between these disorders. We therefore describe a woman with SSC who developed typical lesions of generalized morphoea. A year later, invasive ductal carcinoma of breast was diagnosed. We discuss the rare coexistence of systemic sclerosis and morphoea, and the possible implications of breast cancer.

A 47-yr-old woman from North Wales presented in February 2003 with a 10-month history of Raynaud’s phenomenon, arthralgia and mild heartburn. There was no other relevant past or family history, and no reported exposure to chemicals or toxins. On examination, the significant findings were sclerodactyly, a positive ‘prayer sign’ and abnormal nail fold capillaroscopy with capillary dropout. There was no clinical internal organ involvement; haematology, biochemistry, chest X-ray, pulmonary function tests and 2D echocardiogram results were normal. The ANA was positive at 1:160, in a nucleolar pattern. Limited cutaneous SSC was diagnosed.

In June 2003, she developed progressively enlarging intensely pruritic, hyperpigmented lesions on the back of both knees, the posterior aspect of both thighs and the outer aspect of the upper arms bilaterally. Examination showed extensive, confluent, hyperpigmented, indurated lesions with a violaceous, erythematous border typical of generalized morphoea. The surrounding skin was normal and there was no extension of sclerodactyly or any other clinical development. Methotrexate 15 mg once a week was started.

In June 2004, she reported a lump in the upper outer quadrant of left breast, present for 3 months. The patient had undergone pan-hysterectomy in 1994 and was on hormone replacement therapy (HRT). Histology confirmed an invasive ductal carcinoma of the breast. HRT was discontinued. Left mastectomy with epirubicin for 3 months, then anastrozole 1 mg daily. By October 2004, there was a marked improvement in the morphoea. Induration of the skin and subcutaneous tissue had receded completely with only residual hyperpigmentation. Arthralgia and sclerodactyly remained unchanged. Methotrexate was continued.

Although considered to be rare, coexistence of SSC and LS has been reported in the past. Soma et al. [2] reported that 6.7% of 135 SSC patients at presentation had additional lesions of LS. Mizutani et al. [3] described recurrent morphoea lesions over 6 yr in a patient with SSC. Conversely, Rosenberg et al. [4] reported a positive ANA in 63% of children with LS. The scleroderma specific anticytokeratin antibody was detected in three of 25 patients with LS in another study [5]. Mariçq [6] reported two of 27 LS patients with Raynaud’s and fully established SSC.

SSC, but not LS, has been associated with approximately a two-fold increase in malignancy, the greatest risk being for lung cancer [7]. The results have been non-uniform and inconclusive for breast cancer. As reviewed in [7], a large population-based Australian study reported an insignificant increase whereas a Swedish study found no increase in breast cancer in SSC. Furthermore, a USA study reported no increase in breast or lung cancer but rather a non-significant decrease in risk of both. Although by itself not associated with increased risk of malignancy, LS may occur following radiotherapy for breast or other cancers.

The mechanisms explaining a relationship between SSC and cancer are unknown. A multitude of factors such as exposure to environmental agents, prior genetic damage, immunosuppressive therapy or paraneoplastic phenomenon have been implicated as potential common links between the two conditions [8]. Breast cancer in our patient developed within 2 yr of the onset of scleroderma. She had symptoms of breast cancer before methotrexate therapy was instituted and had never received radiotherapy. It is likely that the 10 yr of HRT was a contributing risk factor. HRT, however, is associated with an increased risk of lobular but not ductal carcinoma [9]. It remains unexplained why the morphoea improved dramatically within months of treating the breast cancer. While this may have been in response to cancer chemotherapy, another possibility was the eradication of the breast tumour per se since the morphoea in this setting was unusual and may have represented a paraneoplastic syndrome.

In summary, this case highlights the interesting association between SSC and LS. Such cases, although rare, might provide important clues to the pathogenesis of scleroderma. Development of breast cancer in such a rare clinical situation points to the possibility of common links between the three conditions.

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as the immunomodulatory drugs typically used for MS will be ineffective for relapse prevention in NMO and severe refractory attacks in NMO usually necessitate rescue plasmapheresis or more effective immunosuppressive therapies [3].

In the management of this suspected case of NMO, the authors made use of investigatory tools such as cerebrospinal fluid analysis and MRI of the brain/spinal cord in diagnosis. However, as indicated by de Seze et al. [2], even when all these investigatory armaments are summoned there is still the pitfall of misdiagnosis and the inability to distinguish the two. Recently, using the technique of dual immunostaining, Lennon et al. [4] have demonstrated that a serum autoantibody marker, NMO-IgG, is highly specific for neuromyelitis optica and can significantly curtail the diagnostic ambiguity.

Perhaps, with the aid of a specific autoantibody marker, the management of this patient might have been more streamlined.

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Devic’s syndrome in systemic lupus erythematosus and probable antiphospholipid syndrome: reply

We thank Drs Chan and Liu for their interest in our case report. We entirely agree that it is fundamental to distinguish between neuromyelitis optica caused by an inflammatory/demyelinating process and a genuine case of multiple sclerosis. In fact, this was our main concern.

Our patient had clear evidence of lupus with multisystem involvement and had several clinical and laboratory signs of severe lupus activity, despite being strongly immunosuppressed. Her clinical presentation was not characteristic of multiple sclerosis: optic neuritis was unilateral; blindness was permanent; evoked potentials were delayed but also had short amplitude; and MRI lesions had no enhancement after gadolinium administration and were not progressive in time and space.

In our opinion, the main diagnostic issue was the differentiation between an inflammatory and/or a thrombotic mechanism. Antiphospholipid syndrome is common in lupus and is frequently associated with central nervous system involvement, namely transverse myelitis [1–3]. Also, abrupt-onset, unilateral and irreversible optic neuritis is more commonly ischaemic than inflammatory/demyelinating [1]. Our patient had transiently positive anti-cardiolipin antibody and presented with stroke, thrombocytopenia and extensive livedo reticularis, suggesting underlying antiphospholipid syndrome. A good response to anticoagulation has been reported in similar cases [2, 4].

Overall, we felt that a demyelinating syndrome as part of her lupus with secondary antiphospholipid syndrome was more probable than classical multiple sclerosis occurring coincidentally in this patient. We agree with Drs Chan and Liu that it would be interesting to study the specific autoantibody marker for neuromyelitis optica (NMO-IgG) [5]—this may well give further help in characterizing these clinically difficult patients.

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Effect of rituximab in refractory SLE: inhibition of Th1?

Sir, The report by Tokunaga et al. [1] clearly demonstrates that rituximab, a chimeric monoclonal antibody specific for human CD20, is highly effective against the life-threatening disease systemic lupus erythematosus (SLE). Furthermore, the authors demonstrate that rituximab not only reduces B-cell numbers and IgG levels, but also down-regulates CD40 and CD80 on B cells of treated patients. Interestingly, all of the five patients they studied had central nervous system (CNS) involvement (two had consciousness disorder and three had sensory disorder).

The balance between T-helper type 1 cells (Th1 cells) and Th2 cells in SLE patients remains controversial. Some reports have suggested that SLE is a disease in which the actions of peripheral