focus score of one lymphocyte focus for 4 mm² 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 thymine supplements that returned her thyroid function tests to normal.

Prevalence of neuropathy in patients with SS ranges from 10 to 50%. Polyneuropathy can be the first clinical manifestation of SS and may even precede sicca symptoms in 40% of patients. However, less frequently, cranial neuropathy can occur with a predisposition to involvement of the trigeminal nerve. The vasculitic damage to vaso nervorum documented by pathological studies is associated with a higher incidence of serum anti-SS-A (Ro) antibodies. The association of SS with autoimmune thyroid disease (AITD) is well recognized. ATTD and SS share similarities in the immunopathology in addition to their genetic linkage to the HLA-DR/DQ4 alleles. Only nine cases of facial nerve involvement associated with SS have been described previously.

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 ATTD has not been reported hitherto.

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q. Antitrypsin phenotypic variability is not associated with ANCA in southern Chinese

q. Antitrypsin (α1AT) is a 52 kDa protein encoded by a gene locus Pi on chromosomal segment 1q42.1. 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 α1AT 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, 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 α1AT alleles, not α1AT, with ANCA.

These were largely studies of white subjects, which may not necessarily be extrapolated to all populations. α1AT variant phenotypes may have predisposition to PR3-ANCA, but the same association may not exist for MPO-ANCA. In populations with a low prevalence of α1AT variant phenotypes, the pattern of ANCA could differ from that in white subjects where such variants are prevalent. We set out therefore to establish the distribution of α1AT 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 immunofluorescence, followed by enzyme linked immunosorbent assays (ELISA) for anti-PR3 and anti-MPO. α1AT phenotypes were determined by isoelectric focusing, the results of which were compared with those of healthy Chinese adults.

A total of 137 samples from ANCA+ (either anti-MPO or anti-PR3 positive by ELISA) patients were evaluated, 67 (62%) 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 homozgyous 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 homozgyous M (for example, M1 M2), and 4 (3%) heterozygous for M and a variant allele. Table 1 and 2 show the allelic and phenotypic frequencies. In the healthy controls (n=1085), 717 (66.1%) were homozgyous M (for example, M1 M2), and 4 (3%) heterozygous for M and a variant allele. Table 1 and 2 show the allelic and phenotypic frequencies. In the healthy controls (n=1085), 717 (66.1%) were homozgyous M (for example, M1 M2), and 4 (3%) heterozygous for M and a variant allele. Table 1 and 2 show the allelic and phenotypic frequencies. In the healthy controls (n=1085), 717 (66.1%) were homozgyous M (for example, M1 M2), and 4 (3%) heterozygous for M and a variant allele.
associated diseases in southern Chinese among whom anti-MPO predominated.

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A case of cholesterol embolism with ANCA treated with corticosteroid and cyclophosphamide

We report a case of a patient with cholesterol embolism who showed positive for both myeloperoxidase antineutrophil cytoplasmatic antibody (MPO-ANCA) and proteasome 3 antineutrophil cytoplasmatic antibody (PR3-ANCA) and who was treated with prednisolone (PSL) and cyclophosphamide.

A 50 year old man underwent cardiac catheterisation for back pain. The examination disclosed 90% stenosis of the right coronary artery and a saccular aneurysm in the thoracic aorta. The patient underwent percutaneous transluminal coronary angioplasty and the aneurysm was wrapped with an artificial blood vessel. Postoperatively, the patient had a fever, pleural effusion, abdominal pain, and increased white blood cell (WBC) count, C reactive protein (CRP), and serum creatine. Both of blood and pleural effusion exudate were negative. PSL 15 mg/day was started. However, acute progression of renal failure required haemodialysis.

The patient was transferred to our hospital. Physical examination showed a temperature of 38.0°C and blood pressure of 178/98 mmHg. Cyanosis was noted in both heels and all toes with necrosis and ulcers at the tips of the fifth toes. He had an increased erythrocyte sedimentation rate (ESR) of 82 mm/1st

h. Anaemia was noted with a red blood cell count of 2500×10⁹/l, while the patient’s WBC count was high at 12×10⁹/l. His platelet count (304×10⁹/l) was within the normal range. Biochemistry showed high levels of blood urea nitrogen (10.0 mmol/l of urea), creatinine (710 µmol/l), and CRP (11.3 mg/l). Complements components were within normal ranges. PR3-ANCA and MPO-ANCA were high at 82E and 29E, respectively.

After admission to hospital, circulatory disturbance in his toes worsened. A diagnosis of ANCA associated vasculitis was made based on systemic inflammatory findings and high levels of WBC, CRP, PR3-ANCA, and MPO-ANCA. High dose steroid treatment was started. Biopsies of the right heel skin and thigh quadriceps showed cholesterol embolism (fig 1). However, PSL treatment was continued together with three courses of cyclophosphamide pulse treatment because of persistent fever and high ANCA values. The treatment reduced the fever and toe necrosis, and the ulcers improved. ANCA gradually decreased to normal. The PSL dosage was reduced to 15 mg/day and the patient was discharged.

Cholesterol embolism predominantly affects elderly men with a history of hypertension, atherosclerotic vascular diseases, and renal insufficiency at the time of diagnosis. At least 31% of patients had a preceding history of anticoagulant use or the antecedent performance of a vascular procedure affecting the arterial circulation. The presence of these cholesterol embolisms within the vascular lumen triggers a characteristic localised inflammatory and endothelial vascular reaction. The inflammatory changes resulting from cholesterol embolism may be responsible for many of the systemic manifestations such as fever, weight loss, myalgias, leucocytosis, eosinophilia, and a raised ESR. Thus cholesterol embolism is referred to as both vasculitis look-alikes and pseudovasculitis syndrome. The prognosis is poor, particularly in the presence of acute renal failure.

Three ANCA positive cases of cholesterol embolism have been described. Peat and Mathisson reported an ANCA positive patient with dyspnoea and haemoptysis after acute deterioration of renal function. Cyclophosphamide and PSL improved the systo-moms, but cyclophosphamide was discontinued and the PSL dose was reduced because renal and skin biopsies showed cholesterol embolisms. Subsequently, the patient died of intractable cardiac failure.

Kaplan-Pavlovic et al reported two cases of renal failure with positive MPO-ANCA. The details are unknown for one patient. The other patient was treated with corticosteroid alone. This patient required haemodialysis and amputation of the toes. Although their treatment did not result in the improvement of vasculitis, the combination of PSL and cyclophosphamide was effective in our patient with ANCA.

This result suggests that active treatment with corticosteroid and cyclophosphamide should be considered in ANCA positive cases of cholesterol embolism.

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Figure 1 Skin biopsy specimen showing cholesterol embolism in arterioles within subcutaneous tissues (haematoxylin and eosin, × 400).

Matters arising, Letters