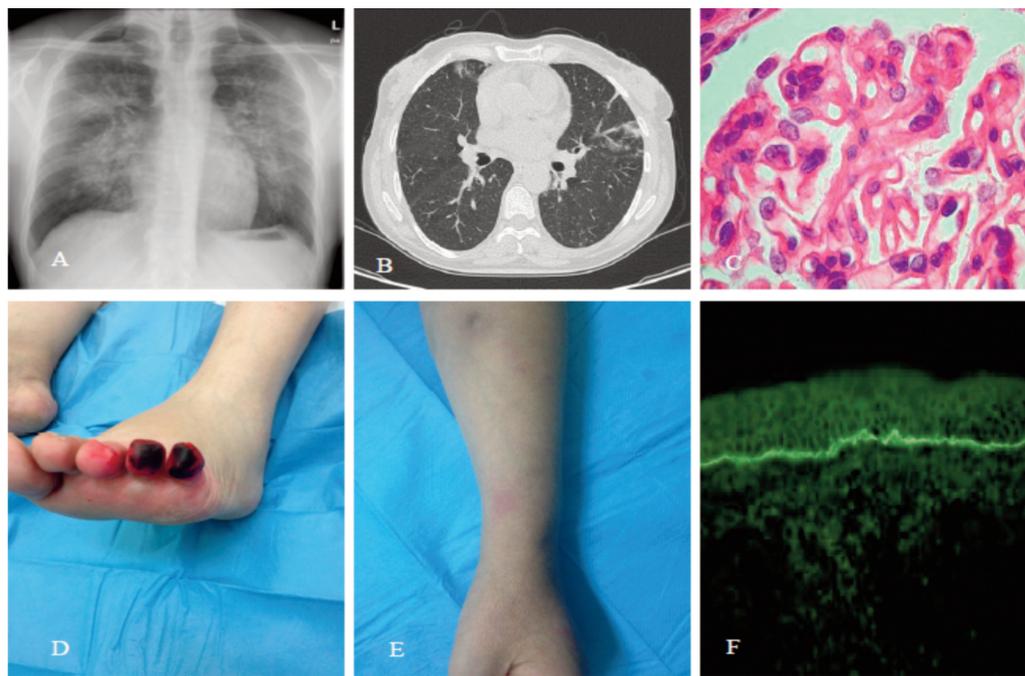
## CASE REPORT

A 52-year old woman was admitted to our hospital because of mild pyrexia, fatigue, night sweats and polyarthralgia. She was of rural origin and she reported frequent close

contacts with household animals. Her past medical history was uneventful apart from a four-year long lasting history of Raynaud's phenomenon and arthralgia.

Physical examination revealed diffusely decreased breath sounds at chest auscultation, soft hepatic enlargement and bilateral arthritis of wrists and metacarpophalangeal joints. Investigations showed mild anaemia, with neutrophil leucocytosis and raised erythrocyte sedimentation rate and C-reactive protein. Chest X-ray revealed bilateral pulmonary infiltrates (Figure 1A) appearing as diffuse patchy ground-glass opacities at the chest HRCT (Figure 1B). Routine and bronchioloalveolar lavage liquid cultures were negative for common aerobic and anaerobic bacteria, opportunistic and acid-fast bacilli.

Demonstration of a >4-fold rise of IgM type antibodies against phase II *C. burnetii* antigens prompted us to diagnose acute Q fever. Antimicrobial therapy with tetracycline was then started. Routine echocardiography performed to unmasking asymptomatic endocardial involvement, unexpectedly revealed a significant pericardial effusion. In parallel, autoantibodies testing documented positivity for ANA, anti-Smith, anti-DNA and anti-RNP antibodies. Rheumatoid factor was negative and complement fractions levels appeared mildly reduced. Systemic lupus erythematosus (SLE) was then



**Figure 1.** Clinical, instrumental and histopathologic features. Chest X-ray reveals bilateral pulmonary infiltrates (A) appearing as diffuse patchy ground-glass opacities at the chest HRCT (B). After two months of follow-up the patient developed a nephrotic syndrome due to membranous glomerulonephritis (C) and critical limb ischemia related to severe Raynaud phenomena (D) and livedo reticularis (E). Lupus band was also positive (F).

suspected and a medium-dosage prednisone (10 mg/day) was added to antimicrobial therapy.

At 2-weeks follow-up chest/pericardial disease and arthritis appeared to be dramatically improved and serology for *C. burnetii* markedly reduced. Patient was discharged with the diagnosis of Q fever and suspected SLE and a medium-dosage therapy with prednisone was suggested.

Early after two months of follow-up the patient developed critical limb ischemia related to severe Raynaud phenomena (Figure 1D), livedo reticularis (Figure 1E) and nephrotic syndrome due to membranous glomerulonephritis (Figure 1C). Continuous treatment with endovenous prostacycline analogue, and heavy immunosuppression with steroids and mycophenolate mophetil leads to improvement of vascular and glomerular features. Serological testing for *C. burnetii* performed at 3 months showed a negative result for IgM and a significant reduction of IgG. Lupus band test returned also positive (Figure 1F). A diagnosis of acute Q fever in a patient with “full blown” SLE was then made.

## DISCUSSION

Acute Q fever clinical presentation may be very protean varying from asymptomatic or a flu-like syndrome as in the Derrick’s original description to a critical illness with a fatality case rate of 1-2%<sup>4</sup>.

However, some clinical features of acute Q fever such as rash, arthralgia, pleural and pericardial effusion, and false positivity for ANA and antiphospholipid antibodies (reported to be as high as 30%)<sup>3</sup> may lead to a misdiagnosis of a systemic connective tissue disease.

In this case differential diagnosis between Q fever with false positivity for autoantibodies, or alternatively SLE with false positivity for *C. burnetii* serology or, ultimately, Q fever in the course of a mild SLE appeared

to be challenging: we inclined to favour the latter hypothesis owing to the presence of specific finding of Q fever (atypical pneumonia) side by side with typical features of SLE (Raynaud’s phenomenon, low complement fractions and positivity at medium high levels for anti-DNA antibodies). Short course of medium dosage-steroids has been advocated in case of incomplete response to antibiotic therapy in Q fever and may obviously have accelerated the improvement of SLE features in our patient. Hydroxycloquine use carrying beneficial effect in the treatment of both disease was not prescribed on the basis of known patient’s condition of glucose 6-phosphate dehydrogenase deficiency.

## CONCLUSIONS

Q fever should be considered as a differential diagnosis of SLE in patient presenting with atypical pneumonia along with specific feature of SLE in subjects exposed to risk factors for acute *C. burnetii* infection.

## CONFLICT OF INTERESTS:

The Authors declare that they have no conflict of interests.

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