CASE REPORT

Infective endocarditis initially presenting with a dermatomyositis-like syndrome

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SUMMARY

Infective endocarditis (IE) may present with rheumatological manifestations such as myalgias, arthralgias, arthritis and back pain. However, muscle inflammation is rare. We present a case of a 68-year-old Hispanic man who presented with 1-month history of tiredness, weight loss, fever, myalgias, muscle weakness and dysphagia to solid food. On physical examination he had severe weakness in the proximal upper and lower extremities, and erythematous eruption involving the upper eyelids, neck and metacarpophalangeal joints. Creatine kinase levels were markedly elevated at 15 809 U/L. MRI of the right thigh revealed intermuscular and intramuscular oedema. Muscle biopsy showed acute necrotising suppurative perimyositis. Blood cultures were positive for methicillin-resistant *Staphylococcus aureus*. A transoesophageal echocardiogram revealed vegetations in the pulmonic valve. All clinical manifestations were resolved completely with broad-spectrum antibiotics. This case suggests that IE should be considered in the differential diagnosis of a patient presenting with inflammatory myopathy.

BACKGROUND

Infective endocarditis (IE) is an infectious disease of the endocardium which leads to heart valve vegetations and destruction.¹ There are multiple risk factors for developing IE, including dental and surgical procedures that predispose to bacteraemia. Symptoms may be subtle and non-specific. IE may present with rheumatological manifestations in 25–44% of patients.^{2–7} The most common musculo-skeletal manifestations are myalgias, arthralgias, arthritis and back pain. However, muscle inflammation is rare. We present a case of a Hispanic man who presented with a dermatomyositis-like syndrome as the initial presentation of IE.

CASE PRESENTATION

A 68-year-old Puerto Rican man with hypertension and benign prostatic hyperplasia was presented in September 2010 with 1-month history of tiredness, unintentional weight loss of 10 pounds, fever, myalgias, proximal muscle weakness, dysphagia to solid food and facial and neck rash. He was sexually active with a single partner. He did not smoke or use illicit drugs. There was no history of alcohol abuse, blood transfusions or trauma.

Physical examination indicated weakness in the proximal upper (2/5 strength) and proximal lower (3/5 strength) extremities, and an erythematous rash involving the upper eyelids, neck and the

metacarpophalangeal joints. He did not have a petechial rash, subungual (splinter) haemorrhages, Osler nodes or Janeway lesions. The cardiopulmonary examination was normal. Deep tendon reflexes in the upper and lower extremities were normal. The remainder of the physical examination was unremarkable.

INVESTIGATIONS

The laboratory evaluation revealed a creatine kinase (CK) of 15 809 U/L (normal range 15–105 U/L), aspartate aminotransferase of 519 IU/L (normal range 10–59 U/L), alanine aminotransferase of 167 IU/L (normal range 13–40 U/L) and lactate dehydrogenase of 113 IU/L (normal range 110–210 U/L). The blood white cell count (WCC) was 10 300/mm³, the haemoglobin was 12.9 g/dL and the platelet count was 402 000/mm³. The erythrocyte sedimentation rate was 28 mm/h. Urinalysis showed 30–35 red blood cells (RBC)/high-power field (hpf), 15–20 WCC/hpf, 0–2 granular casts/hpf, few bacteria, moderate leucocyte esterase and 1+ protein. The level of prostate-specific antigen was elevated at 21 ng/mL. Thyroid-stimulating hormone level was normal. Antinuclear antibodies (ANA) were positive at 1:320, with a homogeneous pattern. Anti-dsDNA, anti-Smith, anti-RNP, anti-SSA, anti-SSB, anti-neutrophil cytoplasmic, anti-MI2, anti-Jo-1 and anti-SRP antibodies were negative. C3 and C4 complement levels were normal. Cryoglobulins were not detected. HIV, hepatitis B and hepatitis C virus tests were non-reactive. The Venereal Disease Research Laboratory test for syphilis was non-reactive. Blood culture was negative. The chest radiograph and CT scan were normal.

DIFFERENTIAL DIAGNOSIS

The clinical presentation and laboratory findings were highly suggestive of dermatomyositis. Other potential aetiologies including viral illnesses, drug-induced myopathies, endocrine disorders, electrolyte disturbances, neurological disorders, malignancy-related myopathies or other connective tissue disorders were excluded on clinical grounds.

TREATMENT

The patient was treated with 60 mg methylprednisolone every 12 h for two consecutive days. He had a rapid response to corticosteroids, as demonstrated by improved muscle strength in the proximal upper extremities (4/5) and the proximal lower extremities (4/5). He was eventually discharged with 30 mg prednisone twice daily and

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500 mg ciprofloxacin twice daily. The latter was for a urinary tract infection. On discharge the WCC was 11 400/mm³ and CK levels were 13 538 U/L.

Electromyography and nerve conduction tests and muscle biopsy were scheduled after discharge but the patient returned to the hospital 2.5 weeks later due to worsening of proximal muscular weakness and dysphagia, and pain in the right hand and in the right lower extremity. Physical examination indicated that muscular strength in the proximal upper extremities was 1/5. The muscular strength in the proximal lower extremities was 0/5. In addition, he had tenderness in the posterior aspect of his right thigh. Deep tendon reflexes in the upper and lower extremities were decreased (1/4). Urinalysis showed 10–15 RBC/hpf, greater than 50 WCC/hpf, and many bacteria. The medical treatment at that time included 750 mg intravenous levofloxacin daily, 60 mg intravenous methylprednisolone twice daily and 2 g/kg immunoglobulin (IVIG) for 5 days.

On the third day of corticosteroid treatment, he developed a right-hand abscess and swelling on the right posterior thigh, which extended to the right buttock. His WCC increased from 11 400 to 21 700/mm³, and CK decreased from 13 538 to 1811 U/L (normal range 15–105 U/L). Blood, urine and right-hand abscess cultures were positive for methicillin-resistant *Staphylococcus aureus*. MRI of the right thigh revealed oedema with fluid-sensitive sequences between intermuscular and intramuscular fasciae extending through the anterior, posterior and medial compartments of the thigh. These findings were consistent with diffuse myositis of the thigh and associated areas of myonecrosis (figure 1). A biopsy of the right quadriceps muscle showed features of acute necrotising suppurative perimyositis (figure 2). Further evaluation included a CT scan, which showed multiple, bilateral pulmonary nodules. A transoesophageal echocardiogram revealed small echogenic densities (<10 mm) in the pulmonic valve. The left ventricular function was preserved. No other abnormalities were found. A venous Doppler of the lower extremities was negative.

The patient was diagnosed with IE. He had no history of intravenous drug use, congenital or structural heart disease, or recent pulmonary, cardiac, gastrointestinal or genitourinary invasive procedures or surgeries. Antibiotics were changed to vancomycin and meropenem. After 3 days of broad-spectrum antibiotic treatment, MRI showed less subcutaneous intermuscular and intramuscular oedema and reduced the size of necrotic

Figure 2 Tissue section stained with H&E (400x) showing preserved striate muscle fibres with extensive infiltration of neutrophils and necrotic debris.

collections (figure 3). The corticosteroid therapy was rapidly tapered down and discontinued 2 weeks later.

OUTCOME AND FOLLOW-UP

The patient showed notable improvement of symptoms by day 42 with intravenous antibiotics, as demonstrated by the increased strength in the proximal upper (5/5) and lower (5/5) extremities and normalisation of the CK level (131 U/L). Systemic corticosteroids were rapidly tapered and discontinued a few weeks later. The patient was asymptomatic at a follow-up visit 2 months after discharge with an unremarkable physical examination and no signs of ongoing infection or muscular weakness. Two years later the patient has remained clinically stable.

DISCUSSION

Rheumatic manifestations may occur in patients with systemic infections such as IE. Our patient presented with clinical features of dermatomyositis including proximal muscle weakness, dysphagia, skin lesions mimicking heliotrope rash and Gottron's sign, elevated muscle enzymes and positive ANA. His symptoms became worse after systemic corticosteroids and IVIG. Further

Figure 1 MRI showing intermuscular and intramuscular oedema with high signal intensities in the fluid-sensitive sequences extending through the anterior, posterior and medial compartments of the thigh.

Figure 3 MRI of the thigh done after 3 days of broad-spectrum antibiotics showing a partial resolution of subcutaneous intermuscular and intramuscular oedema and a decreased size of necrotic collections.

investigation established the diagnosis of IE complicated with myonecrosis.

The overall prevalence of musculoskeletal manifestations in patients with IE varies between 25% and 44%.^{2–7} Myalgias, arthralgias, arthritis and back pain are the most common musculoskeletal symptoms. Other manifestations include sacroiliitis, tendonitis and bursitis. Our patient presented with myalgias but also with severe proximal muscle weakness which is very unusual in IE. This clinical presentation together with cutaneous lesions and elevation of muscle enzymes prompted the initial diagnosis of dermatomyositis instead of IE.

Patients with IE may develop clinical features that mimic rheumatic autoimmune diseases.⁸ In the past, non-cardiac manifestations of IE were thought to be caused solely by septic emboli or metastatic abscesses. However, several studies have shown that peripheral manifestations of IE are linked to immune complex disease.^{9–12} Autoimmune phenomena occur as the result of high levels of agglutinating, complement-fixing bactericidal antibodies and cryoglobulins, which might promote the formation of immune complexes. Some manifestations of such phenomena in IE include glomerulonephritis and cutaneous manifestations such as Osler nodes and subungual haemorrhages.¹² Immune complex disease appears to occur more frequently in IE than in other bacteraemia disorders as Bayer *et al* showed that 90% of patients with IE have detectable levels of circulating immune complexes compared to 50% of septic patients without IE.⁹ Interestingly, patients with right-sided IE have higher levels of immune complexes than those with right-sided IE.¹⁰ The successful treatment of IE results in decreased levels of immune complexes and autoantibodies, and consequently, in the resolution of peripheral complications.¹¹ Our patient had right-sided IE and dermatomyositis-like manifestations that resolved completely after antibiotic therapy. However, it is difficult to assess if muscle manifestations (proximal muscle weakness and dysphagia) in our case were caused by infectious myositis or by immune complex disease. MRI and muscle biopsy were performed in the right thigh but not in the other affected limbs. Although levels of circulating immune complexes were not measured in our patient, autoantibodies were detected. Our patient had high titres of antinuclear antibodies; titres as high as 1:640 have been reported for patients with IE.¹²

On the other hand, patients with polymyositis/dermatomyositis have a greater risk for infection, including IE. In a retrospective study of 192 patients with inflammatory muscle disease, 53 (27.6%) had major infections.¹³ Aspiration pneumonia followed by opportunistic infections were the most common and major infections. Independent predictors of major infection were age >45 years at the onset of inflammatory muscle disease, the presence of arthritis/arthralgia, coexistent interstitial lung disease and current use of azathioprine or intravenous immunoglobulins. In our case, however, true dermatomyositis was very unlikely for several reasons. First, the patient did not improve with corticosteroid therapy. Second, clinical manifestations completely resolved with antibiotic therapy. Third, the patient did not develop recurrent manifestations of inflammatory muscle disease for 2 years despite not being treated with immunosuppressive drugs; this would be very unusual for a chronic autoimmune disease such as dermatomyositis. Finally, the muscle biopsy showed pyomyositis, but no histopathological findings of dermatomyositis.

In summary, we report a case of a Hispanic man in whom clinical features of dermatomyositis were the presenting manifestations of IE. The presence of autoimmune phenomena in IE is possible. Thus, clinicians should be aware of autoimmune manifestations in IE as well as features that mimic rheumatic disorders such as inflammatory muscle disease. An accurate assessment of the clinical manifestations and diagnostic work-up are highly recommended in order to avoid delay in treatment and to provide adequate management.

Learning points

- ▶ Infective endocarditis (IE) may present with rheumatic manifestations such as myalgias, arthralgias, arthritis and back pain.
- ▶ Muscle inflammation is very uncommon.
- ▶ We report a 68-year-old man with IE who initially presented with proximal muscle weakness, dysphagia, elevation of muscle enzymes and skin lesions mimicking dermatomyositis.
- ▶ IE should be considered in the differential diagnosis of a patient presenting with inflammatory myopathy.

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