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Case Report

Giant coronary aneurysm in a patient with familial aortic aneurysm/dissection: Medial degeneration extending to coronary artery

Yoshihiro Morino (MD)[a,*,1], Jun Asada (MD)[a], Taiichi Yamamoto (MD)[b], Yoshio Oh-hashi (MD)[a], Atsushi Yamaguchi (MD)[c], Yoshihiro Sakuma (MD)[a], Shinjo Sonoda (MD)[a], Tsutomu Tamura (MD, FJCC)[a]

[a] *Division of Cardiovascular Medicine, Itabashi-chuo Medical Center, 2-12-7 Azusawa, Itabashi-ku, Tokyo 174-8691, Japan*
[b] *Division of Pathology, Itabashi-chuo Medical Center, Tokyo, Japan*
[c] *Division of Cardiovascular Surgery, Itabashi-chuo Medical Center, Tokyo, Japan*

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Summary Familial thoracic aortic aneurysms and dissections occur as part of known syndromes such as Marfan syndrome, but can also be inherited in families in an autosomal dominant manner as an isolated condition. However, involvement of coronary artery aneurysm/dissections is rare in these patient subsets. A 37-year-old male, normal in skeletal appearance, with a distinct family history of aortic dissection, underwent four major cardiovascular surgeries since age 30 due to giant coronary aneurysm and coronary stenosis, abdominal aortic dissection, and Stanford type A aortic dissection. Pathological examinations demonstrated the left main coronary aneurysm was composed of degenerated, lacerated media, with findings of decreased elastic fibers, deposition of mucopolysaccharide, and cystic medial necrosis. These pathological features found in the coronary aneurysm were identical to those of the aortic wall and radial artery segments in this patient. This case suggests that initial coronary evaluation may be warranted in some families to optimize the clinical management of patients affected with this disease.

Introduction

Although medial necrosis usually occurs as an isolated pathologic process in a subject with aortic disease, there are rare reports of families with multiple members with thoracic aortic aneurysms and dissections in the absence of other clinical manifestations of Marfan syndrome or other

* Corresponding author. Current address: Tokai University Hospital, Kanagawa, Japan. Tel.: +81 3 3967 1181; fax: +81 3 3967 0572.
E-mail address: ymorino@is.icc.u-tokai.ac.jp (Y. Morino).
[1] He is currently working at Tokai University Hospital, Kanagawa, Japan.

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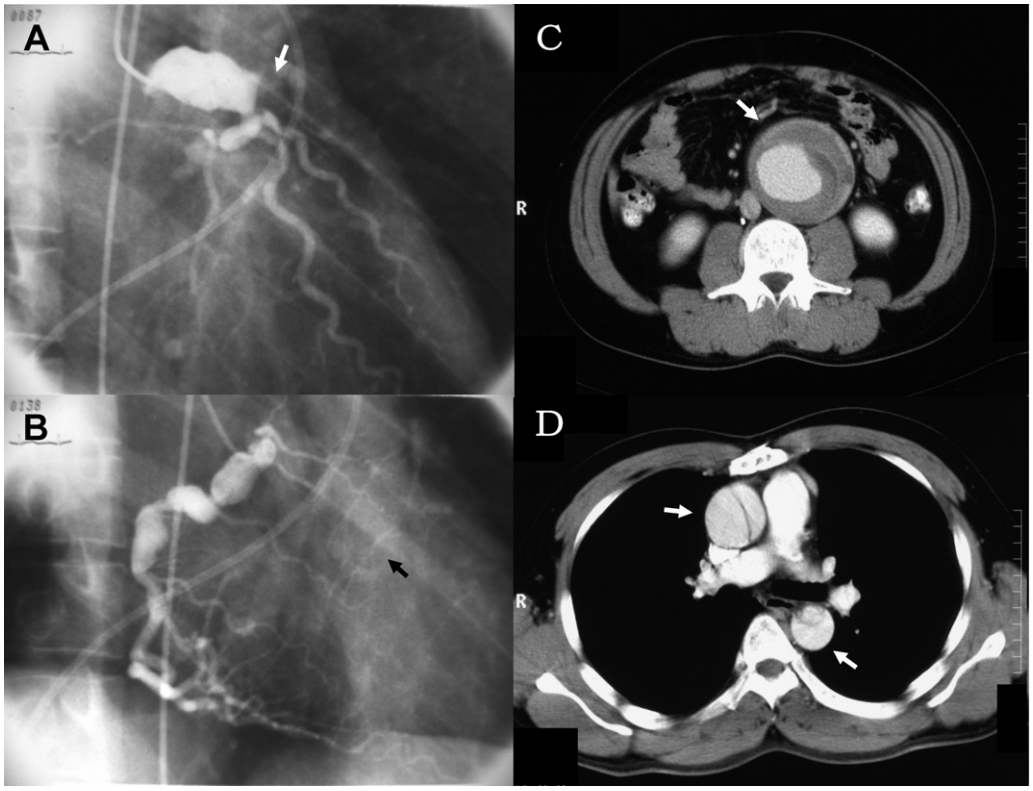

Figure 1 Coronary angiography prior to first bypass surgery showing remarkable aneurysmal formation in left main (A) and the right coronary artery (B). Severe stenosis was observed in the proximal left anterior descending artery (white arrow), filled with collateral circulation from the right coronary artery (black arrow). Computed tomography prior to second and third surgery showing abdominal aortic aneurysm (C) and Stanford type A aortic dissection (D), respectively.

connective-tissue disorders. Between them, extending this pathological degeneration into coronary arteries is considered as infrequent in the literature. We report a case of familial aortic aneurysm and dissection, which was complicated with giant coronary aneurysms. Pathological findings can be compared between a series of surgically resected specimens.

Case report

This case describes a 37-year-old male (172 cm in height and 67 kg in weight), who had repeated episodes of various cardiovascular events, with a distinct family history of aortic dissection. His grandmother died from acute aortic dissection at the age of 81. His mother also suffered from aortic dissection at the age of 57 and took ascending thoracic aortic replacement. All of them, throughout three generations, did not have any characteristic skeletal features of Marfan syndrome.

This patient had gradually worsening effort angina at the age of 30. Coronary angiography revealed giant coronary aneurysm of the left main coronary artery, total occlusion of the proximal left anterior descending artery (LAD), and rosary-bead like appearance in the right coronary artery (RCA) (Fig. 1A and B). Unlike Kawasaki disease [1], any evident calcified findings were not observed in the angiography. At that time, he underwent bypass surgery (bypass grafts: left internal thoracic artery to LAD and saphenous vein graft to diagonal artery and distal left circumflex artery (LCx)) and coronary aneurysmectomy of the left main artery. No surgical revascularization was performed to RCA because no evident stenosis was detected despite multiple aneurysms. Prior to bypass surgery, enhanced computed tomography (CT) demonstrated mild abdominal aortic aneurysm (AAA, 4 cm in maximum diameter). He had no evident past history of Kawasaki disease during childhood and had no coronary risk factors.

At the age of 34, routine follow-up CT of his AAA demonstrated acute enlargement in size (Fig. 1C, 7 cm in maximum diameter), enforcing him to undergo abdominal aortic replacement. Furthermore, 4 months after this surgery, he felt a sudden onset of severe back pain. Emergent CT revealed Stanford type A aortic dissection (Fig. 1D), requiring emergent surgery with ascending thoracic aortic replacement. At the age of 37, he felt recurrence of gradually worsening effort angina. Coronary angiography revealed totally occluded bypass grafts. Therefore, he underwent second bypass surgery (bypass grafts: splenic artery — radial artery graft — LAD and common hepatic artery — saphenous vein graft — LCx).

As described above, he received a total of four major cardiovascular surgeries in his 30s. In order to overview the basic etiology behind these consecutive cardiovascular

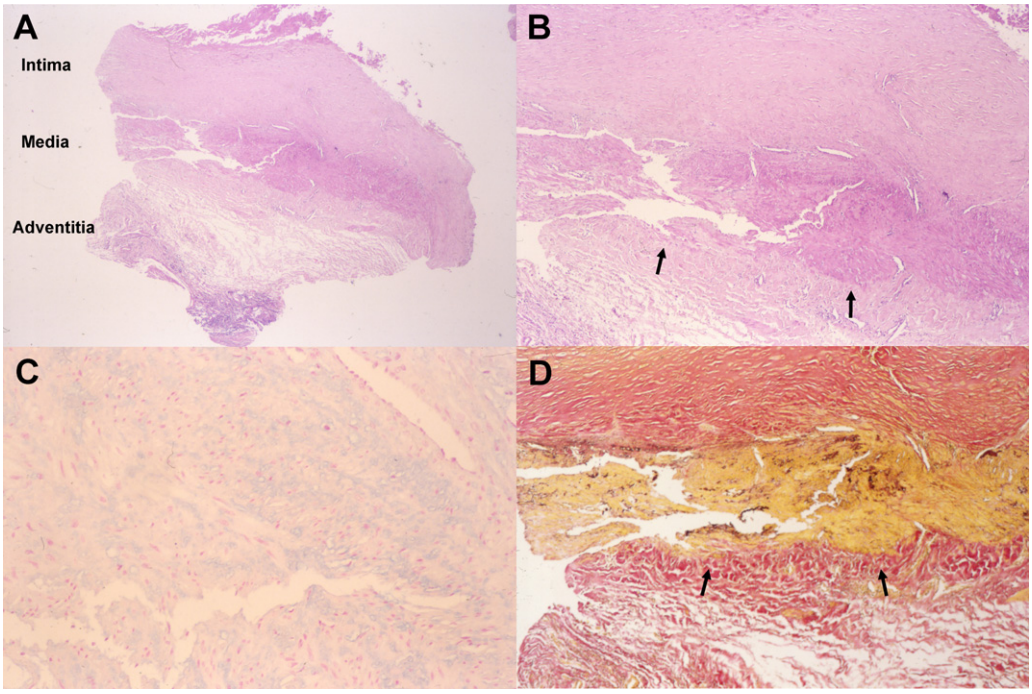

Figure 2 Pathological examination of extracted left main coronary artery. (A) Laceration of media in overview (hematoxylin-eosin stain 13.2×). (B) Loss of the smooth muscle cells at the lacerated media and cystic medial necrosis (arrow) (hematoxylin-eosin stain, 33×). (C) Mucopolysaccharide deposition in the media indicated by blue stains (alcian blue stain, 66×). (D) Decrease and tear of elastic fibers in the media (elastica Van Gieson's stain, 33×).

events, a series of pathological examinations are available. Fig. 2 shows pathological findings of resected left main aneurysm. It was composed of degenerated, lacerated media, with findings of loss of the smooth muscle cells, decreased elastic fibers, and deposition of mucopolysaccharide, consistent with cystic medial necrosis (Fig. 2). These pathological features found in the coronary aneurysm were identical to those of the abdominal aorta (Fig. 3), ascending aorta, and left radial artery segments in this patient. In addition to no typical skeletal abnormalities, he had no lenticular, skin, or lung problems. Thus, despite typical pathological findings and autosomal dominant inheritance pattern, these did not fully meet the established diagnostic criteria of Marfan syndrome.

Discussion

Autosomal dominant inheritance of familial aortic aneurysm and/or dissection, in the absence of other manifestations of Marfan syndrome or other connective-tissue disorders, has been reported since 1977 [2]. Familial aggregation studies indicate that up to 20% of patients with thoracic

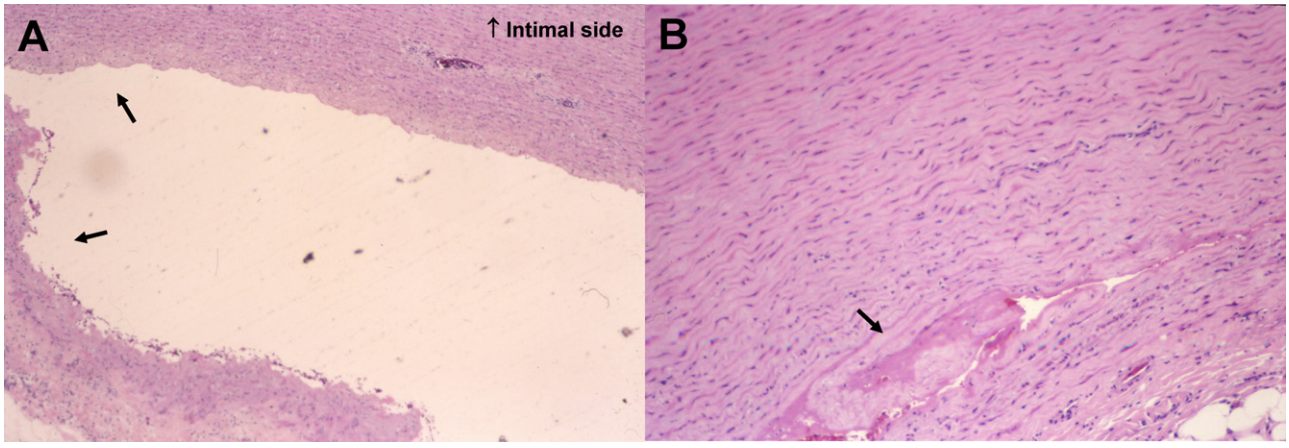

Figure 3 Pathological examination of abdominal aorta. (A) Dissected media in the abdominal aortic aneurismal part (hematoxylin-eosin stain, 33×). (B) Cystic medial necrosis (arrow) seen in the aortic wall in macroscopic normal appearance (hematoxylin-eosin stain, 66×).

aortic aneurysms and dissection who do not have Marfan syndrome have a first-degree relative with the disease [3,4]. In the majority of families, it is inherited in an autosomal dominant manner characterized by decreased penetrance and variable expressivity [5]. Furthermore, several chromosomal loci linked to this disease have been identified [6,7]. This case, with medial degeneration especially ''involving the coronary artery system'', is unique compared with previous reports [5,8], because most cases usually did not involve evident coronary abnormalities. This case suggests that initial coronary evaluation may be warranted in some families to optimize the clinical management of patients affected with this disease. If some specific abnormalities are observed in stress test or other routine non-invasive imaging examinations (echocardiogram or other imaging modalities), coronary CT angiography or even coronary angiography is considered to be necessary.

Further investigations, frequency and prognosis of involvement of medial abnormality in coronary systems, will be required for elucidating actual clinical impact of extension into coronary artery systems as well as establishing optimal strategies for management of this inherited systemic vascular disease.

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