Primary Sjögren’s syndrome associated with chronic periaortitis

Sir, Chronic periaortitis (CP) is a broad term that encompasses three disorders [idiopathic retroperitoneal fibrosis, inflammatory abdominal aortic aneurysm (IAAA) and periarteritis nodosa (PAN)] characterized by similar histopathological alterations, including adventitial and periadventitial inflammation, medial thinning, and advanced atherosclerosis [1].

CP has already been reported in association with various autoimmune disorders, such as Hashimoto’s thyroiditis, and systemic lupus erythematosus (SLE) [1–4].

We describe the first case of primary Sjögren’s syndrome in a patient with an IAAA.

Case report

A 79-year-old male patient was admitted to our hospital because of dull, low-back pain that responded partially to non-steroidal anti-inflammatory drugs, fatigue and anorexia of a few months’ duration. The pain was aggravated by standing and walking.

Past medical history revealed xerophthalmia and xerostomia of 3–4 yrs’ duration.

The patient was taking regularly ramipril 5 mg/day, acetylsalicylic acid 300 mg/day, lansoprazole 30 mg/day and medications for pain including diclofenac 75 mg/day, paracetamol 1 g/day and codeine prn.

On admission, physical examination was unremarkable, except for tenderness over the lumbar–sacral spine and the glutei.

Erythrocyte sedimentation rate (ESR) was 113 mm/1st h and C-reactive protein (CRP) was 13.8 mg/dl (normal values <0.5 mg/dl). A mild chronic disease type anaemia (HB 11.4 g/dl; MCV 90) was noted. Serum creatinine, urine sediment and liver function tests were normal. Rheumatoid factor (111 IU/ml, normal, <20 IU/ml), antinuclear antibodies (titre 1/2560, speckled pattern) and anti-SSA/Ro and anti-La/SSB antibodies tested positive, whereas anti-neutrophil cytoplasmic-antibodies were negative.

Serology for hepatitis C and Epstein–Barr viruses was negative.

X-rays of the lumbar–sacral spine and of the pelvis showed mild osteoarthritis of the spine and hips, which was felt not to be severe enough to account for the patient’s symptoms.

Both abdominal ultrasonography and computed tomography (CT) with enhancement demonstrated an abdominal aortic aneurysm with a diameter of maximum 7 cm.

Endoaneurysmectomy and revascularization were performed to prevent aneurysm rupture. The aneurysm was partially excised and a vascular prosthesis was grafted into the aneurysm of the aortic wall. Histology of the excised aneurysm revealed an IAAA: the aortic wall showed a fibrotic tissue, scattered calcification, atherosclerotic degeneration of the intima and marked adventitial inflammation. The inflammatory infiltrate consisted of lymphocytes, plasma cells and some neutrophils prevalently surrounding the adventitial vasa vasorum (Fig. 1A).

A diagnosis of IAAA was made on the basis of the markedly increased inflammatory markers and of the histological findings.

To evaluate the extent of vasculitis a whole body 18F-fluorodeoxyglucose-positron emission tomography (FDG-PET) was performed. A vascular high-grade (grade 3) uptake in the abdominal aorta and iliac arteries was evident (Fig. 1B).

Twenty days after surgery, the patient was transferred to the Department of Internal Medicine for clinical re-evaluation and further investigations.

Schirmer’s test was positive (2 mm in the right eye and 4 mm in the left eye at 5 min, normal values ≥10 mm/5 min). Minor salivary gland biopsy showed a diffuse inflammatory interstitial
infiltrate consisting of lymphocytes and plasma cells of grade 4 according to Chisholm and Mason [5]. A diagnosis of primary Sjögren’s syndrome was made according to the revised European classification criteria for Sjögren’s syndrome [6]. Prednisolone (1 mg/kg/day) treatment was started with some improvement in the low back pain. The patient was discharged.

Ten days later after his discharge, the patient was referred back to us because of recent onset haematochezia and mild left iliac fossa pain. A repeat abdominal CT-scan with enhancement showed no signs suggestive of aneurismal fistulation, whereas blood tests showed decreased haemoglobin levels (9.2 g/dl) with MCV 86.7 fl.

Blood transfusions were performed to treat the anaemia, however, the patient’s clinical condition rapidly deteriorated and he died soon afterwards. Necropsy demonstrated a left posterior–lateral transmural myocardial infarction, which was judged to be the likely cause of death.

**Discussion**

CP has already been described in association with various autoimmune disorders, including Hashimoto’s thyroiditis, vasculitis, SLE and primary biliary cirrhosis [2–4]. To our knowledge, this is the first reported case of primary Sjögren’s syndrome associated with CP. Recently, a case-control study, which compared inflammatory and non-inflammatory abdominal aortic aneurysms, showed an autoimmune disease in 19% of patients with IAAA, compared with none of the control subjects [2]. Likewise, in a recent prospective study Vaglio et al. found that in 7 (44%) out of 16 cases CP was associated with other autoimmune conditions, namely ANCA-positive renal disease and autoimmune thyroiditis [3].

The association with other autoimmune disorders suggests that CP is a manifestation of a systemic autoimmune process rather than an exaggerated local reaction to atherosclerosis [4]. In this particular case, the presence of constitutional symptoms, raised acute-phase reactants, and increased vascular uptake in the abdominal aorta and/or iliac artery at FDG-PET scan consistent with large-vessel vasculitis further support the concept that IAAA is a systemic inflammatory process [1, 7].

This case, together with the published literature, suggests that patients with CP should be investigated for the presence of underlying autoimmune disorders, including Sjögren’s syndrome.

**Acknowledgements**

The authors gratefully acknowledge Dr Alberto Cavazza (Institute of Pathology) and Dr Annibale Versari (Institute of Nuclear Medicine) for their skilled and kind co-operation.

The authors have declared no conflicts of interest.

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Accepted 15 December 2006

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**Rheumatology** 2007;46:720–721
doi:10.1093/rheumatology/kem011
Advance Access publication 16 February 2007

Neonatal antiphospholipid syndrome associated with heterozygous methylenetetrahydrofolate reductase C677T and prothrombin G20210A gene mutations

Sir, Neonatal antiphospholipid syndrome (APS) is a rare clinical entity characterized by neonatal thrombotic disease due to the transplacental passage of maternal antiphospholipid antibodies (aPL) [1]. While women with aPL show a high incidence of obstetric and fetal complications, the aPL-related thrombosis in their offspring seems to be exceedingly rare. The low frequency of neonatal thrombosis has been attributed to the lack of the most known ‘second hit’ risk factors in infants, and to a low transplacental passage of IgG2 subclass of aPL, which are responsible for most clinical pathogenicity [2]. We describe a neonate who developed ischaemic stroke in the left middle cerebral artery and was found to have positive aPL together with two underlying inherited prothrombotic disorders.