Subclinical vasculitis in polymyalgia rheumatica

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Polymyalgia rheumatica (PMR) is an inflammatory condition of the aging population characterised by pain, stiffness, and symmetrical involvement of shoulder and pelvic girdles. It has been proposed that the primary site of disease may reside outside the synovial joint, but its aetiopathogenesis remains ill understood. There is good indirect evidence that vasculitis is important in the pathogenesis of PMR, based on observations from the closely related disease, giant cell arteritis (GCA). Many patients with GCA have PMR-like symptoms and the converse is also true. One of the characteristics of PMR is its dramatic response to moderate doses of corticosteroid treatment. However, a subset of patients responds inadequately to treatment or has difficulty with dose reduction, the basis for which is not well defined. We report here the case of a patient with PMR which was difficult to treat who was found to have underlying vasculitis on magnetic resonance imaging (MRI), a finding which explains the requirement for the higher dose of steroids.

Case report

A previously healthy 77 year old white woman presented with a three week history of acute onset of aching and stiffness in both shoulders and thighs. There was no history of headache, jaw claudication, or visual impairment. On examination there was tenderness and decreased range of movement of both shoulders, worse on the right, with no evidence of joint synovitis or proximal muscle weakness. She was normotensive with equal peripheral pulses. The remainder of the examination was largely unremarkable. Laboratory investigations showed a mild leucocytosis with raised inflammatory markers (plasma viscosity 2.06 mPa.s, (normal range 1.50–1.72)) and hypergamma-globulinaemia. There was no anaemia. Other investigations, including rheumatoid factor and antinuclear antibody, were also negative. A diagnosis of PMR was made, and treatment was started with a daily dose of 15 mg oral prednisolone.

Clinical response was prompt, with resolution of morning stiffness and full range of movements in both shoulder joints after a week’s treatment. Symptoms returned with widespread polyarthritis after attempting steroid reduction to 10 mg daily of prednisolone. Plasma viscosity remained high 1.85 mPa.s. At this time, gadolinium (Gd) enhanced MRI of the worst affected shoulder was performed to establish the extent of disease. Unexpectedly, this showed a high signal around the right brachial artery, with florid oedema of the adventitia but normal vessel calibre compatible with an acute vasculitis. Despite the absence of clinical symptoms suggestive of GCA, in view of the MRI findings a temporal artery biopsy was performed, which was negative. The steroid dose was increased to 30 mg daily. Twelve months after diagnosis, steroid reduction remains slow and difficult with frequent recurrences of musculoskeletal symptoms, in particular, shoulder girdle pain and stiffness, but no clinical symptoms of vasculitis. Plasma viscosity is now normal (1.67 mPa.s). Repeat MRI shows considerable improvement, but incomplete resolution of the vasculitis (fig 1).

Figure 1 Post-gadolinium axial, T2 weighted, fat suppressed sequences of the patient’s right shoulder before (A) and after (B) steroid treatment. The white arrows outline the intense perivascular oedema or inflammatory change, which improved markedly after treatment. The asterisk represents the humerus. A = anterior; P = posterior; C = chest wall.
Discussion
In summary, this patient presented with clinical symptoms and signs consistent with a diagnosis of PMR. The poor response to moderate doses of corticosteroid treatment raised the possibility of an underlying condition, and MRI examination showed evidence of large vessel vasculitis, consistent with GCA. Histologically, clinically occult vasculitis is seen in up to 20% of temporal artery biopsy specimens in patients with PMR who do not have features of GCA.

Although characteristically involving the temporal artery, GCA can affect any branch of the aorta or indeed other vessels, and this may not lead to clinical symptoms. Therefore, the MRI finding of clinically occult vasculitis in the brachial artery is in keeping with the known features of the PMR and GCA spectrum of disease.

This case highlights the close link between PMR and GCA and explains some of the overlap between both, raising a number of issues. Firstly, failure to respond to moderate doses of corticosteroid treatment in PMR may reflect an underlying vasculitic process, such as GCA, which may require higher doses of steroid treatment. Secondly, vasculitis is probably more common in PMR outside the classically recognised temporal artery distribution and, possibly, as in this case, it could directly contribute to the diffuse nature of symptoms. Preliminary positron emission tomography scanning reports support the concept that large vessel vasculitis may be common in PMR.

Thirdly, withdrawal of steroid in PMR is a difficult issue as it is uncertain whether residual pain pertains to degenerative or inflammatory factors. The resolution and scope of MRI are continually improving, and future studies to determine the extent of clinically occult vasculitis in PMR may help in decisions about treatment.

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