Case Report

Unruptured, Isolated Giant Aneurysm of the Sinus of Valsalva Resulting from Medial Mucoid Degeneration

Hitoshi Inafuku, MD, Yukio Kuniyoshi, MD, PhD, Satoshi Yamashiro, MD, PhD, Katsuya Arakaki, MD, Takaaki Nagano, MD, and Yuji Morishima, MD

We report a quite rare case of unruptured, isolated giant aneurysm of the sinus of Valsalva resulting from medial mucoid degeneration in a young adult woman. A 29-year-old Japanese female diagnosed as having an aneurysm of the sinus of Valsalva and severe aortic regurgitation with no clinical findings of Marfan's syndrome or Ehlers-Danlos syndrome. A modified Bentall's operation was performed successfully, and she was discharged with no complications. A pathological examination revealed marked medial mucoid degeneration of the aneurismal wall. In the literature, most giant aneurysms resulting from mucoid degeneration were found in African young adult females. In this case, there was much mucoid degeneration in the media with no focal destruction of elastic fibers, which was distinct from cystic medial necrosis in Marfan's syndrome. A careful follow-up will be required to detect any other aneurysmal formation in the future. (Ann Thorac Cardiovasc Surg 2009; 15: 203–205)

Key words: aneurysm of the sinus of Valsalva, medial mucoid degeneration, young adult female, Bentall's operation

Introduction

An unruptured, isolated giant aneurysm of the sinus of Valsalva resulting from medial mucoid degeneration is a quite rare cardiac abnormality in young adults in Japan. In this condition, the diagnosis may be late because of an absence of accompanying symptoms. And the aneurysm may cause other complications, such as aortic regurgitation (AR), right ventricular outflow tract obstruction (RVOTO), and coronary artery obstruction. We performed surgery for a giant aneurysm of the sinus of Valsalva, which had caused severe AR.

From Division of Thoracic and Cardiovascular Surgery, Department of Bioregulatory Medicine, Faculty of Medicine, University of the Ryukyus, Okinawa, Japan

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Address reprint requests to Hitoshi Inafuku, MD: Division of Thoracic and Cardiovascular Surgery, Department of Bioregulatory Medicine, Faculty of Medicine, University of the Ryukyus, 207 Uehara, Nishihara, Okinawa 903–0215, Japan.

Case

The patient is a 29-year-old Japanese female having a 6-month history of dyspnea on effort. A systolic heart murmur was noticed during her medical checkup. A giant aneurysm of the sinus of Valsalva and severe AR were observed by echocardiography, and she was referred to us for surgical treatment. Preoperatively, the cardiac catheterization confirmed a giant aneurysm of the sinus of Valsalva, especially dilatation of the right and noncoronary sinuses and severe AR (Seller's grade III), without atrial or ventricular communications (Fig. 1). A computed tomography (CT) demonstrated a 70 × 55 mm giant aneurysm of the sinus of Valsalva. The patient had no clinical evidence of connective tissue disease, such as Marfan's syndrome or Ehlers-Danlos syndrome. Moreover, she also had no clinical or laboratorial evidence of inflammatory disease. Surgical repair for the aneurysm and AR was performed using a usual cardiopulmonary bypass with moderate hypothermia through median sternotomy. The right and noncoronary sinuses were markedly dilated. The aneurismal wall, especially the

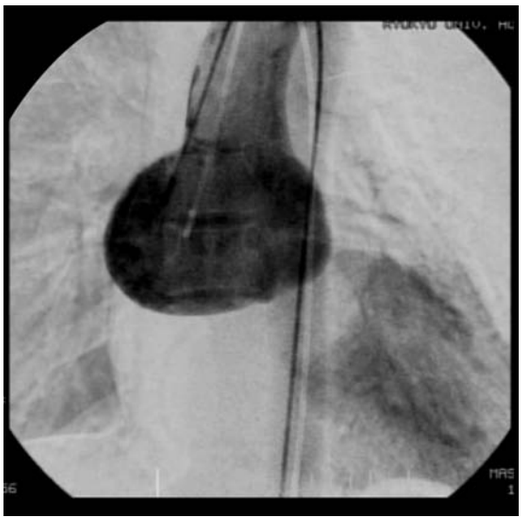

Fig. 1. Aortography revealed a giant aneurysm of the sinus of Valsalva and severe aortic regurgitation.

Fig. 2. Hematoxylin and eosin stains of the aortic wall demonstrated significant mucin deposits in the media (*).

right coronary sinus, was extremely thin and fragile with the right coronary cusp elongation secondary to the stretching of this leaflet, which may cause severe AR. The native valve was therefore thought to be unsuitable for repair. We successfully performed a modified Bentall's type of aortic root reconstruction using the conduit consisting of a bioprosthetic valve and a Dacron graft. The histopathologic examination showed medial mucoid degeneration all around the aneurysmal wall without cystic medial necrosis (Fig. 2). This mucoid degeneration was also demonstrated by additional Alcian blue and elastic van Gieson staining. The postoperative course was uneventful, and she was discharged from our hospital with no complications. No other aneurismal formation was found on her chest or abdominal CT 18 months after the operation.

Comments

Acquired, unruptured, isolated aneurysms of the sinus of Valsalva are uncommon. In particular, the giant extracardiac type is extremely rare and may cause complications, such as aortic insufficiency, coronary artery compression, and right heart failure resulting from RVOTO and arrhythmia.[1–9] The diagnosis in young adults who have an isolated aneurysm of the sinus of Valsalva is relatively difficult until a symptom of the accompanied complications develops. Our present case had suffered from dyspnea on effort as a result of severe AR. She was definitely diagnosed by cardiac catherization as having a giant aneurysm of the sinus of Valsalva and severe AR.

In the recent literature, including our case as summarized in Table 1, 12 cases of unruptured, isolated giant aneurysms (a total of 13 aneurysms) of the sinus of Valsalva were treated surgically.[1–9] Three were associated with hypereosinophilic, Ehlers-Danlos, and Noonan syndromes, respectively.[1,6,9] These aneurysms mostly arose from right and noncoronary sinuses (12/13) and presented complications, namely, AR in 7 (58%), RVOTO in 3 (25%), and compression of the coronary artery in another 3 (25%). In the present case, the aneurysm originated from the right and noncoronary sinuses and developed congestive heart failure resulting from AR caused by a floppy enlargement of the right coronary cusp. So in this case we did a Bentall's type aortic root reconstruction. The microscopic findings of the aneurismal walls in the 12 patients showed mucoid degeneration in 4 (33%), medial necrosis in 2 (16%), medial infiltration of the eosinophils in 2 (16%), and atherosclerotic change in 1 (8%). The mucoid degeneration of the intima and media is a well-described cause of aneurysm in Africans and distinct from an atherosclerotic aneurysm, which predominantly occurs in young adult females and often becomes large before rupture.[10] In the reported 4 patients having mucoid degeneration, 3 (75%) were females. Their mean age was 37 ± 16 (29–63), and the aneurysm's size 7 ± 2 (5–10) cm.

Surgical repair for aneurysms of intimomedial mucoid degeneration is thought to be challenging because of the soft and friable aortic wall and unexplained intraopera-

Table 1. Summary of the recently reported patients, including our surgical case with an isolated, unruptured giant aneurysm of the sinus of Valsalva

Author	Age, sex	Associated syndrome	Location	Size (cm)	Complication	Microscopic finding
Mizushima et al.[1]	24, M	Ehlers-Danlos	R	14	(–)	Cystic medial necrosis
Yasuda et al.[2]	53, F		L	7	AR, RCA compression	Infiltration of eosinophils
	46, F		N	5 × 4	AR	Cystic mucoid degeneration
Tsukui et al.[3]	63, F		N	7	AR, CHF	Mucoid degeneration
Rhew et al.[4]	61, M		R	10 × 10	RVOTO	Atherosclerosis
Chenzbraun et al.[5]	40, M		N	7.5	AMI (LMT occlusion)	
Purnell et al.[6]	49, M	Noonan	R	8.4	AR, RCA compression	Fibrous scar
			N	4.1		and loss of elastin
Yilik et al.[7]	29, M		N	10	Af, CHF	Mucoid degeneration
Joshi et al.[8]	78, M		R	4.2 × 3.5	RVOTO	
	65, M		R	5.9 × 4.9	AR, RVOTO	Cystic medial necrosis
Okinaka et al.[9]	53, F	Hypereosinophilia	R + N	7 × 6	AR, CA aneurysms	Infiltration of eosinophils
Present case	29, F		R + N	7	AR, CHF	Mucoid degeneration

M, male; F, female; R, right; L, left; N, noncoronary sinus; AR, aortic regurgitation; RCA, right coronary artery; CHF, congestive heart failure; RVOTO, right ventricular outflow obstruction; AMI, acute myocardial infarction; LMT, left main trunk; Af, atrial fibrillation; CA, coronary artery.

tive bleeding problems.[10,11] The unruptured, isolated giant aneurysms resulting from mucoid degeneration were considered to develop some severe complications (Table 1) and rupture or dissection.[10,12] Therefore early surgical repair is needed for these conditions, and the operative procedure should be carefully selected, depending on the extent of the abnormal lesion.

Pathologically, our case of mucoid degeneration was different from cystic medial necrosis of Erdheim and G cell, including the cases of Marfan's syndrome. The media in the aneurismal wall of our case showed significant mucoid deposits with no focal destruction of elastic fibers; the media, however, shows cystic necrosis with focal destruction of the elastic fibers of Marfan's syndrome.[12]

We concluded that an unruptured, isolated giant aneurysm of the sinus of Valsalva should be surgically repaired, even if asymptomatic and careful follow-up is needed to detect the pseudoaneurysm at the anastomotic site and the development of new aneurysms elsewhere.

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