CASE REPORT

Red fist and muscle weakness with a rare complication

Iris van Groenigen,¹ Joyce Arnoldus,² Roos Perenboom,³ Alexandre Voskuyl³

¹VUmc, Amsterdam, The Netherlands

²Department of Internal Medicine, ZMC, Zaandam, The Netherlands

³Internal Medicine, VUmc, Amsterdam, The Netherlands

Correspondence to

Iris van Groenigen, i.vangroenigen@vumc.nl

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SUMMARY

A 64-year-old man was referred to our hospital, for a second opinion, with fever, skin lesions and general muscle pain. He has been treated in another hospital with antibiotics on suspicion of erysipelas. A week later skin lesions developed on the metacarpophalangeal and proximal carpophalangeal joints of the hands and nose. His mobility was impaired due to muscle pain and muscle weakness. He also showed proximal muscle atrophy and most importantly a typical heliotrope rash in the eyes. Based on these clinical observations, the most likely diagnosis was dermatomyositis. The diagnosis was confirmed by the presence of increased serum creatine kinase levels and abnormalities in skin and muscle biopsy. Prednisone (70 mg/kg) was initiated, but after 19 days the patient developed a *Pneumocystis jiroveci* pneumonia. He died of respiratory failure a few days later.

BACKGROUND

Dermatomyositis is a rare inflammatory myopathy with skin manifestations. Men and women of all ages may be affected. Symmetric and proximal muscle weakness as well as characteristic skin findings as heliotrope rash, shawl's sign and Gottron's plaques are typical clinical features (figures 1 and 2). Systemic symptoms such as fatigue, fever, weight loss or non-erosive inflammatory polyarthritis may develop, especially as the disease progresses. Interstitial lung disease develops in at least 10% of the cases and can rapidly lead to respiratory failure and even death. Furthermore, patients with dermatomyositis are at risk for malignancies, in particular adenocarcinomas of cervix, ovaries, lung, pancreas, bladder and stomach. The risk of developing cancer in patients diagnosed with dermatomyositis

Figure 2 The patient with heliotrope rash.

decreases with time from its diagnosis. Screening for malignancies in newly diagnosed dermatomyositis is advocated.^{1 2}

The treatment of choice is glucocorticoids and, if needed, glucocorticoid-sparing drugs such as azathioprine and methotrexate.³

Only a few cases have been published about patients with dermatomyositis developing *Pneumocystis jiroveci* pneumonia (PJP).^{4–11} PJP in non-HIV patients has a more acute and fatal outcome compared with HIV-infected patients.¹² In patients without HIV infection, the use of glucocorticoids, especially with other immunosuppressive agents, is one of the main risk factors for developing PJP. We describe a patient with dermatomyositis developing a fatal PJP. This case illustrates the importance considering the diagnosis of dermatomyositis as well as PJP. It also shows the association between lymphocytopenia in patients with dermatomyositis and the risk of developing PJP.

CASE PRESENTATION

In July 2010, a 64-year-old man was referred to our hospital, for a second opinion, with fever, skin lesions and general muscle pain. Six weeks ago, he visited another hospital with submandibular erythema and high fever and was treated with antibiotics on suspicion of erysipelas. A week later skin lesions developed on the metacarpophalangeal (MCP) and proximal carpophalangeal (PIP) joints of the hands, nose and eyes. His mobility was impaired due to muscle pain and muscle weakness. Since the patient was a pigeon breeder, an infectious cause was suspected first. After using antibiotics for 2 weeks, his condition deteriorated and the symptoms progressed. On examination at our hospital, the patient had severe proximal muscle

Figure 1 The patient with Gottron's plaques.

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weakness and could hardly walk. He also showed proximal muscle atrophy and most importantly a typical heliotrope rash in the eyes and Gottron's plaques on the MCP and PIP joints (figures 1 and 2). The patient's history and the physical examination did not reveal any signs suggestive of malignancy.

INVESTIGATIONS

On the basis of these clinical observations, the most likely diagnosis was dermatomyositis. The diagnosis was confirmed by the presence of increased serum creatine kinase levels (1880 IU/L) and abnormalities shown in the skin biopsy (chronic perivascular and interstitial dermatitis with vacuolar changes, oedema and mucine depositions) and in the muscle biopsy (lymphocytic infiltrates in muscle). Immunoserology showed antinuclear antibodies with antibodies against SS-A but not against Jo-1. Interestingly, he also showed lymphocyte counts of $0.34 \times 10^9/L$.

CT scan of the body revealed no signs of malignancy.

TREATMENT

Prednisone (70 mg/day) was initiated and 11 days later the patient started walking without any help and his creatine kinase had decreased to 920 IU/L.

OUTCOME AND FOLLOW-UP

Nineteen days after treatment initiation, he presented with dyspnoea and productive coughing. Chest X-ray showed bilateral interstitial infiltrates, suggesting either interstitial lung disease or a PJP. High-dose trimethoprim-sulfamethoxazole was immediately started. Twenty-four hours later, the patient became respiratory insufficient and was in need of mechanical ventilation. At this point, he was treated with high-dose dexamethason, high-dose trimethoprim-sulfamethoxazole, broad spectrum antibiotics and inotropics. Nevertheless he died of pulmonary failure 2 days later. Bronchoalveolar lavage showed a *P jiroveci*. Autopsy suggested that respiratory insufficiency caused by PJP in a patient with dermatomyositis to be the cause of death. Finally, no signs of malignancy were observed at autopsy.

DISCUSSION

The diagnosis of dermatomyositis in this case was initially difficult because of the suspicion of an infectious focus in this pigeon breeder. At presentation in our hospital, however, the diagnosis of dermatomyositis was made and symptoms rapidly improved after initiating prednisone. Dermatomyositis can be a challenging diagnosis since it is a rare disease and most clinicians do not see it in everyday practice. Therefore, dermatomyositis should always be considered in the presence of heliotrope rash and/or Gottron's plaques and muscle weakness.

Furthermore, after initiating prednisone our patient developed a PJP. Although some cases have been described, it is quite unusual in daily practice to diagnose PJP in patients on glucocorticoids alone. In rheumatic disease 1–2% will develop PJP in the majority of cases in combination with immunosuppressive drugs.⁵ Interestingly the risk for a patient with dermatomyositis to develop PJP might be higher compared with other rheumatological diseases, up to 40%.^{5 13} The cause of such a high risk is unknown, but lymphocytopenia may be a potential risk factor for PJP in patients with dermatomyositis. Viguier *et al* determined lymphocyte counts in patients with dermatomyositis before treatment and up to 12 months after initiating treatment. It was shown that 62% of the patients had lymphocytopenia ($<1 \times 10^9/L$) at diagnosis. Twelve only of 47 patients developed opportunistic infections during the first year following the onset of therapy, of which five cases were of PJP. Patients who

developed all opportunistic infections with significantly lower lymphocyte count at diagnosis were compared with patients who did not develop opportunistic infections. The authors suggest a correlation between the lymphocyte counts $<0.8 \times 10^9/L$ and the development of PJP in patients with dermatomyositis.⁸ In patients with dermatomyositis combined with lymphocytopenia at diagnosis, the estimated risk of PJP might be as high as 40% with a very high mortality rate, and therefore PJP prophylaxis should strongly be considered in this patient group.

Learning points

This case demonstrates that in every patient with heliotrope rash in the eyes and Gottron's plaques on hand joints in combination with muscle weakness, the diagnosis of dermatomyositis should be considered. Furthermore, it underlines the risk of fatal outcome due to *Pneumocystis jiroveci* pneumonia (PJP) in a patient with dermatomyositis and lymphocytopenia being treated with glucocorticoids only. PJP prophylaxis should be seriously considered in patients with dermatomyositis and lymphocytopenia. Further studies are warranted for PJP prophylaxis in this patient population to better define patients at risk for developing PJP.

Competing interests None.

Patient consent Obtained.

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