Clinical vignette

Giant cell arteritis followed by idiopathic retroperitoneal fibrosis in the same patient—an unexpected positron emission tomography finding

A 78-year-old male patient was diagnosed with GCA, confirmed by positive histology of the temporal artery. Treatment with prednisone 1 mg/kg of body weight was initiated, tapered and stopped after 2 years. Two years later, with unremarkable regular controls, the patient presented with lumbar pain, elevated ESR (41 mm/h) and CRP (27 mg/l). PET-CT scan (Fig. 1A) was performed, showing moderately elevated fluoro-deoxy-glucose (FDG) uptake in both femoral arteries and a retroperitoneal metabolically active mass partially obstructing the left urether. MRI scans of the abdomen (Fig. 1B) were consistent with the diagnosis of idiopathic retroperitoneal fibrosis (IRF) with left-sided grade I hydronephrosis. IgG\textsubscript{4} was initially elevated to 2.00 g/l (normal range 0.08-1.4 g/l) subsiding to 0.62 g/l under treatment with prednisone (1 mg/kg body weight). With 5 mg of prednisone per day, both diseases have remained inactive for the past 6 months.

This case describes a patient suffering from both GCA and IRF, an IgG\textsubscript{4}-related sclerosing disease often associated with elevation of the IgG\textsubscript{4} subclass. IRF has not yet been described in patients with GCA. However, similarities in histopathology with inflammation in the medial and adventitial layers of the aorta suggest common yet unproven pathogenetic mechanisms for IRF and GCA.

Disclosure statement: The authors have declared no conflicts of interest.

Peter A. R. Tölle\textsuperscript{1}, Frederike Kesten\textsuperscript{2} and Thomas Daikeler\textsuperscript{3}

\textsuperscript{1}Department of Rheumatology, Bethesda Hospital,
\textsuperscript{2}Outpatient Department of Medicine and \textsuperscript{3}Department of Rheumatology, University Hospital of Basle, Basle, Switzerland.

Correspondence to: Peter Tölle, Department of Rheumatology, Bethesda Hospital, Gellertstrasse 144, Postfach, 4020 Basle, Switzerland.
E-mail: petertoeille@web.de; peter.toelle@bethesda.ch

(A) The PET scan shows focal FDG enhancement periaortical at the level of the aortic bifurcation (difficult to discriminate from the adjacent urether) and retention of nuclide activity due to hydronephrosis. (B) The transverse MRI of the abdomen with gadolinium-enhanced T1-weighted, fat-saturated fast spin echo shows marked periaortic uptake of contrast material just proximal to the aortic bifurcation.