An unusual presentation of giant cell arteritis in a middle-aged woman

Sirs,

A previously healthy 56-year-old saleswoman, non-smoker, presented with deteriorating left arm weakness with no prior trauma. At presentation, she was afebrile. Her left arm was mildly colder than the right and she could not abduct it beyond 45°. The left radial and brachial artery pulse intensity was diminished, while arterial blood-pressure (130/80mmHg) and deep-tendon reflexes were comparable in both arms. Review of systems and rest of physical examination was unremarkable. Laboratory tests revealed normocytic anaemia (Hb:9.6g/dl, MCV:86fL), thrombocytosis (platelets: 525000/μL), high erythrocyte-sedimentation-rate (ESR: 130mm/h) and C-reactive-protein (CRP: 154mg/l, normal value <5mg/l). Repeated blood cultures and perinuclear ANCA (anti-PR3 and anti-MPO) were negative, while serum IgG4/IgG2/IgE levels where within normal limits. Chest radiograph was unrevealing and left shoulder magnetic-resonance-imaging did not show supraspinatus pathology. Abdominal ultrasound ruled out the possibility of an occult tumour or abscess.

Due to persisting laboratory findings suggesting systemic inflammation a temporal artery biopsy was performed. Histology (H&E staining) showed relatively intact media, intensely thickened intima and adventitia with severe luminal narrowing. The internal elastic lamina was disrupted and dense inflammatory cells involving the outer intima, media, and inner adventitia with sporadic giant cells of Langhans-type were present, findings compatible with the pathologic lesion seen in giant cell arteritis (GCA). Anti-IgG4 immunostaining did not identify increased number of IgG4-positive plasma cells. Based on the positive temporal artery biopsy, the clinical manifestations and the compatible laboratory findings, the diagnosis of giant cell arteritis was undertaken. In order to assess the extent of arterial vasculature involvement in this GCA patient ^{18}Fluorodeoxyglucose-positron-emission-tomography (PET) was performed which showed intense tracer uptake throughout both carotid, axillary and subclavian arteries as well as the thoracic and abdominal aorta, and the inguinal arteries (Fig. 1).

Oral prednisolone (60 mg/day) was initiated with prompt amelioration of left arm weakness and normalisation of inflammatory indices within 8 weeks. After four months of treatment, when prednisolone was 20mg/day the patient presented relapse of left arm weakness and increased inflammatory markers. Subcutaneous tocilizumab 162 mg/week was added to the patient's therapeutic regimen. Clinical manifestations subsided and prednisolone was tapered to 5 mg/day, without relapse, after six months of tocilizumab treatment.

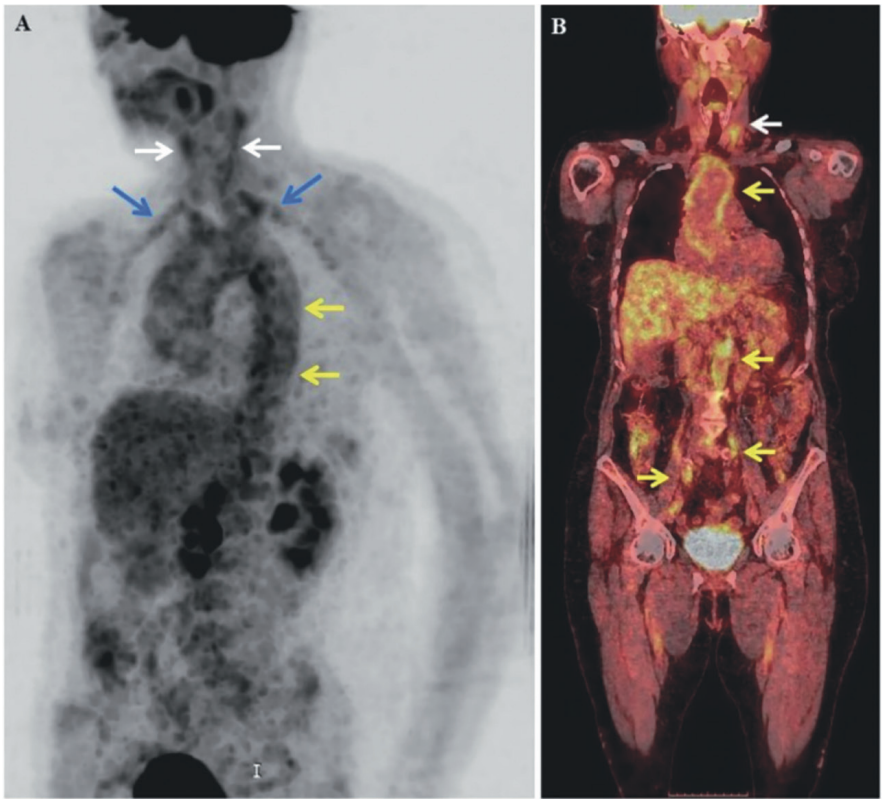

Fig. 1. Maximum intensity projection image (MIP): (**A**) and coronal fused ^{18}Fluorodeoxyglucose positron emission tomography (PET)/CT image (**B**) of a 56-year-old female patient with biopsy-proven giant cell arteritis showing involvement of large-vessels with intense uptake throughout both carotid (white arrows) and subclavian arteries (blue arrows), the thoracic and abdominal aorta, the superior mesenteric artery and the inguinal arteries (yellow arrows).

This case is presented to remind physicians that in all patients with suspicious signs of GCA or even confirmed GCA, a thorough vascular examination, beyond the temporal arteries, should be performed. Autopsy and imaging studies have shown that in many GCA patients, large vessels, like the aorta and its major branches, are affected in addition to cranial arteries (1, 2). Furthermore, this biopsy-proven GCA case with large-vessel involvement could raise the argument whether-or-not our patient has GCA-Takayasu's arteritis (TAK) overlap. Although GCA and TAK are traditionally considered distinct large-vessel vasculitis (LVV), based on differences in age of onset, ethnic distribution, clinical manifestations and predilection of affected arteries, several studies have vitiated the discriminatory boundaries between these two entities. TAK has been reported in older patients (3, 4) and imaging studies in cranial GCA patients, particularly those aged between 50–60 years, have revealed large-vessel inflammation in approximately 60% of them (5). Nonetheless, regarding at least the management of these two LVV, this argument is only of philosophical value. Both disorders to-date are treated in a similar manner, consisting initially of high glucocorticoid doses (1mg/kg/day) with slow, steady tapering. Upon relapse interleukin-6-receptor antagonist is an alternative treatment option (6).

The current classification criteria for GCA and TAK serve well their purpose in distinguishing among the classical forms of these two entities. Yet, they are inadequate in classifying patients with LVV between 40-50 years of age, as well as older individuals in whom cranial symptoms are absent. Efforts are needed to expand the classification of LVV into subgroups possibly with genotypic studies.

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