(ACHA) levels in patients with various atherosclerotic vascular disorders.[6] In this study ACHA levels of patients with SLE were compared with those of healthy donors.

Sixty eight patients (64 women, four men), aged 39.4 (10.6) (mean (SD)) years, who fulfilled at least four of the diagnostic criteria established by the American Rheumatism Association for SLE[7] and 60 healthy donors (55 women, five men), aged 42.6 (8.12), were enrolled into the study. The SLEDAI (SLE Disease Activity Index) score was used to measure disease activity.[8] Patients were considered to have active lupus if they scored at least 2 on the modified SLEDAI scale (calculated by omitting anti-dsDNA and complement from SLEDAI) and prompt treatment was obviously indicated to control their symptoms.

The level of cholesterol-specific antibodies was measured by a solid phase enzyme immunoassay described earlier.[6] Polystyrene plates (Greiner, Frickenhausen, Germany) were coated with 5 µg/well cholesterol dissolved in 100 µl absolute ethanol and incubated at 4°C for 24 hours. After washing with phosphate buffered saline (PBS) and blocking with 0.1% casein (Reanal, Budapest, Hungary) in PBS, the wells were incubated with 100 µl samples of serum diluted 1:800 in PBS containing 0.1% casein. The binding of ACHA was detected by anti-human horseradish peroxidase conjugated γ-chain-specific rabbit antibodies (DAKO, Glostrup, Denmark); and with *o*-phenylenediamine (Sigma, St Louis, USA) using H_2O_2 as substrate. Optical density was measured at 492 nm (reference at 620 nm), and the mean of duplicates was calculated. Serial dilutions of purified immunoglobulin were used as standards in all experiments. Data obtained as optical density values were expressed in arbitrary units per millilitre (AU/ml), related to the standard curve. Our previous observations,[6] in accordance with those of others,[9] demonstrated the specificity of ACHA to cholesterol. The inter- and intra-assay variations of this method were 18.7% and 9.5%, respectively.

Differences between the parameters measured in controls and patients with SLE, between patients with active and inactive SLE, and those between patients with and without previous vascular events were calculated with the Mann-Whitney-test. The χ^2 test was used to estimate the discriminative power of ACHA between patients with SLE and healthy donors. The correlation of individual parameters with each other and with the SLEDAI score was calculated using Spearman's rank correlation test.

Twelve samples (18%) were classified as having been obtained from patients with active SLE and 56/68 (82%) from patients with inactive disease. Three of the 12 "active" samples were from patients with SLE active in more than one organ system and nine were from patients with disease activity in one organ system only (musculoskeletal (four), central nervous system (two), renal (two), and cutaneous (one)).

The difference between ACHA levels measured in the control group and in patients with SLE was significant (p<0.001) (fig 1). High ACHA levels, in the upper quartile occurred in 29/68 (43%) patients with SLE and 13/60 (22%) controls (p=0.012). ACHA levels (mean (SD)) of patients with active (97 (81) AU/ml) or inactive (64 (38) AU/ml) SLE did not differ significantly (p=0.21) and did not correlate with the SLEDAI score

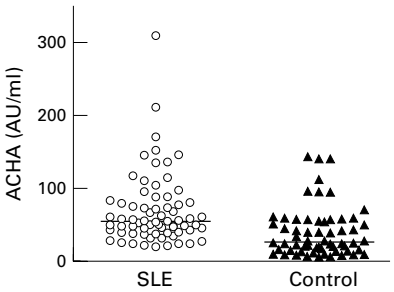

Figure 1 Individual anticholesterol antibody (ACHA) levels (mean (SD)) of patients with systemic lupus erythematosus (SLE) (69 (49) AU/ml) and of healthy donors (38 (34) AU/ml). Horizontal lines show median values.

(r_s=0.22, p=0.066). No correlation was found between the ACHA level and other parameters (such as anti-dsDNA, CH_{50}, C3, and C4, data not shown). No significant differences were found between patients treated with corticosteroids for inactive disease or those with inactive disease and not receiving corticosteroid treatment (p=0.174). The latter group had not received corticosteroids for at least one year before blood samples were obtained. The difference between the ACHA levels of patients who had or had not experienced vascular events was not significant.

The observed increases of ACHA levels may be related to underlying chronic inflammatory disease. A number of conditions such as dyslipoproteinaemia,[1–3] nephrotic syndrome, and changes in cholesterol membrane domains[10] might have elicited the increase of ACHA levels. Corticosteroid treatment might also have stimulated ACHA production, though according to our observations this intervention does not alter ACHA levels significantly. A preventive role against atherosclerosis in patients with SLE has been attributed to raised ACHA levels, though these may also exert an atherogenic effect.[6] Further studies are necessary to clarify the role of ACHA in the changes of lipid metabolism ascertained in patients with SLE.

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Intravenous immunoglobulin for treatment of gastrointestinal haemorrhage in dermatomyositis

Polymyositis (PM) and dermatomyositis (DM) are systemic inflammatory disorders affecting skeletal muscles and other organs, especially the digestive tract.[1–8] Oesophageal motor disturbances are common, occurring in as many as 25–60% of patients with PM/DM.[5] Gastrointestinal disease is less recognised in PM/DM, though it may be responsible for life threatening complications—for example, dramatic haemorrhage, perforation, pseudo-obstruction, pneumatosis cystoides intestinalis, and spontaneous abdominal haematoma[1–4 6–8]. We recently observed a new case, which is of particular interest. The patient who had DM refractory to steroids and both gastrointestinal haemorrhage related to vasculitis and oesophageal impairment due to DM experienced a rapid and complete resolution of all clinical manifestations after intravenous immunoglobulin treatment was started.

An 18 year old man had DM evolving from March 1999. The diagnosis of DM was made by the Bohan and Peter criteria[9 10]: (*a*) symmetrical muscle weakness. Muscle power was gauged for eight proximal muscles (neck flexors, trapezius, deltoid, biceps, psoas, maximus and medius gluteus, and quadriceps) by a modification of the British Medical Research Council Grading system,[5] resulting in a theoretical maximum score of 88 points. Muscle power of the patient was 73 points; (*b*) increased serum muscle enzymes—that is, creatine kinase (CK) 1700 U/l (normal 5–130) and aldolase 7.2 U/l (normal 0.5–3.1); (*c*) myopathic changes on electromyography; (*d*) muscle damage on histological examination; and (*e*) characteristic dermatological manifestations—that is, heliotrope rash, periungual erythema, and poikiloderma.

Autoantibody screen was positive for antinuclear antibodies (ANA) with a value of 1/1000. Investigations, including pulmonary function tests, computed tomography scan of

the lungs, echocardiography, and abdominal ultrasound, were normal. Treatment with prednisone was started at a dose of 1 mg/kg daily. As both the clinical and biochemical status continued to deteriorate gradually, the steroid regimen was increased to a dose of 1.5 mg/kg a day in June 1999.

In July 1999 the patient presented with a two week history of dysphagia and melaena evolving from one day. On admission, his general condition was poor and abdominal palpation was tender. Physical examination also showed cutaneous manifestations of DM and muscle weakness affecting both arms and legs. Muscle power of the patient was 65 points. Laboratory findings were as follows: erythrocyte sedimentation rate 50 mm/1st h, C reactive protein 30 mg/l, haemoglobin 6.6 mmol/l, mean corpuscular volume 90 fl, reticulocytes 150×10^9/l, white blood cell count 10×10^9/l, platelet count 490×10^9/l, CK 3000 U/l, and aldolase 13.5 U/l. Findings of renal and liver tests, total protein, and albumin levels were normal. Autoantibody screening was positive for ANA >1/1000 with a speckled pattern; other tests, particularly for anti-Jo1 antibody, rheumatoid factors, anticardiolipin and antiphospholipid antibodies, lupus-like anticoagulant, antineutrophil cytoplasmic antibodies, and cryoglobulin, were negative. Oesophageal manometry showed decreased peristalsis in the upper third of the oesophageal body and normal pressure in both upper and lower oesophageal sphincters. Gastroscopy demonstrated multiple small ulcerations affecting the stomach and the duodenum, with histology showing vasculitis of the small sized vessels.

A diagnosis of gastrointestinal haemorrhage related to vasculitis and oesophageal impairment due to DM was made. The patient was given intravenous immunoglobulin at a dose of 1 g/kg for two consecutive days monthly for six months. Prednisone was simultaneously decreased gradually to 5 mg every 15 days. The patient had no gastrointestinal haemorrhage recurrence, swallowing disorders and muscle strength improved rapidly, and the dermatological signs cleared.

In November 1999 methotrexate treatment was started at a dose of 30 mg weekly. At one year follow up, the patient remains free of digestive, cutaneous, and muscle symptoms with methotrexate at a dose of 30 mg weekly and 12 mg prednisone daily.

Although oesophageal motor abnormalities predominate in patients with PM/DM and have been extensively described, involvement of the gastrointestinal tract is considered to be less common.[1–8] In a review of 96 patients with DM, Downey *et al* found that only four patients had gastrointestinal manifestations.[2] Our findings confirm that gastrointestinal impairment is a major cause of morbidity in PM/DM, as our patient presented with life threatening gastrointestinal haemorrhage. A diagnosis of gastrointestinal vasculitis related to DM could reasonably be made for our patient because the onset of DM clinical deterioration and gastrointestinal vasculitis was concomitant and the search for other causes of vasculitis (notably systemic vasculitides or other connective tissue disorders) proved negative.

Our report further highlights the importance of recognising gastrointestinal complications at an early stage in PM/DM, resulting in accurate diagnosis and management, and therefore decreasing both morbidity and mortality. The pathological mechanisms of gastrointestinal involvement are still not clearly understood in PM/DM, though it may be related to vasculitis of small sized vessels, leading to ischaemia, haemorrhage, and perforation of the gastrointestinal wall.[2 3 8] Moreover, the present case is original, as our patient with DM and life threatening digestive impairment received intravenous immunoglobulin treatment, which prevented gastrointestinal haemorrhage recurring and produced dramatic and rapid remission of swallowing disorders. Previous authors have also mentioned a favourable outcome with intravenous immunoglobulin treatment in patients with systemic vasculitis—for example, Churg-Strauss vasculitis, microscopic polyangiitis, or systemic lupus erythematosus.[11–14] In this instance, a limitation was the concomitant continuation of steroids during the entire period of intravenous immunoglobulin, making it difficult to be certain that the patient's clinical improvement was only attributable to intravenous immunoglobulin treatment. However, the improvement of all gastrointestinal symptoms may reasonably be related to intravenous immunoglobulin infusions in our patient with DM because the gastrointestinal manifestations deteriorated persistently despite high doses of prednisone as a single treatment. The beneficial effect of the accompanying methotrexate treatment could also be excluded, as this later drug was started at the five month follow up of the patient.

Finally, our findings indicate that intravenous immunoglobulin should be considered the best treatment in both gastrointestinal haemorrhage related to vasculitis and oesophageal dysfunction due to steroid refractory DM, such a treatment offering the advantages of short term efficacy and good tolerance. However, no definite conclusion can be drawn and further controlled trials with a large number of patients with PM/DM are required to establish optimal doses and effective management.

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Sjögren's syndrome: an unusual cause of Bell's palsy

The most common form of facial paralysis is idiopathic—that is, Bell's palsy. Sjögren's syndrome (SS), a chronic inflammatory disorder characterised by lymphocytic infiltration of exocrine glands resulting in the so called "sicca complex", is a rare secondary cause of this self limiting illness. Primary SS includes mostly peripheral, and to a lesser extent cranial, autonomic neuropathy and central nervous system involvement.[1] A patient with unilateral facial palsy, autoimmune hypothyroidism, and Sjögren's syndrome is presented.

A 41 year old woman developed right sided facial numbness, described as "dentist anaesthesia for tooth extraction". One day later she had a reduced sense of taste and right facial weakness. General physical examination was not remarkable. Neurological examination showed anisocoria, peripheral right sided facial paresis, reduced sense of taste on the right half of the tongue, and dysaesthesia in the region of the second segment of the right trigeminal nerve.

Although the erythrocyte sedimentation rate (ESR) was 30 mm/1st h, routine laboratory investigations were normal. Liquor examination, including IgG, IgG index, and oligoclonal or extra bands on electrophoresis, was not abnormal. Screening tests for herpes simplex and varicella virus, syphilis, and borrelia in the serum and in the liquor were negative. Magnetic resonance imaging (MRI) of the brain showed no abnormalities. Nerve conduction studies showed peripheral facial paresis. She recovered spontaneously from her symptoms within several days. Conversely, during follow up the raised ESR persisted.

Review of her medical history uncovered complaints of burning eyes and dry mouth, slight weight gain, and cold intolerance. There was no history of arthralgias or skin lesions. She denied using any drugs previously. A strongly positive anti-extranuclear antigen (ENA)/SS-A antibodies test was shown on further investigation, with negative tests for antinuclear factor/anti-nDNA antibodies/ rheumatoid factor and RNP/SS-B/Sm antibodies. Rose-Bengal staining showed corneal punctate lesions (van Bijsterveld score 6). Lower labial biopsy showed histopathological findings matching the diagnosis of SS with a