Isolated vasculitis of the lower extremities in a patient with polymyalgia rheumatica and giant cell arteritis

Sirs,
A 69-year-old woman, with a history of arterial hypertension with ACE inhibitor therapy and no other risk factors for vascular affections, was admitted to our Neurology Department because of sudden onset of diplopia. Past medical history revealed pain and stiffness in her shoulders and pelvis of about two months' duration. Ophthalmic examination ruled out ischaemic optical neuritis. Brain CT scan, MR, and MR angiography were all negative. Laboratory tests showed elevated ESR (91mm/1h) and C-reactive protein (12.5mg/dl, normal values <0.5mg/dl). Autoimmunity serology (rheumatoid factor, ANA, ENA, and ANCA) was negative. The patient was reviewed by a rheumatologist. On direct questioning, she denied manifestations suggestive of giant cell arteritis (GCA) such as new onset of headache, jaw claudication, or limb claudication. Examination of the temporal arteries was unremarkable. Colour-Doppler sonography of the temporal arteries, aorta, and of its main branches showed no signs of arteritis.

We suspected polymyalgia rheumatica (PMR) and GCA with secondary diplopia. The patient refused to undergo a temporal artery biopsy. 18F-fluorodeoxyglucose positron emission tomography with CT (18FDG PET/CT) showed increased FDG uptake in the areas of the cervical and lumbar inter Spinous bursae, in the shoulders and in the trocanteric region consistent with bursitis as well as in the femoral and tibial arteries on both sides (grade 3 on a 0-3 scale) consistent with vasculitis (1).

PMR with GCA affecting the arteries of the lower extremities was diagnosed. The patient had a dramatic clinical and laboratory response to 50mg/day prednisone. GCA and PMR are common and often overlapping conditions in western caucasians aged 50 or over (2). PMR may present as an isolated entity, but may also frequently be associated with GCA (3). In particular, PMR can be the presenting manifestation of “silent” GCA without clinically overt cranial manifestations (4). PET/CT may have a role in detecting large- vessel vasculitis in some patients presenting with PMR features in whom GCA is suspected, especially 1) in patients with PMR who do not achieve an adequate response to prednisone dosages of 10 to 20mg/day, 2) in patients with PMR and persistent elevated inflammatory markers, and 3) in patients with PMR and severe constitutional symptoms (4). As this case shows, PET/CT can reveal extra-cranial arterial involvement in patients with GCA. Extra-cranial arterial involvement in GCA with or without PMR is probably more frequent than usually suspected (4). Colour-Doppler sonography can also be useful to detect early large-vessel involvement in GCA (5) and is a reliable technique in expert hands (6).

In the literature, claudication of the lower limbs has been reported in association with both diseases. However, lower limb arterial involvement may be underestimated because it is often asymptomatic or pauci-symptomatic (7-9).

Advanced age is a risk factor for peripheral vascular disease, mainly in combination with other risk factors such as diabetes mellitus, atherosclerosis, obesity, and smoking. However, the presence of a systemic inflammatory disease may considerably accelerate the atherosclerotic process (8). In patients affected by GCA/PMR that do not have other risk factors, arteritis may cause per se lower extremity stenoses (whether clinically overt or not) similarly to what occurs in the temporal arteries (8).

Regarding PMR, PET/CT can document widespread inflammation with involvement of the bursae. Bursitis is thought to be responsible for the shoulder and pelvic pain and stiffness that characterise PMR (10, 11).

Fig. 1. 18FDG PET/CT: increased FDG uptake in the areas of the cervical and lumbar interspinous bursae, in the shoulders and in the trocanteric region consistent with bursitis as well as in the femoral and tibial arteries on both sides (grade 3 on a 0-3 scale) consistent with vasculitis.

References

7. KERMANI TA, MATTESON EL, HUNDER GG, WAR-

Unità Operativa di Reumatologia and 2Unità Operativa di Medicina Nucleare, Arcispedale S Maria Nuova, Reggio Emilia, Italy.
Address correspondence to: Dr Giuseppe Germano, Arcispedale S. Maria Nuova, Viale Risorgimento 80, 42100 Reggio Emilia, Italy. E-mail: giuseppe.germano@asmn.re.it
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