

Case report

Low back pain as presenting manifestation of giant cell arteritis associated to abdominal aortitis

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ABSTRACT

Giant cell arteritis (GCA) is the most common vasculitis in Western countries in individuals over the age of 50. The diagnosis is relatively straightforward when typical features, such as headache, jaw claudication or other ischemic complications are present. Although atypical presentations of GCA have been described, herein we report for first time low back pain as the presenting manifestation of this vasculitis. We also emphasize the importance of considering the use of positron emission tomography (PET) in the evaluation of GCA patients presenting without "overt" cranial ischemic manifestations.

Introduction

Giant cell (temporal) arteritis (GCA) is the most common primary systemic vasculitis in Western countries, in particular in the elderly (1). It is characterized by the granulomatous involvement of large and medium-sized blood vessels of the aorta with predilection for the extracranial arteries of the carotid artery (2, 3). The diagnosis of this condition is relatively straightforward when typical features, such as headache, jaw claudication or other ischemic complications are present (2). In some cases, however, GCA may present without "overt" cranial ischemic manifestations (3, 4). Herein, we report for first time low back pain as the presenting manifestation of this vasculitis.

Case report

We describe the case of a 64-year-old woman who presented to hospital in March 2005 because of severe low back pain.

Her past medical history was unremarkable except for the presence of autoim-

mune hypothyroidism in 1984 that was successfully controlled by substituted thyroid hormones, and an uncomplicated surgical intervention for interauricular communication in 1994.

Prior to the onset of back pain and between March 2003 and June 2004 she had been periodically followed at the out-patient Rheumatology Clinic because of a clinical picture compatible with polymyalgia rheumatica (proximal muscular pain involving the shoulder and hip girdles along with stiffness and elevation of inflammatory laboratory parameters) which improved dramatically following an initial dose of 15 mg/prednisolone/day. Afterwards, corticosteroid therapy was progressively tapered until complete discontinuation 15 months later.

In January 2005, she began to complain of severe low-back pain and asthenia. On admission she described great problems to move or resting in the same position in bed during the night. She also recalled a weight loss of 5 kg since the onset of backache (8 weeks before presenting to hospital). No fever, radicular pain, cramps or other related symptoms were described.

On the physical examination the temperature was 37.8°C, and blood pressure was 150/90 mm Hg. The temporal arteries were normal to palpation without decreased pulses or nodules. No murmurs were heard, and chest and abdominal examination was also normal. A lumbar root stretch test was negative. However, vertebral percussion was slightly painful over the last lumbar levels. The examination of peripheral joints did not show synovitis and the neurological examination was also normal.

Her biological parameters at the time of admission were consistent with

an inflammatory process with ESR 56mm/h, C-reactive protein 5.6 mg/dl, mild anemia (hemoglobin 11.5 gm/dl) with normal leukocyte and platelet counts, abnormal polyclonal profile of gammaglobulins and a mild cholestatic pattern. The renal function (serum creatinine 0.82 mg/dl) was normal and the immunological tests, including ANA and rheumatoid factor, only showed positive results for IgG anticardiolipin antibodies at low titer.

At that time a differential diagnosis to exclude an infectious process, malignancy or an inflammatory disease was conducted. Urine and blood cultures (n = 12), serological tests for hepatitis B and C, HIV, cytomegalovirus, parvovirus B19, mononucleosis, borrelia, Brucellosis-Wright and atypical bacteria were all negative and a chest radiograph was normal. A transthoracic echocardiography showed a slight dilatation of right cavities but it failed to disclose valve vegetations. Leukocyte-labelled scintigraphy was also normal. Lumbar CT scan and lumbar MRI with Gadolinium no signs of spondylodiscitis.

Tumoral markers (CEA, CA125, CA19-9, and CA15-3), urinary Bence-Jones test, and bone marrow aspiration were normal or negative and bone scintigraphy excluded metastatic lesions. Gastroscopy and colonoscopy studies as well as gynecological examination, cytological tests and mammography did not disclose the presence of cancer. A thoracic and abdominal CT scan showed mild aortic wall thickening.

Since no infection or tumoral disease became evident at that time, an inflammatory disease was considered and, due to this, an 18-F-fluorodeoxyglucose-positron emission tomography (FDG-PET) scan and a temporal artery biopsy were performed. FDG-PET scan revealed an inflammatory process involving the wall of the whole thoracic and abdominal aortic artery as well as in its cervical (carotid and subclavian) and iliac branches (Fig. 1). The temporal artery biopsy showed inflammatory changes of the arterial wall with the presence of mononuclear and multinucleated giant cells along with disruption of the internal elastic lamina and intimal edema. Aortic aneurysmal

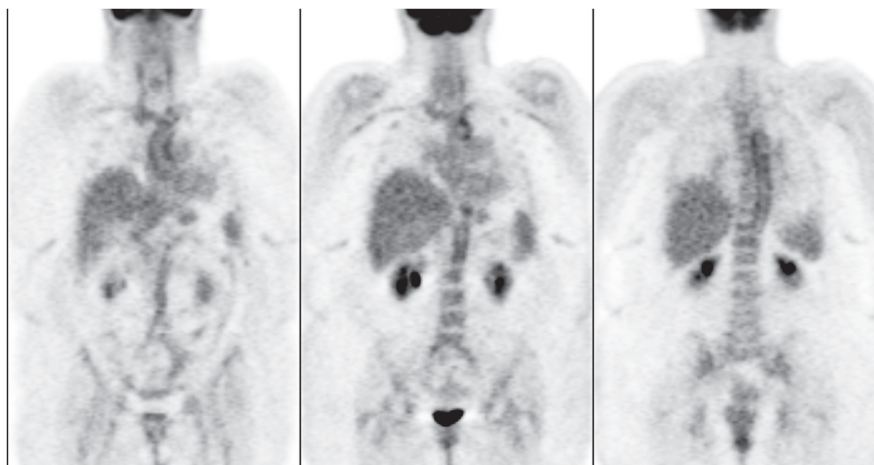


Fig. 1. FDG-PET scan showing an inflammatory process involving the wall of the whole thoracic and abdominal aortic artery as well as its cervical (carotid and subclavian) and iliac branches.

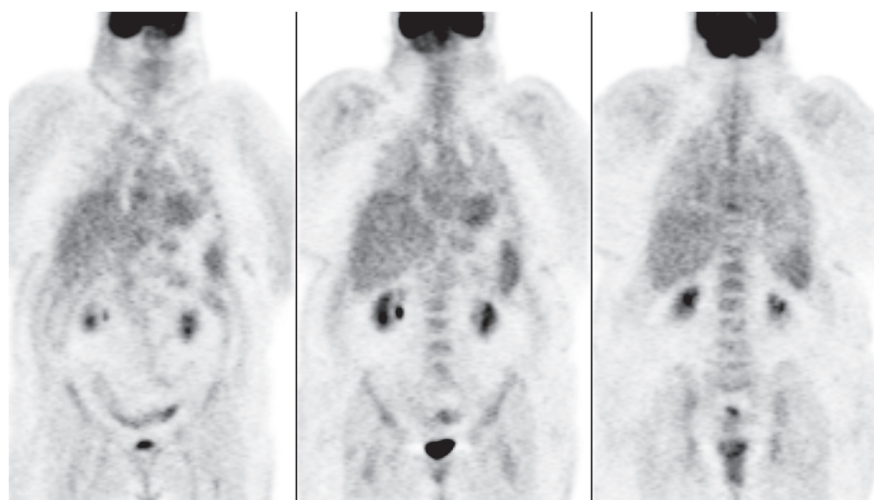


Fig. 2. FDG-PET scan 90 days after the onset of corticosteroid therapy. A negative scan was consistent with response to therapy.

disease was excluded by an angio-CT-scan that did not disclose the presence of aneurysms in the thoracic and abdominal regions.

A diagnosis of biopsy-proven giant cell arteritis was made and initially treatment with methyl-prednisolone (60 mg daily) was started. Also, to avoid vascular complications, antiaggregation therapy (aspirin 150 mg per day) was also given. Following methylprednisolone therapy a dramatic improvement of her back pain was observed (within the first 48 hours after the onset). In addition, rapid normalization of the ESR and CRP was achieved. Seven days later she was switched to oral prednisolone (60 mg/day) and discharged from the hospital free of symptoms. Afterwards, progressive reduction of corticosteroid

therapy was undertaken without having disease relapses or new episodes of back pain. A new FDG-PET scan performed 90 days after the onset of corticosteroid therapy was consistent with the clinical improvement (Fig. 2).

Discussion

Although in most cases a diagnosis of GCA is relatively straightforward due to the presence of cranial ischemic complications (2), sometimes patients may present without "overt" disease manifestations (3, 4). In these cases the search for conditions other than vasculitis is mandatory.

Herein, to the best of our knowledge, we report the first case of a patient with acute low back pain as the presenting clinical feature related to an abdominal

aortitis without aneurysmal process in the setting of with biopsy-proven GCA. In our case, aortic inflammation was confirmed by a FDG-PET scan. This fact underlines the potential role of this technique in the early diagnosis of large vessel vasculitides such as GCA.

The incidence of aortic complications in CGA is estimated to be up to 14% with a female predominance, being the cause of death in 3-12% of these patients (5). Three types of clinical manifestations have been classically described; aortic arch syndrome with diminished brachial pulses and hypertension, aortic valvular insufficiency, and aortic aneurysm and dissection (5, 6). Aortic inflammation may be the presenting feature of GCA or, more commonly, it may occur in patients with pre-existing GCA, often when corticosteroid therapy was reduced or discontinued (5, 6). Recent observations emphasize the importance of performing extended follow-up to GCA since aortic aneurysmal disease and aortic dissection may be observed long time after corticosteroid therapy had been discontinued (5, 6). This is especially true in patients presenting with high inflammatory response at the time of GCA diagnosis (6). However, due to the lack of specificity in its presentation, the diagnosis of aortitis is often hard to be established. In this regard, other inflammatory conditions, such as chronic periaortitis complicated by retroperitoneal fibrosis, are well known to cause lumbar pain at the beginning or during the course of the disease (7, 8). As recently pointed out (7, 8), low back pain may be the presenting symptom as referral pain from an inflamed aortic wall.

Several reports have emphasized the potential value of FDG-PET scan to detect extracranial involvement of GCA and to monitor GCA patients with atypical

presentations (9-11). FDG-PET shows metabolic images and contributes to a better understanding of the pathogenesis of the vasculitic process. In our case it revealed diffuse abnormal FDG arterial uptake of the whole aortic tree, in particular in the abdominal segment. In this regard, a recent study suggests that PET is highly effective in assessing the activity and extent of large-vessel vasculitis with a clinical correlation with CRP and ESR levels (12). PET may also contribute to the evaluation of the response to therapy in vasculitides and disease recurrence (13).

Although FDG-PET is a useful technique for the diagnosis of primary large and medium size blood vessel vasculitis, it may not be sensitive enough to detect vasculitis in arteries smaller than 4 mm of diameter and it was not accurate in vessels of temporal artery size. Since GCA is a vasculitic process affecting cranial vessels, FDG-PET should not replace clinical criteria and, in particular, a positive temporal artery biopsy to achieve a diagnosis of this condition.

In conclusion, the present case highlights the potential use of FDG-PET as an additional tool for the diagnosis of patients with GCA presenting atypical symptoms.

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