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

focus score of one lymphocyte focus for 4 mm^2 salivary gland tissue. Additionally, thyroid function tests showed a raised thyroid stimulating hormone (11 mU/l), low free thyroxine 4 (13.0 pmol/l) with positive antithyroid microsomal antibodies and negative antithyroglobulin antibodies.

The clinical, serological, and histopathological manifestations fulfilled the European study group criteria for the diagnosis of SS. The patient was treated with artificial tears and thyroxine supplements that returned her thyroid function tests to normal.

Prevalence of neuropathy in patients with SS ranges from 10 to 50%.[2] Polyneuropathy can be the first clinical manifestation of SS and may even precede sicca symptoms in 40% of patients.[3] However, less frequently, cranial neuropathy can occur with a predisposition to involvement of the trigeminal nerve.[4] The vasculitic damage to vaso nervorum documented by pathological studies is associated with a higher incidence of serum anti-SS-A (Ro) antibodies.[5] The association of SS with autoimmune thyroid disease (AITD) is well recognised.[6 7] AITD and SS share similarities in the immunopathology in addition to their genetic linkage to the HLA-DR3/DR4 alleles.[7] Only nine cases of facial nerve involvement associated with SS have been described previously.[1 8–10]

This case illustrates how facial palsy disclosed the primary SS as an underlying systemic disorder. To our knowledge the combination Bell's palsy as presenting feature in a patient with SS, and hypothyroidism secondary to AITD has not been reported hitherto.

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α_1 Antitrypsin phenotypic variability is not associated with ANCA in southern Chinese

α_1 Antitrypsin (α_1AT) is a 52 kDa proteinase encoded by a gene locus Pi on chromosomal segment 14q32.1. It is a natural inhibitor of proteinase 3 (PR3), a neutrophil granular protein and a major autoantigen of antineutrophil cytoplasmic antibody (ANCA). The function of α_1AT is in turn restricted by myeloperoxidase (MPO), another autoantigen of ANCA. The interplay between the enzymes, inhibitors, and the autoantibodies is implicated in the dynamics of the vasculitic process,[1] resulting in a whole spectrum of clinical conditions ranging from systemic granulomatous diseases to kidney limited glomerulonephritis. There have been reports of the correlation of specific α_1AT alleles, notably Pi^Z, with ANCA.[2–4] These were largely studies of white subjects, which may not necessarily be extrapolated to all populations.

α_1AT variant phenotypes may have predisposed to PR3-ANCA, but the same association may not exist for MPO-ANCA. In populations with a low prevalence of α_1AT variant phenotypes, the pattern of ANCA could differ from that in white subjects where such variants prevail. We set out therefore to establish the distribution of α_1AT in patients with the two main forms of ANCA (anti-PR3 positive and anti-MPO positive). Blood samples of patients with vasculitis received at the immunology section of the Department of Pathology, Queen Mary Hospital, Hong Kong, were tested for ANCA by indirect immunofluoresence, followed by enzyme linked immunosorbent assays (ELISA) for anti-PR3 and anti-MPO. α_1AT phenotypes were determined by isoelectric focusing, the results of which were compared with those of healthy Chinese adults.

A total of 157 samples from ANCA+ (either anti-MPO or anti-PR3 positive by ELISA) patients were evaluated, 60 (38%) of which were positive for anti-PR3 and 97 (62%) for anti-MPO by ELISA. All were Chinese patients with a clinical diagnosis of vasculitis. The male to female ratio was 0.76 (0.94 for anti-PR3 positive and 0.67 for anti-MPO positive patients). The mean age of the two groups was 52.4 and 59.4 years, respectively. A total of 103 (66%) were homozygous M, 50 (32%) heterozygous M (for example, M1 M2), and 4 (3%) heterozygous for M and a variant allele. Tables 1 and 2 show the allelic and phenotypic frequencies. In the healthy controls (n=1085), 717 (66.1%) were homozygous for an M phenotype. Allelic variants were rare, accounting for only 0.7% of all alleles. The α_1AT deficiency variant Pi^Z was absent in both the study group and the healthy control group. There was no significant difference in the proportion of homozygous and heterozygous M phenotypes between the normal and the ANCA+ group (exact χ^2 test, p=0.56) and between anti-PR3 and anti-MPO (exact χ^2 test, p=0.80). ANCA+ patients had a higher proportion of variant alleles, but this did not reach significance (1.26% *v* 0.70%; exact χ^2 test, p=0.10).

The rarity of the Pi^Z allele in oriental[5] and black populations[6] has been previously reported, a finding which is confirmed for Chinese patients in this study. We found no association of α_1AT variant phenotypes with ANCA in Chinese patients. It is interesting to note the higher number of anti-MPO positive patients in the study group. In a separate study the anti-MPO to anti-PR3 ratio in Chinese patients diagnosed over a defined period was 1.4:1 (the reverse of the situation in white populations, where PR3-ANCA positive Wegener's granulomatosis is much more common).[7] Even for the Chinese patients who tested positive for PR3-ANCA, the positive predictive value for Wegener's granulomatosis was less than 25%. The low prevalence of α_1AT variant phenotypes may be one factor behind the uncommon presence of anti-PR3 in Chinese people. We conclude that α_1AT does not have a significant role in ANCA

Table 1 α_1 Antitrypsin phenotypes in ANCA (anti-PR3* or anti-MPO*) positive patients*

Phenotypes	*All ANCA+ No (%)*	*Anti-PR3+ No (%)*	*Anti-MPO+ No (%)*
Homozygous M			
M1 M1	99(63)	36(60)	63(65)
M2 M2	4(3)	2(3)	2(2)
M3 M3	0(0)	0(0)	0(0)
M4 M4	0(0)	0(0)	0(0)
Heterozygous M			
M1 M2	48(31)	20(33)	28(29)
M1 M3	0(0)	0(0)	0(0)
M2 M3	2(1)	0(0.0)	2(2)
Other heterozygous			
M1 S	2(1)	1(2)	1(1)
M2 S	0(0)	0(0)	0(0)
M1 other	2(1)	1(2)	1(1)
M2 other	0(0)	0(0)	0(0)
Total	157(100)	60(100)	97(100)

*ANCA = antineutrophil cytoplasmic antibody; PR3 = proteinase 3; MPO = myeloperoxidase.

Table 2 α_1 Antitrypsin alleles in ANCA (anti-PR3* or anti-MPO*) positive patients*

Allele	*All ANCA+ No (%)*	*Anti-PR3+ No (%)*	*Anti-MPO+ No (%)*
M1	250(80)	94(78)	156(80)
M2	58(18)	24(20)	34(18)
M3	2(1)	0(0)	2(1)
M4	0(0)	0(0)	0(0)
S	2(1)	1(1)	1(0.5)
Z	0(0)	0(0)	0(0)
Other	2(1)	1(1)	1(0.5)
Total	314(100)	120(100)	194(100)

*ANCA = antineutrophil cytoplasmic antibody; PR3 = proteinase 3; MPO = myeloperoxidase.