LESSON OF THE MONTH

Dermatomyositis—how far to go!

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Case history

A 73 year old lady was referred with a four week history of proximal muscle weakness, myalgia and an erythematous, oedematous rash affecting her face, upper chest, and back. Systemic enquiry and general examination was otherwise unremarkable. Initial investigations revealed a normal full blood count, erythrocyte sedimentation rate, C reactive protein, urea and electrolytes, liver function test, thyroid function tests, and urine analysis. The creatine kinase (CK) was increased to 3806 (normal range 24–170 U/l). Although Mi-2 antibody testing was not carried out she did have an IgG ANA level of 100. The rest of the autoimmune screen was negative. A needle muscle biopsy showed type 2-fibre atrophy with perivascular inflammatory changes but no direct evidence of myositis.

She was given high dose oral prednisolone (60 mg daily) and received two pulses of intravenous methylprednisolone (1 g) and cyclophosphamide (1 g). Although the CK level responded rapidly to this treatment her clinical response was more gradual. Azathioprine was introduced and the prednisolone dose was gradually reduced to 30 mg daily with plans to reduce the dose further by 5 mg/month. She had a normal chest radiograph, sigmoidoscopy, barium enema, and abdominal ultrasound.

Three months after the onset of her dermatomyositis she was seen urgently with acute thoracolumbar back pain. Radiographs confirmed slight vertebral wedging of D12. A myeloma screen and biochemical bone profile were normal. An isotope bone scan showed increased activity at D12 but no other areas of abnormality. We concluded that the probable explanation of these results, in a post-menopausal lady who had several other risk factors for osteoporosis (obesity, immobility, and high dose corticosteroids), was one of an osteoporotic fracture rather than an underlying malignant process. The dose of prednisolone was reduced from 10 to 7.5 mg daily and cyclical etidronate started. She had experienced nausea since starting azathioprine, this was withdrawn and her nausea settled.

Three months later she was readmitted with a recurrence of her rash and myositis (CK, 666). Systemic enquiry, general examination, and routine investigations were normal. In particular there was neither a history of weight loss (which had actually increased with corticosteroids) nor gastrointestinal symptoms. A repeat

needle muscle biopsy showed type II muscle atrophy. She was given prednisolone 40 mg daily again and methotrexate 10 mg weekly. Once again her CK normalised despite a slow clinical response. On this occasion she developed respiratory muscle involvement and severe cricopharyngeal dysphagia. During endoscopy, for percutaneous enteroscopic gastrostomy feeding, an inoperable poorly differentiated adenocarcinoma was discovered.

Discussion

We have described a patient who presented with probable dermatomyositis and occult gastric cancer. The concern that we had missed the opportunity for an early diagnosis prompted us to re-examine the available literature to assess how far we should investigate the next patient who presents with dermatomyositis.

An association between cancer and dermatomyositis was first suggested in 1916.¹ Although further case reports followed it was not until 1951 that anybody attempted to quantify any association.² A number of further studies variously reported an incidence of malignancy between 7%–34%.^{3–6} In their seminal papers describing criteria for the diagnosis of dermatomyositis and polymyositis, Peter and Bohan reported an incidence of only 8.5%.^{7,8} This figure was at odds with a number of authors who later reported prevalence rates between 30% and 50%.^{9–11} In an attempt to make sense of these discrepant results, a large population based study was set up to provide an accurate estimate of the risk of cancer in patients with dermatomyositis.¹² Among the 392 patients with dermatomyositis, 61 cancers were diagnosed in 59 patients (15%). The authors concluded that this equated to a relative risk of cancer of 2.4 for male and 3.4 for female patients. In 1994 Zantos *et al* confirmed (pooled odds ratio of 4.4) the association in their meta-analysis of published case-control and cohort studies evaluating the association of myositis and malignancy.¹³

A wide variety of different malignancies have been reported in patients with dermatomyositis. Although several authors have highlighted an apparent overrepresentation of gynaecological malignancies,^{14,15} Callen¹⁶ and Zantos¹³ conclude that the distribution approximates to that found in an age matched general population.

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Table 1 Investigations in patients with dermatomyositis

Investigation	Chest radiography	Mammogram	Abdominal ultrasound	Faecal occult blood	Gastroscopy	Barium enema	Cervical smear	Computed tomography chest/abdomen	PSA
Author	No	Yes	Yes	Yes	No	No	Yes	No	No
Snaith ²²	Yes	Yes	Yes (female)	Yes	No	No	No	No	Yes
Callen ¹⁴	Yes	Yes	No	No	No	No	No	No	No
Richardson ²³	Yes	No	Yes	Yes	Yes	Yes	No	Yes	No
Spiera ²⁴	Yes	Yes	Yes	Yes	No	No	No	No	No

The above investigations are in addition to a full history and examination (including breast, rectal, and pelvic examination) and routine tests (which we have interpreted as FBC, LFT, bone profile, and urine analysis). Any abnormality should be followed up independently.

The issue of how far to investigate patients with dermatomyositis will often depend on the presence of symptoms, clinical signs or abnormal routine blood tests. There is little doubt that any symptoms or abnormalities should be followed up appropriately. Our patient illustrates the dilemma when confronted by someone with dermatomyositis who has no localising symptoms, signs or routine investigations. During her first admission sigmoidoscopy, barium enema, chest radiography, and ultrasound examination of her pelvis and abdomen were normal. In the absence of localising signs there did not seem to be any indication to proceed with further investigations. When she was re-admitted she had further routine blood tests and a repeat abdominal ultrasound despite the lack of clinical indications of underlying malignancy. Upper gastrointestinal endoscopy was only performed as a prelude to percutaneous enteroscopic gastrostomy feeding.

Bohan and Peter first questioned the value of an extensive search for occult malignancy.⁷ Several authors have since confirmed that extensive investigations are of little value.^{17–18} Although the logical interpretation of this evidence is to investigate patients on the basis of abnormal symptoms, signs or baseline investigations, the management plans of a number of authors are at odds with this approach (table 1).

Several factors are thought to influence the incidence of coexisting malignancy. Increasing age,¹² female sex, normal CK levels,¹⁹ and refractory or recurrent disease are all thought to increase the likelihood of occult malignancy. In contrast, features of overlap connective tissue disease and the presence of myositis associated antibodies are thought to reduce this risk.⁸

Gastric cancer has an incidence of between 8–20/100 000. Early, potentially curable, disease is usually asymptomatic.²⁰ Populations with high prevalence rates have therefore initiated mass screening programmes²¹ in an attempt to reduce mortality.

In conclusion, we would like to suggest that asymptomatic patients with dermatomyositis have an age specific examination for occult malignancy. The potential benefits of detecting early disease probably justify the inclusion of endoscopy in the age specific screening of patients with dermatomyositis. We would also emphasise that there may be a delay of up to five years before any associated malignancy develops.^{12–14} Consequently a heightened awareness of the possibility of

malignancy should be maintained. Finally, our patient has reminded us of another important lesson; the benefits of bone protection in at risk patients starting long term corticosteroid treatment.

The lessons

- Recurrent or refractory dermatomyositis should prompt a search for occult malignancy.
- Upper gastrointestinal endoscopy should be considered as part of the age specific routine assessment of patients with dermatomyositis.
- An investigation plan in the asymptomatic patient with dermatomyositis might include chest radiographs, faecal occult blood tests, sigmoidoscopy, gastroscopy and in addition prostatic specific antigen in a male and Ca-125, abdominal and pelvic ultrasound, and mammography in a female patient.
- Patients with dermatomyositis who are treated with high dose corticosteroids should receive prophylaxis against osteoporosis.

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Figure 1 Syphilitic dactylitis.
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