Aortic angiosarcoma masquerading as inflammatory aortitis

Sirs,

Primary malignant tumor 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 embolization, 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 (18F-FDG) uptake of the intra luminal thrombus, with SUV\textsubscript{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 immature, grade III, mildly differentiated angiosarcoma with tumor-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 tumoral embolization 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 tumor. 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 tumor 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 thrombosis showing high uptake on 18F-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\textsubscript{max} cutoff of 3.63 had 72% sensitivity and 90% specificity for differentiating benign from tumoral thrombosis (10). Finally, 18F-FDG-PET/CT scan highlights the intensity of the arterial wall inflammation with arteritis. In our case, the absence of PET-scan findings compatible...
with vasculitis or atherosclerotic disease and the uptake within the thrombus on 18F-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.

The patient consent was obtained.

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References