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CASE REPORT

Dermatomyositis with concurrent hepatitis B and schistosomiasis infection

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Introduction

Dermatomyositis is an idiopathic inflammatory myopathy characterised by progressive, symmetrical proximal muscle weakness and cutaneous changes (Kee et al, 2009). Dermatomyositis causes a complement-mediated microangiopathy in the perifascicular regions of proximal muscles, resulting in capillary destruction, inflammatory cell stress and hypoperfusion. The aetiopathogenesis of this condition remains poorly understood, but is presumed to be autoimmune, with the majority of patients responding to steroid treatment (Stübgen, 2014).

Several triggers have been proposed as possible causes of dermatomyositis. Existing literature describes parasitic causes of rheumatological disease, including reactive inflammatory myositis triggered by schistosomal infection (McGill, 2003). A case of dermatomyositis coexisting with hepatitis B, in the context of liver hepatocellular carcinoma, has been reported (Kee et al, 2009).

Case report

A 54-year-old Afro-Caribbean man presented with a 5-week history of 12 kg weight loss, shortness of breath on exertion, night sweats, fever, lethargy and bilateral leg pain with weakness. Past medical history included hypercholesterolaemia, hypertension and previous recurrent malaria, managed with regular valsartan, amlodipine and spironolactone. He was born and now lived in the UK, although he lived in Nigeria from the ages of 10 to 26 years.

On examination, he had a heliotrope rash in the periorbital region, cutaneous rash over the shoulders, finger clubbing and ragged nailfold cuticles. Neurological examination demonstrated a bilateral weakness of shoulder abduction and hip flexion (3/5 on the MRC scale) and truncal weakness. Sensation, coordination and gait were normal, and his cranial nerves were intact. Cardiovascular, respiratory, abdominal and lymph node examinations were normal. There were no other obvious vasculitic features or evidence of peripheral synovitis.

Initial investigations showed a normocytic anaemia (haemoglobin 119 g/litre, normal range 130–170 g/litre) and normal mean cell volume 87.5 fl (normal range 83–101 fl). C-reactive protein level (89 mg/litre, normal range <5 mg/litre) and erythrocyte sedimentation rate (60 mm/hour, normal range 1–20 mm/hour) were both raised. Troponin T level 1099 ng/litre (normal range 0–14 ng/litre) and creatine kinase level (10 733 units/litre, normal range 40–320 units/litre) were markedly elevated. Autoimmune serology revealed positive anti-Jo-1 antibodies but all other auto-antibodies were negative. Infectious disease screening revealed acute or acute-on-chronic hepatitis B infection (positive hepatitis B surface antigen, core antigen and e antibody). Schistosomiasis antibody was detected on enzyme-linked immunosorbent assay (ELISA). Blood cultures for microscopy, culture and sensitivity, HIV and tuberculosis serology were negative. Urine dipstick confirmed myoglobinuria.

A cardiac transthoracic echocardiogram and cardiac magnetic resonance imaging were both normal, and serum troponin I level was within normal range (25.2 ng/litre, normal range <34.2 ng/litre). Computed tomography pulmonary angiogram showed calcified granulomas and bilateral ground glass opacification. Ultrasound of the liver showed four hyperechoic foci in the right lobe suggestive of granulomas and hepatomegaly (190 mm at the mid-clavicular line). In the context of new anti-Jo-1-positive dermatomyositis, a malignant trigger was excluded with computed tomography of the abdomen and pelvis and whole-body fluorodeoxyglucose-positron emission tomography. Magnetic resonance imaging of the left hip showed myositis, with skeletal muscle biopsy confirming fibre necrosis and myophagocytosis. Immunohistochemical staining showed frequent CD68 positive macrophages with very few scattered T lymphocytes.

Case report (continued)

He was treated with pulsed methylprednisolone (for active myositis), tenofovir (in view of positive hepatitis B serology), praziquantel (for schistosomiasis) and co-amoxiclav (for possible superimposed bacterial infection). With treatment, he showed marked clinical improvement of symptoms and biochemical parameters. He was discharged home on methotrexate and a weaning regimen of oral prednisolone.

At 2 months follow up, he remained well without muscular or cutaneous symptoms. He continued to take prednisolone 35 mg once daily as further dose reductions triggered recurrence of febrile illness, and he remained on methotrexate and tenofovir. His other regular medications included anti-hypertensive agents, oral iron and folic acid supplementation. His hepatitis B virus DNA load was decreasing since commencing tenofovir, with Fibroscan pending to assess for fibrosis.

This article describes the clinical presentation and management of a 54-year old male patient who was diagnosed with polymyositis with concurrent acute-on-chronic hepatitis B and schistosomal infections. To the authors' knowledge this is the first case report of biopsy-confirmed dermatomyositis in a patient with active schistosomiasis and hepatitis B. This case posed a diagnostic challenge, given its non-specific initial presentation and the significant overlap in clinical symptoms between the three disease processes identified. Furthermore, it presented a potentially complex therapeutic challenge, as the three separate disease processes required active immunosuppressive treatment in a patient with risk of reactivation of infection and reduced functional reserve.

Discussion

Several potential immunological triggers for dermatomyositis have been proposed, including infections and malignancy (Hill et al, 2001; Bax et al, 2021). The literature supports an association between parasitic infection and dermatomyositis, as described in this case. However, there is no strong evidence as to whether patients develop an immunological response to a parasite that triggers dermatomyositis (as per McGill, 2003), or whether patients with subclinical polymyositis and deranged immune responses are susceptible to parasitic infections (as per Behan et al, 1983). McGill (2003) emphasised the importance of screening for chronic low-grade symptoms as well as pulmonary and cutaneous extra-articular features of idiopathic inflammatory myopathy in patients with joint disease and a history of travel to tropical or subtropical regions.

With regard to hepatitis B, Pittsley et al (1978) described a case of hepatitis B virus infection causing clinical features of dermatomyositis and resolving with prednisolone as a viral prodrome mimicking dermatomyositis. More recently, Kee et al (2009) described a case of confirmed dermatomyositis in the context of hepatitis B virus and hepatocellular carcinoma. While dermatomyositis in the presence of malignancy is well-documented (Hill et al, 2001), there remains no association between hepatitis B virus and dermatomyositis in the absence of malignancy in the literature. The risk of hepatitis B reactivation in patients treated with immunosuppressive therapy should also be considered.

In this case, the patient's ongoing low grade fevers despite treatment were of continued concern to the clinical team. It was unclear whether the fevers were caused by his schistosomiasis, dermatomyositis or acute on chronic hepatitis B virus infection. Similarly, this patient's acute shortness of breath could represent a bronchospasm of Katayama fever (Doherty et al, 1996), or simply a decompensation of the granulomatous disease and ground-glass opacification secondary to dermatomyositis identified on his computed tomography pulmonary angiogram. Ultimately, both symptoms resolved with steroids and anti-parasitic agents.

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Learning points

- Dermatomyositis/anti-Jo-1 positive anti-synthetase syndrome is part of the idiopathic inflammatory myopathies, a group of rare diseases related to dermatomyositis and polymyositis. Clinically, it can demonstrate cutaneous, lung and cardiac involvement.
- Screening for common infections in patients presenting with dermatomyositis should be considered in those from high-risk areas, and screening for viral hepatitis is essential before starting immunosuppressive therapy.
- Early diagnosis can be difficult but correlates with better clinical outcomes and long-term prognosis, so a high index of suspicion should lead clinicians to arrange appropriate investigations in a timely fashion to ensure accurate diagnosis.
- Elevated levels of cardiac troponin T, but not troponin I, are associated with elevated levels of creatine kinase in idiopathic inflammatory myopathy and suggestive of a skeletal muscle rather than a cardiac cause. Differentiating cardiac involvement in myopathy is essential by obtaining a troponin I serum level, and considering further cardiac investigations such as echocardiogram and cardiac magnetic resonance imaging.

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