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Case report

About two intra-aortic masses

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1. Introduction

Primary aortic malignant tumors are extremely rare; only 157 cases have been reported up to now. Their diagnosis is not easy because the clinical presentation is often non-specific, mimicking multiple other diseases. And the too often delayed diagnosis makes the prognosis poor and treatment hazardous. We describe two patients presenting with an intra-aortic mass. The clinical and radiological signs are similar. The first one is an intra-aortic sarcoma and the second one suggests an aortic thrombus. We will then discuss the management we did, suggesting that sometimes, such as life threatening situations, thrombolysis should be considered.

2. Case reports

Patient 1: A 64 year-old woman is admitted for weight loss, general fatigue and high erythrocyte sedimentation rate (ESR). At admission, the physical examination is normal except for a major clubbing of her left fingers. A thoraco-abdominal CT shows an intra-mural thrombus of the aortic arch and the descending aorta (see Fig. 1A1,); this is confirmed by a transoesophageal echocardiography. The Magnetic Resonance Imaging describes the thickening of the aortic wall with an intra-luminal thrombotic material. The Pet-scan describes an aortic inflammatory process. Although temporal arterial biopsy is normal, corticosteroid therapy for a possible Horton's disease and warfarin are started with a good initial clinical response and a normalization of inflammatory parameters. After 3 months, the ESR increases again. The patient presents now with a left Claude Bernard Horner's sign. A new thoracic CT shows the progression of the intra-luminal thrombus to the descending aorta, and an aneurysm of the subclavian left artery (see Fig. 1, A2). The general status worsens rapidly and the patient develops now a right hemiplegia and diffuse abdominal pain. The urgent surgical procedure demonstrates multiple aneurysms of the ascending and descending aorta as well as of the left subclavian artery. Multiple biopsies disclose an aortic unclassified sarcoma. Our patient dies in the operating room with multiple emboli in the right coronary artery.

Patient 2: An 82 year-old woman, known for type 2 diabetes, atherosclerosis and peripheral arteritis, is admitted for dyspnea, post-prandial abdominal pain and general fatigue. She presents with a systolic aortic murmur and absence of the right radial pulsus. The laboratory test results are normal. A thoraco-abdominal CT shows an intra-aortic mass in the aortic arch (see Fig. 1, B1). Within 2 days, a diffuse livedo reticularis as well as bilateral cyanosis of her toes are noted and our patient goes rapidly into a haemodynamic shock. Surgical treatment of her aortic thrombosis is not applied because of the poor general condition. Thrombolysis is finally given with an improvement of her general status and disappearance of the livedo reticularis. The thoracic CT confirms the total

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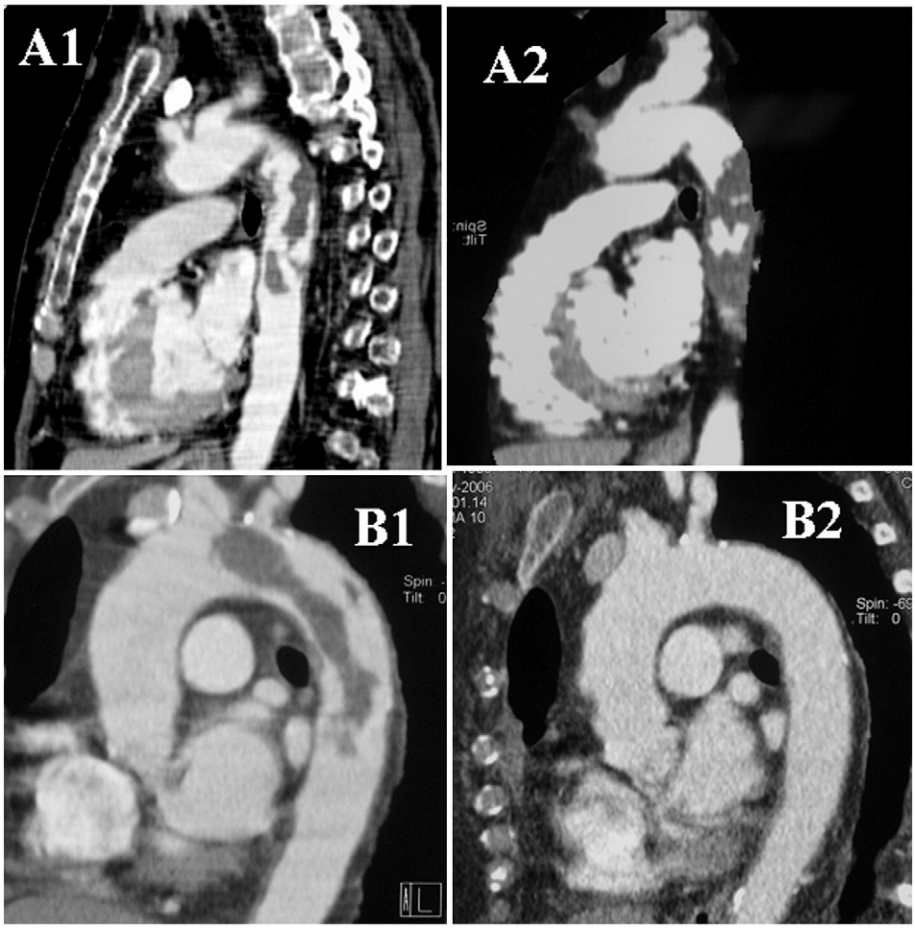

Fig. 1. Thoracic CT of our two patients with intra-aortic mass. A1: patient 1 at admission; A2, patient 1 two months later. B1: patient 2 at admission; B2, patient 2 after thrombolysis.

disappearance of the intra-luminal mass, suggesting its thrombotic nature (see Fig. 1, B2).

3. Discussion

The first case is an aortic sarcoma that was misinterpreted as a vasculitis. The patient disclosed no feature of atherosclerosis and had clinical and biological signs of inflammation, but a normal temporal arterial biopsy. Sarcoma was finally suspected because of the worsening of her clinical and radiological conditions despite the corticosteroids.

Primary malignant tumors of the aorta are extremely rare: 157 cases have been reported in the literature up to now with a male:female ratio of 2:1. They happen at any age and involve more frequently the thoracic or the abdominal aorta. Sarcoma is the most frequent and involves more frequently the intima [1,2].

Symptoms and clinical findings are often related to the type of primary tumor [1–3]. Tumors primarily involving the intima grow into the aortic lumen, causing obstruction and tumoral embolism. Local aortic obstruction can be responsible for claudication, mesenteric angina, hypertension; tumoral embolization leads to peripheral ischemia, mimicking cardiovascular emboli; tumors arising in the media or adventitia present with less precise symptoms such as back or abdominal pain. They may also present as an aortic aneurysm.

Primary aortic tumors occasionally produce systemic symptoms such as fever and elevation of sedimentation rate, mimicking malignancy disease or vasculitis; patients with signs of arteritis not responding to steroid therapy should undergo further examination for vascular malignancy. Most of the cases are diagnosed post-mortem. Intra-aortic biopsy is rarely performed.

Metastases are common, localizing particularly in the bone, the kidneys, the liver, the adrenal glands and the lung [2]. Because of the delayed diagnosis and frequent metastases, the mean survival rate is less than 16 months. But aggressive attitude, including early surgical therapy, postoperative irradiation and systemic chemotherapy is though to prolong survival for several months [1,2,6].

Magnetic resonance imaging (MRI) is the most specific test for the diagnosis of an aortic tumor [4]. Transesophageal echocardiography is also a valuable method to distinguish between an aortic tumor and an atheromatous plaque [5]. Aortography must be avoided because of the risk of embolization.

In our second patient, thrombotic nature of the intra-aortic mass is suspected by the other features of atherosclerosis and is confirmed by its disappearance after a thrombolytic treatment. Aortic thrombosis can complicate general atherosclerosis and can lead to multisystemic complications such as cholesterol emboli dissemination with livedo reticularis, renal failure and haemodynamic shock. Because general status was degrading rapidly with multiple organ failure, and because a local surgical intervention on aortic cross was considered as useless, we performed a last resort general thrombolysis. This thrombolysis was successful with disparition not only of livedo reticularis and cyanosis but also of the intra-aortic mass. So, in some cases, particularly in life threatening situations in which there is no other therapeutic option, thrombolysis should be discussed.

4. Learning points

- Primary aortic tumor should be suspected when an intra-aortic mass is seen with no other feature of atherosclerosis or when the "vasculitis" does not respond to steroid therapy. MRI helps in distinguishing between aortic tumor and thrombotic material. Prognosis of aortic tumors, despite aggressive therapy, is poor.
- An intra-aortic mass in presence of other features of atherosclerosis highly suggests its thrombotic nature and may be, in particular circumstances such as life threatening, susceptible to thrombolysis.

References

[1] Seelig M, Klinler P, Oldenburg A, Blackshear JL. Angiosarcoma of the aorta: report of a case and review of the literature. J Vasc Surg 1998;28:732–7.
[2] Bohner H, Luther B, Braunstein S, Beer S, Sandmann W. Primary malignant tumors of the aorta: clinical presentation, treatment, and course of different entities. J Vasc Surg 2003;38:1430–3.
[3] Pompilio G, Tartara P, Varesi C, Biglioli P. Intimal-type primary sarcoma of the thoracic aorta: an unusual case presenting with left arm embolization. Eur J Cardio-thorac Surg 2002;21:574–6.
[4] Higgins R, Posner MC, Moosa HH, Staley C, Pataki KI, Mendelow H. Mesenteric infarction secondary to tumor emboli from primary aortic sarcoma. Guidelines for diagnosis and management. Cancer 1991;68:1622–7.
[5] Rhee MY, Myong NH, Park YB. Primary intimal sarcoma of the aorta: role of transesophageal echocardiography. Circ J 2002;66(1):111–3.
[6] Chiche L, Mongredien B, Brocheriou I, Kieffer E. Primary tumors of the thoracoabdominal aorta: surgical treatment of 5 patients and review of the literature. Ann Vasc Surg 2003;17(4):354–64.