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Skin rash in a jaundiced patient

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A 62-year-old woman presented with a violaceous, pruritic skin rash affecting her face, central chest, forearms and thighs (figure 1). She had lost 12.7 kg over the last two months. She also complained of difficulty in combing her hair, and had mild dysphagia to solid food.

On examination, in addition to the rash, she was found to be jaundiced, with proximal muscle tenderness and weakness. There were no palpable organs or lymph node enlargement.

Laboratory findings were as follows: haemoglobin 11.0 g/dl, packed-cell volume 0.31, white blood cell count $13.8 \times 10^9/l$, platelets $149 \times 10^9/l$, erythrocyte sedimentation rate 72 mm/h, total bilirubin 107 $\mu\text{mol/l}$, alanine transaminase 70 IU/l, aspartate transaminase 67 IU/l, lactate dehydrogenase 420 IU/l. Abdominal computed tomography (CT) scan is shown in figure 2.

Figure 1

Figure 2

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Questions

- 1 What is the skin rash due to?
- 2 What other investigations should be performed?
- 3 What abnormality is shown on the abdominal CT scan?
- 4 What is the probable cause of her dysphagia?
- 5 What is the underlying diagnosis?

Answers

QUESTION 1

Dermatomyositis. The rash usually is a helio-trope rash (oedematous violaceous changes in the peri-orbital areas). Similar changes may be seen on the dorsal aspects of the knees, elbows, knuckles, neck, central chest, or the erythema may be more generalised. Gottron's papules (erythematous dermal papules overlying the pharyngeal joints) are pathognomonic of dermatomyositis. Other changes include peri-ungual telangiectasia, cuticular overgrowth, nail-fold infarct, poikiloderma (patchy discolouration of the skin), and erythematous scaly pruritic photodistributed plaques.

QUESTION 2

Muscle enzymes (ie, creatine kinase, lactic dehydrogenase, aldolase, alanine transaminase and myoglobin) should be measured, electromyography performed, a skin and/or muscle biopsy carried out, and antinuclear antibodies, specific antibodies (anti-Jo-1) measured.

QUESTION 3

CT scan of the abdomen revealed enlargement of the head of pancreas with porta-hepatis and para-aortic lymph node enlargement, and intrahepatic biliary dilatation.

QUESTION 4

Almost all of these patients have high dysphagia with nasal regurgitation. Weakness of the pharyngeal and upper oesophageal muscles as a part of paraneoplastic neurological syndromes including Lambert-Eaton myasthenic syndromes, myasthenia gravis, polymyositis-dermatomyositis. Dysphagia could also be due to candida, herpes simplex, or cytomegalovirus infection.

QUESTION 5

The underlying diagnosis was adenocarcinoma of the head of pancreas.

Discussion

Since an association between polymyositis and cancer was first proposed in 1916,¹ its validity has been in dispute. Dermatomyositis is an inflammatory proximal myopathy that is part of a group of disorders known as idiopathic inflammatory myopathies. Included in this designation are polymyositis, an entity which also bears a relationship with cancer, and inclusion body myositis, a disorder not linked to cancer.² Almost any type of malignancy (box 1) may be found in patients suffering from dermatomyositis.³ Paraneoplastic neurological syndromes are now thought to result in part from the cross-reaction of antitumour antio-

Malignancy associated with dermatomyositis/polymyositis

- lymphoma
- breast carcinoma
- nasopharyngeal carcinoma
- pancreatic carcinoma
- ovarian carcinoma
- colorectal carcinoma
- melanoma
- oat cell carcinoma
- stomach carcinoma
- renal cell carcinoma

Box 1

Summary points

- the risk of carcinoma is increased in patients with dermatomyositis/polymyositis
- in patients with dermatomyositis there is also a high rate of mortality from cancer
- paraneoplastic neurological syndromes result in part from the cross-reaction of antitumour antibodies with antigens present in neural tissue
- dermatomyositis may occur before, after, or concurrent with cancer
- eradication of the tumour may ameliorate the symptoms of dermatomyositis while worsening may herald recurrence

Box 2

dies with antigens also present in neural tissue.⁴

Dermatomyositis may occur before, after, or concurrent with cancer. Eradication of the tumour may result in improvement in the signs and symptoms of dermatomyositis. Worsening may herald a recurrence. About 25% of patients have an associated malignancy.⁵ Symmetric proximal muscle weakness with or without tenderness typifies the myopathy. Creatine kinase is generally elevated and electromyography has characteristic abnormalities.

Amyopathic dermatomyositis is a disease of unknown origin characterised by the specific skin lesions of dermatomyositis but without clinical or laboratory evidence of myopathy.⁶ It has been suggested that the incidence of cancer is higher in patients with a normal creatine kinase level.⁷

Final diagnosis

Adenocarcinoma of the head of pancreas associated with dermatomyositis.

Keywords: pancreatic carcinoma, dermatomyositis, paraneoplastic syndrome

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