A 56-year-old, previously healthy man presented with a painless pulsatile tumor of the right neck (Figure 1A). The remaining clinical and neurological examination was unremarkable. Duplex sonography, CT angiography, and digital subtraction angiography revealed large aneurysms of both common and internal carotid arteries (Figure 1B and 1C). Aneurysms of the brachiocephalic and both subclavian arteries were also seen. An abdominal CT showed aneurysms of the infrarenal aorta (Ø 4.8 cm), the superior mesenteric, and both common iliac arteries (Figure 2A). A chest CT was not performed. However, on reevaluation of the abdominal CT three months later, the uppermost section showed the right coronary artery with a diameter of 33 mm (Figure 2B and 2C) that had initially not been recognized. Besides an elevated erythrocyte sedimentation rate (65 mm/h) and C-reactive protein level (41.4 mg/L), laboratory tests were normal, without evidence for vasculitis secondary to other inflammatory diseases (blood count, liver and renal function tests, VDRL, p-ANCA, c-ANCA, antibodies against myeloperoxidase or proteinase 3, ANA, ds-DNA, C3, C4, CH 50).

Two months later, the aneurysmatic right carotid artery was surgically replaced by a Dacron graft. Immunosuppressive therapy with prednisone (1 mg/kg per day) was started under the assumption of a large vessels vasculitis. Two days later, the patient suddenly died after a sharp thoracic pain and collapse despite cardiopulmonary resuscitation. A postmortem examination revealed a ruptured giant aneurysm (Ø 7 cm) of the right coronary artery and a ruptured hemopericardium (Figure 3A), with four liters of sanguineous liquid in the thoracic cavity. All three coronary arteries had aneurysms with large parietal thrombi. On histological examination, vasculitis of the aorta, its branches, and of medium-sized arteries in the liver and lung was found (Figure 3B).

In this case, Takayasu arteritis is the most convincing diagnosis. Inflammation and subsequent neointimal proliferation result in stenotic or occlusive lesions, whereas destruction of the elastica and muscularis may cause dilation or aneurysms.1 Involvement of the coronary arteries is uncommon,2–4 and coronary aneurysms are extremely rare.5 Either coronary ischemia or aneurysm rupture may be fatal. Unfortunately, no surgical or catheter intervention would have been a reasonable option for this patient for surviving this massive destructive vasculitic process.
Figure 1. A, Lateral neck with a pulsatile tumor on the right. B, Digital subtraction angiography (a.p.) of the right common and internal carotid arteries. The aneurysm is 2.0 cm × 7.0 cm. C, Axial sliced CT, showing right internal carotid artery 1 cm from the bifurcation.

Figure 2. Three-dimensional reconstruction of abdominal multi-slice CT-angiography (A), showing elongation and aneurysms of the abdominal aorta, the superior mesenteric artery, and both common iliac arteries. Uppermost scan of the abdominal CT without (B) and with (C) iodine contrast medium showing the heart. A round structure with a diameter of 35 mm was apparent in projection of the right coronary artery (40 Hounsfield units) with slight enhancement (93 Hounsfield units), consistent with a partially thrombosed aneurysm.

Figure 3. A, Ruptured aneurysm (arrow) with a diameter of 7 cm at the origin of the right coronary artery and a parietal thrombus. The length of the rupture site (arrow) measured 2 cm. B, Photomicrographs of a coronary artery showing extensive destruction of the media and infiltration of the entire artery wall with lymphocytes, plasma cells, and granulocytes, but without granulomas. Note the narrowing of the vasa vasorum (arrow) with fibromuscular thickening of its wall (magnification ×12.5; hematoxylin-eosin stain). Immunohistochemical staining revealed a predominance of CD4+ T lymphocytes and VS38+ plasma cells (not shown).
A Swelling of the Right Neck and Sudden Death
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