Aortic angiosarcoma masquerading as inflammatory aortitis

Sirs,

Primary malignant tumour of the aorta is rare and difficult to diagnose, with fewer than 200 cases reported (1). The most frequent clinical presentations of aortic sarcomas are peripheral embolisation, local arterial occlusion or aneurysmal disease, often leading to delayed diagnosis (2). General systemic symptoms are rare and can erroneously lead to a diagnosis of vasculitis and subsequent immunosuppressive therapy (3). We report the case of a 73-year-old woman who presented for persistent elevated C-reactive protein level lasting for 1 year and a 4-month history of fever, weight loss and fatigue. She was otherwise healthy, with no cardiovascular risk factors of atherosclerosis. A thoraco-abdominal CT examination had been performed 1 year before the current admission, and revealed an atheroma-like ulceration of the aortic arch (Fig. 1, A1). With the persistence of symptoms, another CT scan showed a large intraluminal thrombus of the descending aorta with thickening of the aortic wall (Fig. 1, A2-A3). MR angiography images (Fig. 1, B1-B3) were initially interpreted as aortitis. Due to a suspicion of inflammatory aortitis complicated by a thrombus, oral corticosteroids were initiated but did not result in any clinical or biological improvement, so treatment was stopped. A PET-CT scan showed an increased 18F-fluorodeoxyglucose (^{18}F-FDG) uptake of the intra luminal thrombus, with SUV_{max}=4, whereas there was no uptake of the aortic wall (Fig. 1, C1-C3), suggesting malignancy and making the diagnosis of vasculitis unlikely.

After multidisciplinary team discussion, radical surgical resection of the lesion with aortic prosthetic graft placement was performed. Histopathology revealed an intimal, grade III, mildly differentiated angiosarcoma with tumour-free resection margins. No other treatment was initiated. PET-CT imaging performed 3 months after the surgery showed no aortic recurrence but bone metastasis and bilateral tumoural embolisation of iliac arteries. Chemotherapy with taxol was started thereafter.

Aortic angiosarcoma can mimic atherosclerotic disease or thromboembolic disease. Indeed, CT images of angiosarcoma might show linear plaques adhering to the aortic wall or irregular bulging into the lumen with no contrast enhancement of the tumour. However, the absence of diffuse aortic wall involvement or parietal calcifications are atypical for atherosclerosis (4, 5). With an inhomogeneous lesion with protrusive vegetation, an aortic tumour should be considered (6). On contrast-enhanced CT performed during the arterial enhancement phase, one might observe an irregular filling defect without associated aortic wall thickening or extension beyond the aorta in sarcoma (7). CT findings compatible with vasculitis are homogeneous vessel wall thickening >3.0 mm, mural contrast enhancement and luminal narrowing. Results of MRI of aortic angiosarcoma are heterogeneous, and the presence of a lobulated or irregular mass into the vascular lumen with a hypointense lesion on T1-weighted sequences and hyperintense lesion on T2-weighted sequences has been found to suggest the diagnosis. In case of sarcoma, MRI may show vascular wall enhancement together with enhancement within aortic filling defects (8). Lai *et al*. described 6 cases of malignant venous thrombi showing high uptake on ^{18}F-FDG-PET/CT scans, although the initial CT scan had been compatible with venous thromboembolism (9). FDG avidity is found in both venous thrombosis and malignant thrombus, but uptakes differ significantly, as shown in a retrospective review: a SUV_{max} cut-off of 3.63 had 72% sensitivity and 90% specificity for differentiating benign from tumoural thrombosis (10). Finally, ^{18}F-FDG-PET/CT scan highlights the intensity of the arterial wall inflammation with arteritis. In our case, the absence of PET-scan findings compatible

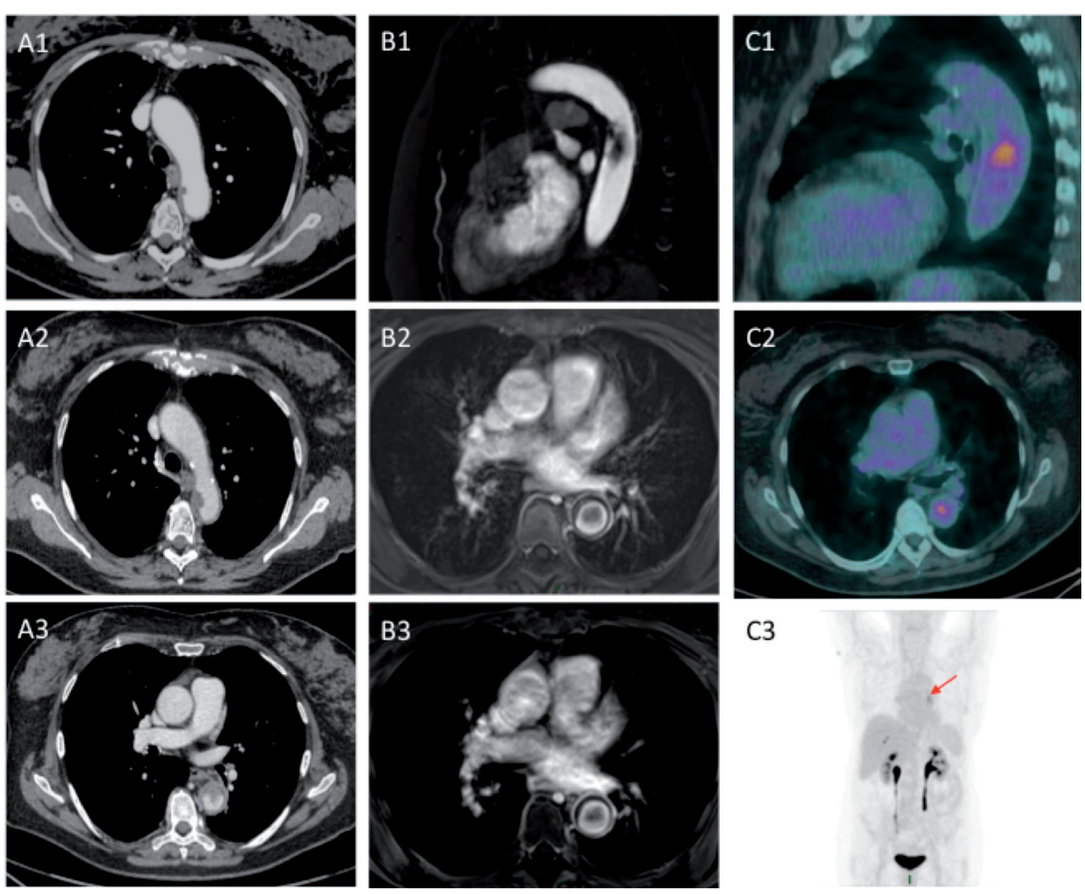

Fig. 1. Multimodality preoperative imaging of the aorta. Contrast-enhanced axial CT performed 1 year earlier, showing a small aortic ulceration with thrombus, suggestive of an atherosclerotic lesion (**A1**). Current thoraco-abdominal CT scan shows a large intra-luminal thrombus of the descending aorta with thickening of the aortic wall (Fig. 1, **A2**-**A3**). MR angiography of the aorta reveals an intraluminal aortic lesion (**B1**), T1-weighted sequences performed 40 seconds (**B2**) and 10 minutes (**B3**) after MRA show a progressive enhancement of the aortic wall without enhancement of the intraluminal aortic lesion. MIP (**C1**) and axial (**C2**) ^{18}F-FDG PET images show intraluminal tracer uptake (SUV_{max} = 4) in the descending aorta sparing the aortic wall. Absence of pathological uptake on large vessel walls (**C3**).
CT: computerised tomography;
FDG: fluorodeoxyglucose;
FISP: fast imaging with steady-state in precession;
MIP: maximum intensity projection;
MR: magnetic resonance;
PET: positron emission tomography;
SUV: standard uptake value.

with vasculitis or atherosclerotic disease and the uptake within the thrombus on ^{18}F-FDG-PET/CT scan was strongly suggestive of alternative diagnoses.

In conclusion, our case highlights the misleading presentation of aortic angiosarcoma due to lack of specific clinical and radiological signs. PET scan might be helpful in difficult situation.

Patient consent was obtained.

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