



Disseminated *Mycobacterium avium* infection masquerading as longstanding polymyositis

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Immunosuppression is a risk factor for atypical mycobacterial infections, so suspicion should guide re-evaluation of negative results.

Case report

A 63-year-old lady presented in May 2010 with a three-month history of fever, weight loss, night sweats and a widespread rash. The initial diagnosis was a relapse of polymyositis. She had anti Jo-1 antibody-related polymyositis diagnosed in 1993 on clinical, serologic and electromyologic evidence. She had been treated with ≥ 15 mg prednisolone since 1993 and weekly oral methotrexate 15 mg since 2007. On examination she had multiple, tender, 10–20 cm diameter, well-demarcated, atrophic areas of erythema over her trunk and limbs, mild truncal weakness, fine inspiratory crackles at both bases, and an ejection systolic murmur in the aortic region.

Blood tests included haemoglobin (Hb) 10.1 g/dL, leucocytes $14.9 \times 10^9/L$, platelets $488 \times 10^9/L$, serum C-reactive protein (CRP) 127 mg/L, alanine transaminase of 23 U/L and a serum creatinine kinase (CK) 240 U/L. She remained positive for antibody directed against Jo-1. Urine culture and three sets of blood cultures were negative. An echocardiogram, chest radiograph, upper gastrointestinal and colonic endoscopies were normal. A CT scan of chest, abdomen and pelvis demonstrated previously documented bibasal pulmonary fibrosis. The clinical differential diagnosis included Sweet's syndrome, Panniculitis and Erythema Elevatum Diutinum. Two skin biopsies demonstrated focal granulomatous inflammation,

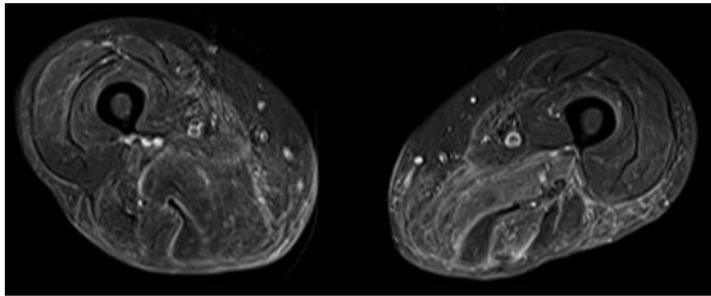
which raised the possibility of erythema nodosum or polymyositis-associated panniculitis. Acid fast stains were not performed and the biopsy was not cultured for mycobacteria at this stage.

The prednisolone was increased to 60 mg/day and methotrexate to 20 mg/week. Within 24 hours her pyrexia settled and symptoms improved. She was discharged from hospital with a steroid tapering regimen. Two weeks later, her CRP had fallen to 41 g/L, and the CK was 207 U/L.

After discharge, attempts to reduce prednisolone below 40 mg were met with a resurgence of symptoms. She was bed-bound and had watery diarrhoea, usually six episodes a day, and had lost approximately 15 kg of weight since the initial illness started. Multiple stool cultures were sent and special stains for mycobacteria were negative. She was re-admitted in August 2010 with high spiking temperatures. The skin lesions were more extensive covering her back, buttocks, thighs and arms, and at this stage a clinical diagnosis of cutaneous atypical mycobacterial infection was considered.

Laboratory tests included Hb 9.8 g/dL, CRP 240 mg/L, CK 21 U/L. The skin was re-biopsied and stains and skin cultures were specifically requested for mycobacterium. Magnetic resonance imaging sequences (MRI) demonstrated generalized fat atrophy of muscles, but low grade oedema of the left adductor magnus (Figure 1), which was biopsied. On gross appearance, the muscle was pale, watery and difficult to identify. Both biopsies demonstrated granulomatous changes with histiocytes packed with acid and alcohol fast bacilli (Figure 2). By contrast

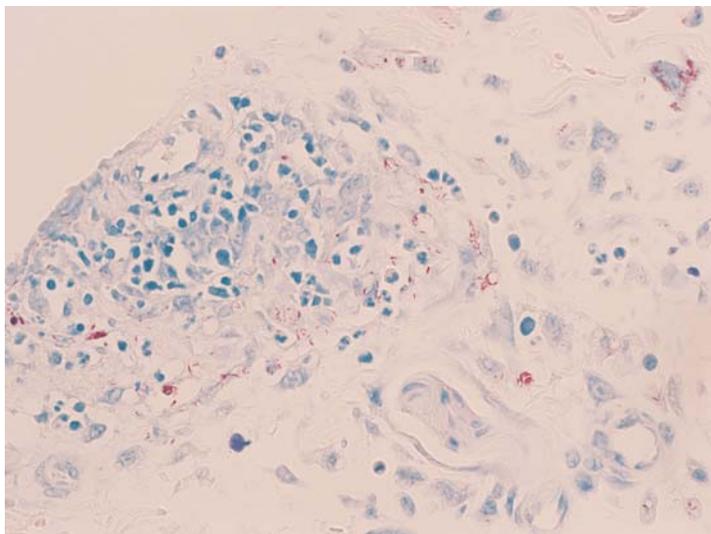
Figure 1
T2 turbo inversion recovery magnitude (TIRM) sequence of thighs demonstrating oedema of the left adductor magnus and semitendinosus



re-examination of the initial skin biopsies identified a single bacillus.

Cultures of the skin and muscle samples isolated *Mycobacterium avium*. On request, the initial stool samples were re-examined and mycobacteria were then identified. No specific sensitivities were reported except that empirical treatment would be sufficient. The patient was started on antimycobacterial chemotherapy (Rifater and Ethambutol with pyridoxine cover) and gradual prednisolone reduction was continued. At six weeks, her skin

Figure 2
Ziehl-Neelsen stain of skin biopsy demonstrating perivascular and loose foamy macrophages stuffed with acid-fast bacilli (magnification $\times 400$)



lesions were considerably improved, the diarrhoea had subsided and she was feeling stronger. Her blood tests were Hb 10.6 g/dL, CRP 48 mg/L, CK <20 U/L. She will need to complete at least 18 months of antimicrobials and is currently on a regimen of ethambutol 900 mg daily, rifampicin 600 mg and clarithromycin 500 mg twice daily. She had a delay in her treatment due to isoniazid or pyrazinamide induced hepatitis.

Discussion

Iatrogenic immunosuppression remains a significant risk factor for the development of atypical mycobacterial infection. A recent review has highlighted the increasing frequency of mycobacterial skin and deep tissue infections.¹ *Mycobacterium abscessus*, *fortuitum*, and *chelonae* are the usual pathogens.² Mycobacterial myositis has been reported in immunosuppressed patients³⁻⁵ and *M. avium* infection has been reported in polymyositis⁶⁻¹⁰ but to our knowledge this is the first instance of *M. avium* infection involving at least the skin, skeletal muscle and gut. Atypical mycobacterial infection can mimic flares of inflammatory conditions, with weight loss, temperatures and malaise, so there should be a high index of suspicion (particularly if immunosuppression is ineffective). It is important to request the correct stains and cultures to get an accurate result. In this case, a negative result was corrected after re-examination of the specimens. Accurate diagnosis of mycobacterial infection is improved by multiple sampling from different tissues.

Further investigation revealed that this patient had mycobacterial infection on her first admission. The original skin biopsies were re-stained to demonstrate acid fast bacillus. This case demonstrates, for the first time, the presence of *Mycobacterium avium* in skin, gut and muscle in an immunosuppressed patient and highlights the need for appropriate investigation. We would recommend a high level of suspicion of mycobacterial infection if granulomatous inflammation is seen in the immunosuppressed.

References

- 1 Lee WJ, Kang SM, Sung H, *et al*. Non-tuberculous mycobacterial infections of the skin: a retrospective study of 29 cases. *J Dermatol* 2010;37:965-72

- 2 Chen HY, Chen CY, Huang CT, *et al.* Skin and soft-tissue infection caused by non-tuberculous mycobacteria in Taiwan, 1997–2008. *Epidemiol Infect* 2011;**139**:121–9
- 3 Shih JY, Hsueh PR, Chang YL, Lin SF, Teng LJ, Luh KT. Pyomyositis due to *Mycobacterium haemophilum* in a patient with polymyositis and long-term steroid use. *Clin Infect Dis* 1998;**26**:505–7
- 4 Lawn SD, Bicanic TA, Macallan DC. Pyomyositis and cutaneous abscesses due to *Mycobacterium avium*: an immune reconstitution manifestation in a patient with AIDS. *Clin Infect Dis* 2004;**38**:461–3
- 5 Leung YY, Choi KW, Ho KM, Kun EW. Disseminated cutaneous infection with *Mycobacterium chelonae* mimicking panniculitis in a patient with dermatomyositis. *Hong Kong Med J* 2005;**11**:515–19
- 6 Lee HT, Su WJ, Chou TY, Chen WS, Su KY, Huang DF. *Mycobacterium avium* complex-induced pleurisy in a patient with amyopathic dermatomyositis and interstitial lung disease after prolonged immunosuppressive therapy. *J Clin Rheumatol* 2009;**15**:193–4
- 7 Nuñez JM, Monteagudo I, López-Longo FJ, Cobeta JC, Rivera J. Disseminated *Mycobacterium avium*-intracellulare infection in a patient with polymyositis. *Arthritis Rheum* 1989;**32**:934–6
- 8 Airio A, Kauppi M, Kautiainen H, Hakala M, Kinnula V. High association of mycobacterial infections with polymyositis in a non-endemic country for tuberculosis. *Ann Rheum Dis* 2007;**66**:1404–5
- 9 Chen IJ, Tsai WP, Wu YJ, *et al.* Infections in polymyositis and dermatomyositis: analysis of 192 cases. *Rheumatology (Oxford)* 2010;**49**:2429–37
- 10 Marie I, Ménard JF, Hachulla E, *et al.* Infectious complications in polymyositis and dermatomyositis: a series of 279 patients. *Semin Arthritis Rheum* 2011;**41**:48–60

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