Coronary aneurysm is usually defined as abnormal dilatation with a diameter greater than 1.5-fold that of adjacent normal segments, and it may be incidentally found in patients undergoing coronary angiography. Coronary aneurysm is reportedly present in 1.4% of postmortem examinations, sometimes multivessel coronary arteries, but rarely left main coronary aneurysm in adults. Coronary aneurysms may be asymptomatic, or they are complicated by thrombosis, rupture of the involved segments, and embolism of the intracoronary thrombus to the distal artery tree producing a small area of infarction. There are various etiologies, such as atherosclerosis, inflammation, coronary artery dissection, trauma, infection, and congenital abnormality. We present an elderly patient who developed multiple coronary aneurysms that were remarkable for the isolated vasculitic lesions found in the coronary media at autopsy.

Case Report

An 81-year-old man was referred in February 1999 for preoperative evaluation of his coronary artery disease. He had been diagnosed with advanced gastric cancer a few weeks previously and had been followed-up for the past 14 years because of angina pectoris. Coronary angiograms in October 1985 showed 99% stenosis of the distal right coronary artery and 75% stenosis of the left diagonal branch (Fig 1). He was treated with isosorbide dinitrate 40 mg/day with relief of anginal attacks and he never had inexplicable fever or skin eruptions.

On admission, his pulse was 84 beats/min and regular, and the blood pressure 134/72 mmHg. Pulmonary breath sounds were diminished at the base of the left lung posteriorly. Complete blood cell count showed hemoglobin 12.3 g/dl, white cell count 5,400/μl, and platelet count 19.1×104/μl. Serum chemistry revealed alanine aminotransferase 64 IU/L, blood urea nitrogen 29 mg/dl, creatinine 0.7 mg/dl, C-reactive protein 2.2 mg/dl, Ig G 2,572 mg/dl.
were negative. Anti-nuclear antibody was 180. Both p- and c-antineutrophil cytoplasmic antibodies (normal, 800–1,800), and Ig M 267 mg/dl (normal, 100–638).

Fig. 3. (a) Right coronary aneurysm showing the extensive inflammatory reaction with massive destruction of the medial layer and concurrent atherosclerotic thickening and mural thrombus in the intimal layer. Capillary blood vessels are seen at the margin between the media and intima of the artery consistent with atheromatous recanalization (Hematoxylin-eosin; ×3). (b) Section of the left coronary aneurysm showing inflammatory infiltrates in the media (grayish area in the middle of the artery), but not in the intima or adventitia (Hematoxylin-eosin; ×3).

Fig. 4. (a) Left coronary artery showing destruction of the internal and external elastic membranes (elastica von Gieson; ×10). (b) Close-up view of the coronary arterial wall showing the inflammatory cells, which are chiefly small lymphocytes and plasma cells (Hematoxylin-eosin; ×50).

(normal, 800–1,800), and Ig M 267 mg/dl (normal, 100–180). Both p- and c-antineutrophil cytoplasmic antibodies were negative. Anti-nuclear antibody was ×40 (speckled type). An electrocardiogram showed normal sinus rhythm and an inverted T wave in lead III. Chest X-ray showed cardiac enlargement (cardiothoracic ratio, 64.8 %) and rightward compression of the trachea by the dilated aortic arch. Computed tomography showed that the ascending and descending aortae were dilated to 5 cm in diameter. Echocardiography revealed normal ventricular wall motion and mild aortic regurgitation. The left ventricular end-diastolic dimension was 51 mm and the % fractional shortening was 37.2%. The patient underwent cardiac catheterization in March 1999 and coronary angiography demonstrated multiple aneurysms alternating with severe stenoses along the entire length of the 3 coronary arteries including the left main truncus (Fig 2), which had developed during the 14-year follow-up. Ultrasonographic evaluations of other peripheral arteries, including the carotid and femoral arteries, revealed simple atherosclerotic plaques without aneurysmal changes. No complications associated with the coronary aneurysms occurred before the patient died of gastric cancer in March 2000.

At autopsy, the heart weighed 450 g and the myocardium contained some small foci of fibrosis. Multiple coronary aneurysms were observed in all the epicardial arteries. On microscopic examination of the coronary arteries, the prominent feature was massive destruction of the medial elements with an extensive inflammatory reaction (Fig 3a,b). The internal and external elastic membranes were destroyed (Fig 4a). The inflammatory cells were chiefly small lymphocytes and plasma cells (Fig 4b) and were confined to the coronary media, not being detected in the intima, adventitia or vasa vasorum. Lymphatic germinal centers, granulomas or giant cells were not found. A moderate degree of atherosclerotic fibrointimal thickening was present and there was a mural thrombus in the right coronary artery. Atherosclerotic intimal thickening was observed in the aortic wall. No evidence of vasculitis was seen in sections of the aorta (Fig 5), peripheral arteries, and other major organs.

Discussion

The present case, an elderly man, developed multiple coronary aneurysms, including the left main truncus, during 14 years of follow-up for angina pectoris, despite his initial coronary angiogram showing no aneurysmal changes. Although there was dilatation of the ascending and descending aortae, there were no aneurysmal changes in other peripheral arteries. Histological examination at autopsy revealed an inflammatory reaction in the coronary medial layer with destruction of the elastic membranes, which presumably caused the aneurysmal dilatation of the vessels. The present case is unique in that cellular infiltrates were confined in the coronary media and were not found in the atherosclerotic coronary intima or the aortic wall. The evidence is strongly against atherosclerosis as the cause of the multiple coronary aneurysms because the inflammation of coronary atherosclerosis is characterized by infiltration of inflammatory cells within the plaque, so we therefore consider that the coronary atherosclerosis is independent of the inflammation in the coronary media.

Systemic vasculitic diseases, such as Kawasaki’s disease, aortitis and polyarteritis nodosa, should be considered as differential diagnosis. Kawasaki’s disease, in which 15% of patients will develop coronary aneurysms5 was excluded because of the lack of a febrile disorder during follow-up after the initial coronary angiography. Aortitis involves the coronary arteries6 but is unlikely to be the underlying disease in the present patient because there was no inflammatory lesion in the aorta or its major branches. Polyarteritis nodosa is a systemic vasculitis that affects the coronary arteries in 50% of the patients7 and microscopically, all layers of the arterial wall are involved, which was not the case for the present patient.

A few cases of ‘isolated coronary vasculitis’ have been described in the literature. Ahronheim et al described a case of coronary adventitial infiltrate causing coronary thrombosis and sudden death8 and Pick et al reported a patient with multiple coronary aneurysms and normal visceral angiography of other organs for whom they considered the find-
ings of isolated coronary vasculitis were most consistent with polyarteritis nodosa.9 Swalwell et al reported 2 autopsy cases of isolated coronary vasculitis that did not have evidence of vasculitis in sections of other organs and the microscopic findings resembled polyarteritis nodosa because the arteritis involved all three layers of the vessel wall.10 The histological features of the present patient are comparable to those of a 68-year-old man reported by Dettmeyer et al.11 The autopsy findings were lymphohistiocytic infiltrates of the coronary media, destruction of the coronary elastic membranes, and distention of the coronary arteries, as well as systemic atherosclerosis, fibrosis of the coronary intima, and no involvement of other organs, which is similar to the present case. They considered an atypical nongranulomatous variant of giant cell arteritis as the differential diagnosis, even though granulomas and polymuclear giant cells were not found.

In conclusion, multiple coronary aneurysms developed during 14 years of follow-up in an elderly patient. There was lymphocytic infiltration of the medial layer and although a definite diagnosis could not be made, we speculate that is an atypical form of vasculitic disease; namely, isolated coronary vasculitis.

References