

Case report

Detection of asymptomatic aortic involvement in ANCA-associated vasculitis using FDG PET/CT

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ABSTRACT

Large-vessel involvement in ANCA-associated vasculitis is very rare. We report here on the case of two patients with ANCA-associated vasculitis and asymptomatic aortic arch involvement diagnosed using fluorodeoxyglucose positron emission tomography with computed radiographic imaging (FDG-PET/CT). Because aortic involvement in ANCA-associated vasculitis is a potentially life-threatening condition, its early detection can be crucial. FDG-PET/CT may also provide new insights into large-vessel involvement as part of the spectrum of ANCA-associated vasculitis.

Introduction

We report here on two cases of asymptomatic aortic arch involvement diagnosed using FDG-PET/CT in the context of ANCA-associated vasculitis.

Case report 1

The first patient was a 57-year-old Caucasian woman who had presented with progressive loss of visual acuity and polyarthralgia for several months. Physical examination revealed an absence of joint swelling and the temporal arteries were normal. Ophthalmologic analysis showed bilateral optic neuropathy and the histological examination of the temporal artery was normal. Laboratory data were consistent with elevated acute phase reactants (C-reactive protein at 40 mg/l) and the presence of perinuclear antineutrophilic cytoplasmic antibodies (p-ANCA), which were directed against myeloperoxidase (MPO) (49 U/L; n<20). A contrast-enhanced CT scan revealed segmental thickening in the aortic arch. Fluorodeoxyglucose positron emission tomography with computed radiographic imaging (FDG-PET/CT) showed a segmen-

tal and intense uptake in the same area, with a maximum Standardised Uptake Value (SUVmax) of 8.5 (Fig. 1A: Maximum Intensity Projection, MIP, 1B: CT, 1C: fused PET/CT). A diagnosis of ANCA-associated vasculitis with aortic arch involvement was made. The patient was treated with a combination of corticosteroids and intravenous cyclophosphamide, which resulted in a clinical improvement (improvement in visual acuity and polyarthralgia), the disappearance of acute phase reactants and the normalisation of p-ANCA. FDG-PET/CT showed the total disappearance of aortic wall uptake after the 4th pulse of cyclophosphamide.

Case report 2

The second patient was a 66-year-old Caucasian man referred to our unit for an unexplained and prolonged inflammatory syndrome. He had a history of a degenerative lumbar canal stenosis and a monoclonal gammopathy of undetermined significance (MGUS). For several months, he had been experiencing paresthesia in the lower limbs and a dry mouth syndrome. Physical examination did not reveal any headaches, joint pain or swelling, and the pulmonary examination was normal. The laboratory data showed C-reactive protein levels at 33 mg/L and fibrinogen at 6.64 g/L. FDG-PET/CT showed a high and segmental FDG uptake in the aortic arch (SUVmax: 5.1). An enhanced CT scan showed eccentric aortic wall thickening (Fig. 1D: Maximum Intensity Projection, MIP, 1E: CT, 1F: fused PET/CT). During this exploration, high titres of p-ANCA directed against MPO were observed (60 UI/L; n<20). Based on these findings, ANCA-associated vasculitis was suspected. A muscle biopsy was performed and revealed small-vessel wall infiltration by inflammatory

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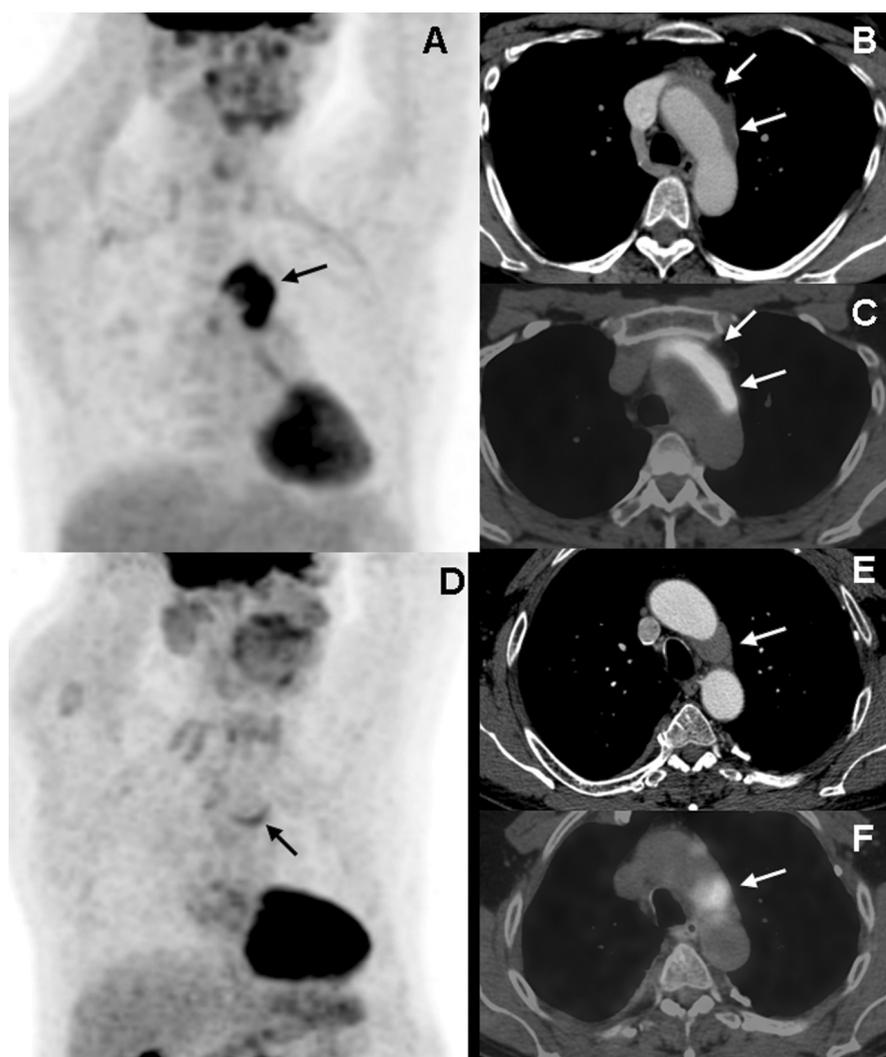


Fig. 1. FDG PET/CT and contrast-enhanced CT findings in the two patients. Imaging findings in the first patient showed segmental thickening in the aortic arch (B, arrows) with an intense uptake in the same area (SUVmax=8.5) (A: Maximum Intensity Projection, C: fused PET/CT, arrows). The imaging findings in the second patient were similar: eccentric aortic wall thickening (E, arrow) with high FDG uptake (SUVmax: 5.1, D, MIP, F: fused PET/CT, arrows).

cells, without any necrosis. Treatment consisted in intravenous cyclophosphamide and corticosteroids, followed by azathioprine maintenance therapy. ANCA-associated vasculitis, which includes granulomatosis with polyangiitis vasculitis (previously Wegener's disease), microscopic polyangiitis, Churg-Strauss syndrome and idiopathic pauci-immune glomerulonephritis, constitutes small-vessel vasculitis associated with the presence of ANCA (c or p-ANCA) directed against either proteinase-3 or myeloperoxidase (1). Large-vessel involvement in ANCA-associated vasculitis is exceptional, with fewer than 20 reported cases (1, 2). The characteristics of the few patients with large-vessel in-

volvement described in the literature differ from those with large-vessel disease such as Takayasu arteritis. Indeed, it mainly occurs in non-Asian patients, often over the age of 50 years; there is no predominance of female patients and peri-aortic lesions are more frequent (1). Thus, perivascular soft tissue masses, aneurism, and dissection with aortic insufficiency and rupture have been described in the context of ANCA-associated large-vessel disease (3-6). The pathogenic process may either involve an accumulation of inflammatory cells in the aortic wall, or aortic wall vasculitis (1, 7, 8). Large-vessel involvement may be observed without there being obvious clinical symptoms

of small-vessel vasculitis (4), as in the cases we describe, and may even precede the manifestations resulting from small-vessel injuries (1, 7).

Conventional imaging techniques such as enhanced CT, MRI or ultrasound scans, can contribute to the diagnosis of large-vessel injuries. However, the findings may be non-specific and not enable a distinction to be made between active lesions and inactive sequelae. FDG-PET/CT is a sensitive and specific whole-body imaging tool for large-vessel vasculitis, especially in patients who are not receiving immunosuppressive drugs (9, 10). It increases overall diagnostic accuracy when combined with conventional imaging modalities (9, 10). FDG-PET/CT is also a tool that can be of value to follow up the efficacy of immunosuppressive therapies, particularly in a context of large-vessel vasculitis.

To our knowledge, this is the first report on the use of FDG-PET/CT for the diagnosis of asymptomatic aortitis in ANCA-associated vasculitis. Insofar, as aortic involvement in ANCA-associated vasculitis is a potentially life-threatening condition, its early detection can be crucial. These two cases highlight the usefulness of FDG PET/CT in detecting asymptomatic aortic involvement. As described with giant cell arteritis, FDG-PET/CT may be of prognostic value regarding late aortic complications (11). FDG-PET/CT may also provide new insights into large-vessel involvement as part of the spectrum of ANCA-associated vasculitis.

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