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CASE REPORT

Polymyalgia rheumatica as the manifestation of unclassified aortitis

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Abstract Polymyalgia rheumatica (PMR) frequently occurs with giant cell arteritis (GCA). We report here two cases of PMR with aortitis in the absence of diminished pulse and manifestations related to GCA. Contrasted CT, MR angiography, and F-18-deoxyglucose positron emission tomography showed aortitis without stenosis that is not classified into any of large vasculitides. It should be acknowledged that aortitis might present as PMR and imaging studies are recommended.

Keywords Aortitis · Giant cell arteritis · Polymyalgia rheumatica · Takayasu arteritis

Introduction

Polymyalgia rheumatica (PMR) is an inflammatory disorder of unknown etiology characterized by pain and stiffness in the neck, shoulder, and pelvic girdles [1]. The diagnosis relies upon clinical symptoms such as shoulder pain, morning stiffness, and elevated erythrocyte sedimentation rate (ESR) in the absence of any other inflammatory disease. Low-dose prednisolone is highly effective in reversing the symptoms. Giant cell arteritis (GCA) is an inflammatory vasculopathy usually affecting large and medium-sized arteries. GCA is closely related to PMR and the presence of biopsy-proven GCA has been demonstrated in 16–21% of patients with PMR; symptoms of PMR may, therefore, also be viewed as clinical features of GCA [1]. Patients with Takayasu arteritis present with fever, malaise, weight loss, visual impairment, and marked elevation of ESR and CRP with decreased pulse in one or both brachial arteries, according to the American College of Rheumatology classification criteria [2]. However, Takayasu patients with pulseless radial arteries are at an advanced stage of their disease, and this criterion should probably be revised in the light of more modern vascular imaging technology. We report on two patients in whom symptoms of PMR were associated with unclassified aortitis that were interpreted as common features of Takayasu arteritis and large-vessel GCA, a variant type of GCA that affects subclavian and axillary arteries and the aorta [3].

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Case report

Case 1

A 61-year-old man was admitted to our department with a one-month history of stiffness of neck, shoulder, and proximal muscles followed by high fever. Shoulders, upper arms, and thighs were tender to touch but tone and power were conserved. The patient did not complain of headaches, jaw claudication, or visual disturbances. His temporal arteries did not show any local sign of inflammation and his blood pressure was 112/50 mmHg in both arms.

Laboratory investigations showed a white blood cell count of 13,000 per microliter, ESR 120 mm/first h and CRP 11.57 mg/dl (normal value less than 0.24 mg/dl). Blood and urine cultures and a pharyngeal swab were negative. Serologic tests for syphilis were negative. Myeloperoxidase antineutrophil cytoplasmic antibody (ANCA) and proteinase-3 ANCA were negative. The patient fulfilled five of the

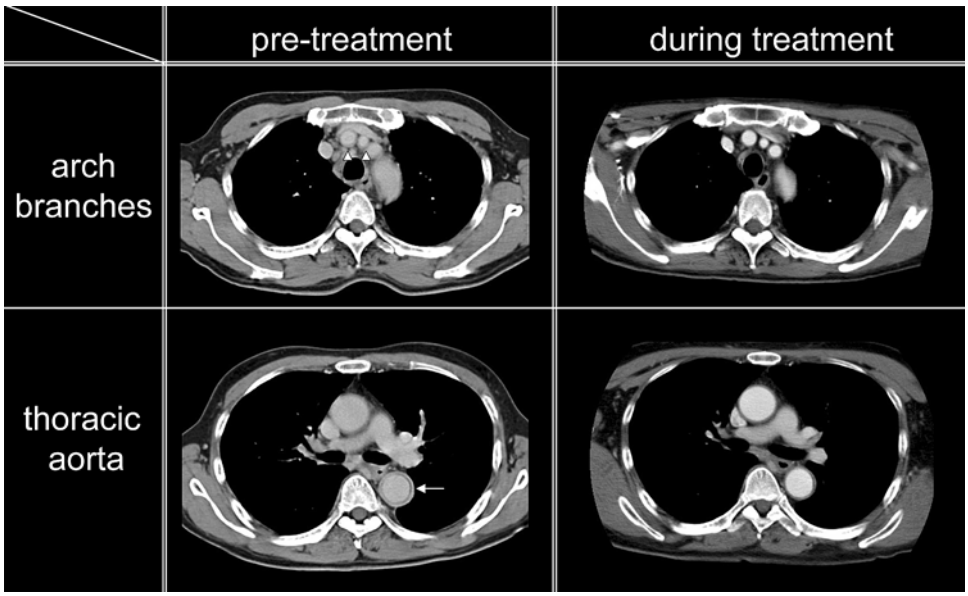

Fig. 1 Thickening of the wall of aortic arch branches (*upper panels*, *arrowhead*) and thoracic aorta (*lower panels*, *arrow*) before administration of prednisolone (*left panels*) and normalized arterial wall during treatment (*right panels*) in the contrasted CT images of case 1

seven Bird classification criteria for PMR: bilateral shoulder pain or stiffness, onset of illness within two weeks, initial ESR over 40 mm first h, morning stiffness exceeding an hour, age older than 65 years, depression and/or loss of weight, bilateral upper arm tenderness [4].

Contrasted computerized tomography (CT) scan and magnetic resonance angiography (MRA) revealed thickening of the aortic arch wall and its branches of the descending and abdominal aorta in the absence of annular ectasias, aneurysms, or calcification (CT images in Fig. 1, left panel). No stenoses were noted in the aorta or the aortic branches. Involvement of temporal arteries was not detected by MRA. F-18-deoxyglucose (FDG) positron emission tomography (PET) study revealed markedly increased uptake in the wall of aortic arch branches and in that of the thoracic and abdominal aorta, indicating inflammation in the vessel walls (Fig. 2, left panel). The patient expressed HLA-B*400201/440301 and HLA-DRB1*090102/130201 alleles. The patient was diagnosed as having PMR and unclassified aortitis, and treated with 30 mg prednisolone daily, leading to prompt remission of all clinical symptoms and normalization of CRP and ESR levels. Four weeks after the initiation of corticosteroid treatment imaging showed thinning of the vessel walls (Figs. 1, right panels, and 2, right panels). After 12 months of treatment with low-dose prednisolone (7.5 mg daily) symptoms have not recurred and CT scan has not revealed any recurrence of aortitis.

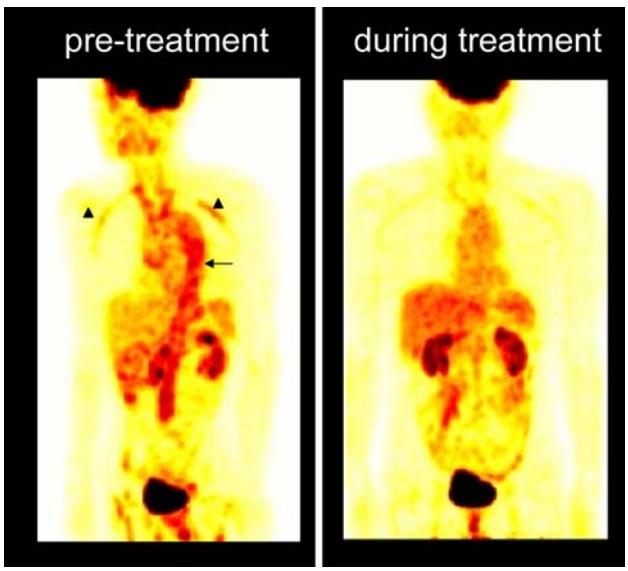

Fig. 2 Increased uptake of 18F-glucose in the thoracic and abdominal aorta (*arrow*) and in the subclavian arteries (*arrowhead*) before administration of prednisolone (*left panel*) and significant decrease in arterial uptake during treatment (*right panel*) of case 1

Case 2

A 63-year-old woman attended our clinic complaining of myalgia and stiffness of her neck and shoulders. Shoulders and upper arms were tender to touch. ESR was elevated at 90 mm first h and CRP at 6.01 mg/dl. She was diagnosed with PMR having satisfied four of the seven Bird criteria. Prednisolone (15 mg daily) rapidly improved her muscle symptoms, but the symptoms of PMR recurred when prednisolone was reduced to 7.5 mg/day six weeks later. On physical examination a bruit was heard at the right side of the neck. Contrasted CT scan and MRA revealed slight thickening of the wall of thoracic and abdominal aorta, and mild stenosis of the right internal carotid artery (Fig. 3). She did not have any clinical signs of GCA. She expressed

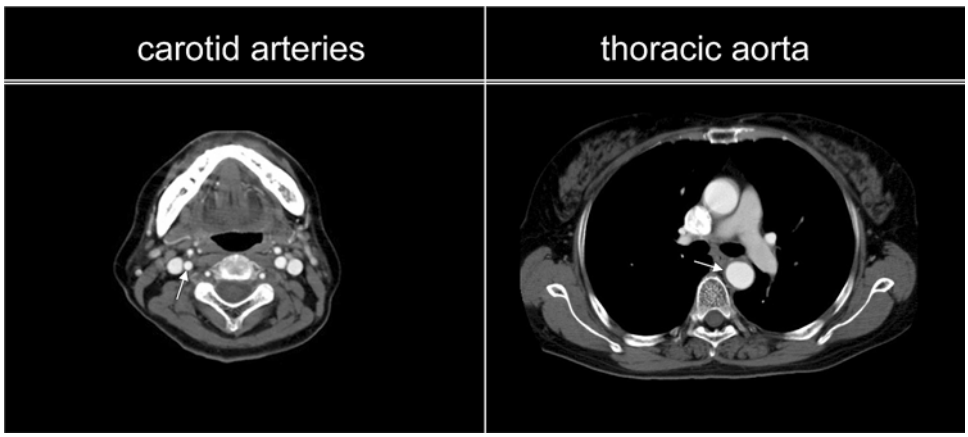

Fig. 3 Mild stenosis of right internal carotid artery (*left panel*, *white arrow*) and thickening of the wall of thoracic aorta (*right panel*, *white arrow*) in the contrasted CT images of case 2

HLA-B*070201/400201 and HLA-DRB1*010101/150101 alleles. She was diagnosed with PMR and unclassified aortitis. She was treated with 10 mg oral prednisolone daily, leading to a prompt response. Thereafter clinical remission was sustained with 7.5 mg prednisolone daily for 10 months.

Discussion

We have described two cases of aortitis presenting as PMR. Current imaging studies using MRA and PET have shown that some cases of PMR refractory to a low dose of corticosteroids underlie large-vessel vasculitis [5, 6]; it may, therefore, not be uncommon for patients with aortitis to present clinically as PMR. GCA and Takayasu arteritis share some clinical or histopathological aspects, and the Chapel Hill Consensus Conference on the nomenclature of systemic vasculitides classified these two diseases as large-vessel vasculitis [7].

However, epidemiology and clinical manifestations of Takayasu arteritis and GCA are significantly different. Takayasu arteritis occurs more frequently in Japan than Europe whereas GCA is less common in Japan compared with other populations [8]. Takayasu arteritis affects women nine times more frequently than men, and approximately 80% of female patients in Japan are below 40 years of age [9]. In contrast, GCA occurs in people aged over 50 years. Focal symptoms such as headache, scalp tenderness, and jaw claudication are common in GCA, whereas these manifestations are unusual in Takayasu arteritis. Moreover, symptoms of PMR are common clinical features in GCA. The muscle stiffness in PMR may relate to systemic inflammation because of an excessively activated innate immune system, presumably sharing physiopathology with GCA [10]. Only 10% of GCA patients have aortic lesions compared with almost all patients with Takayasu arteritis [11]. Large-vessel GCA is a minor subset in which subclavian, axillary, and brachial arteries are mainly involved. Inflammatory infiltrates in temporal arteries were free in 42% of the patients [3]. Headache and jaw claudication are unusual in these patients, but PMR occurs in large-vessel GCA and in cranial (classical) GCA. However, no study has assessed a possible link between PMR and Takayasu arteritis. Patients with large-vessel GCA express an HLA-DRB1*0404 allele whereas patients with cranial GCA express an HLA-DRB1*0401 allele, although both of these alleles are less frequent in the Japanese population than in US individuals [8]. Takayasu arteritis is associated with HLA-B52, B39.2, DRB1*1502, DQB1*0601, and DPB1*0901 [9]. Thus, HLA-B and DRB1 allele patterns may be helpful for distinguishing between large-vessel GCA and Takayasu arteritis.

Our patients were over 50 years of age at onset and had neither jaw claudication nor headache. Their temporal arteries were normal on physical examination and no stenosis was seen on MRA. Contrasted CT, MRA, and PET scan enabled us to demonstrate aortic involvement before the development of stenosis leading to faints, visual impairment, or upper arm claudication due to insufficient blood supply. A number of reports have demonstrated inflammatory findings of Takayasu arteritis by CT scan and MRA [12]. Some cases of GCA with atypical or systemic complaints also have aortic lesions detected by MRA and PET scan [5, 6]. The same imaging systems in our patients clearly demonstrated a thickened aortic wall, and increased FDG in case 1 definitely suggests active inflammation of Takayasu arteritis or large-vessel GCA. These two vasculitides are, however, very close entities in view of symptoms and imaging studies despite differences in age at onset and association of HLA [3]. Our patients did not express any HLA alleles associated with Takayasu arteritis or large-vessel GCA. Taken together, we considered the aortic involvement of our Japanese patients as aortitis that could not be clearly classified into either Takayasu arteritis or large-vessel GCA. It should be kept in mind that an unclassified aortitis may present as PMR in patients aged

over 60 years. A thorough physical examination and extensive imaging evaluation including FDG–PET are recommended to assess large-vessel involvement in patients with PMR.

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