Clinical Case Report

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Nontraumatic avulsion of aortic valve commissure as a cause of acute aortic valve regurgitation

A case report

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Abstract

Background: Avulsion of the aortic valve commissure as a cause of acute aortic valve regurgitation is mostly due to trauma, infective endocarditis, or ascending aortic dissection. Nontraumatic avulsion of the aortic valve commissure is very rare. We reviewed the literature and analyzed potential risk factors of nontraumatic avulsion.

Case presentation: An 80-year-old male with hypertension was seen in the emergency department with acute onset dyspnea. Echocardiogram revealed left ventricular hypertrophy with adequate systolic function, prolapse of the noncoronary cusp, and incomplete coaptation of the right coronary and noncoronary cusps with severe aortic valve regurgitation. Surgery revealed an avulsion between the left coronary and noncoronary cusps. Histopathology examination of the aortic valve showed myxoid degeneration, fibrosis, and calcification. Examination of the ascending aorta revealed myxoid degeneration and fragmentation of elastic fibers. Aortic valve replacement was performed, and the patient was alive and well 4 years after surgery. A review of the literature showed that more than three-fourths of the similar cases occurred in males, and about half in patients with hypertension and those 60 years of age or older.

Conclusions: In the case of acute aortic regurgitation without a history of trauma, infection, or valvotomy, when 2 prolapsed aortic cusps are observed by echocardiography in the absence of an intimal tear of the ascending aorta, an avulsion of the aortic commissure should be suspected, especially in males with hypertension who are 60 years of age or older.

Abbreviations: ARD = aortic root dilatation, ARR = aortic root replacement, AVR = aortic valve replacement, F = female, L = left coronary cusp, M = male, N = noncoronary cusp, NA = not available, R = right coronary cusp.

Keywords: acute aortic valve regurgitation, aortic valve prolapse, avulsion of aortic valve commissure

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1. Introduction

Acute aortic valve regurgitation (AR) caused by detachment of the aortic valve commissure from the aortic wall, also known as avulsion of the aortic valve commissure, is usually related to blunt chest trauma, infective endocarditis, or ascending aortic dissection.[1] Spontaneous aortic valve commissure avulsion is a rare condition. It had not been mentioned in recent guidelines or in cardiology textbooks as a cause of acute AR.[2,3] We present a case of acute AR resulting from nontraumatic avulsion of the aortic valve commissure, and review the relevant literature with a focus on potential risk factors and echocardiographic findings.

2. Case report

An 80-year-old male presented to the emergency department with dizziness and shortness of breath for 1 day. He had hypertension, but denied any history of dyspnea, fever, chills, or chest pain. Physical examination revealed tachycardia, a systolic-diastolic murmur grade 2/6 over the aortic area/left lower sternal border, bilateral rales of lungs without lower legs pitting edema, and a distended urinary bladder. An electrocardiogram showed sinus tachycardia and complete right bundle branch block. Chest X-ray showed an increase in the cardiothoracic ratio with pulmonary edema. Echocardiography revealed a hypertrophic interventricular septum, concentric left ventricular hypertrophy (LVH), adequate left ventricular systolic function with an ejection fraction of 68%, prolapse of the noncoronary cusp (NCC) (Fig. 1A), and incomplete coaptation of right coronary cusp (RCC) and NCC (Fig. 1B and C) with severe AR (Fig. 1D).

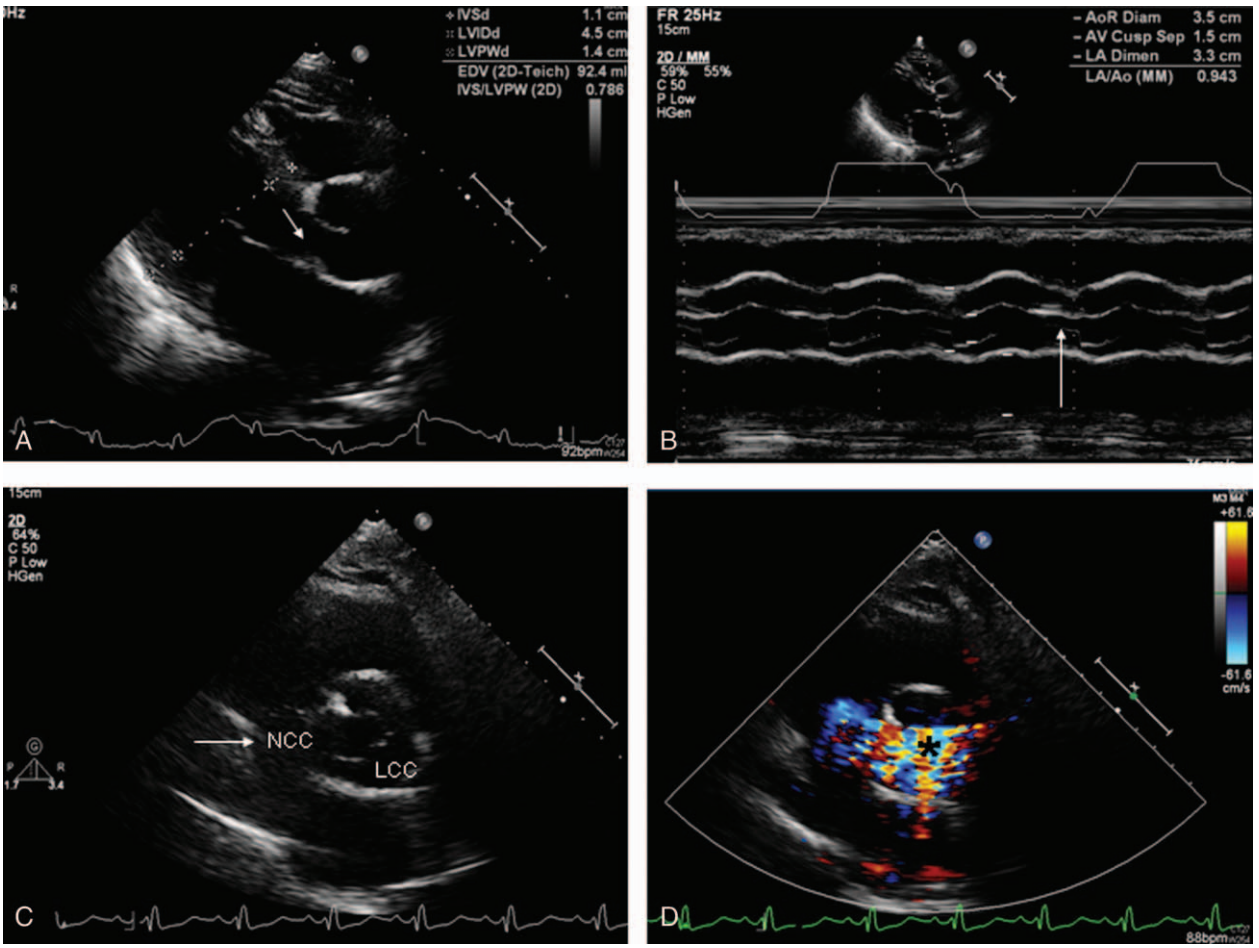

Figure 1. (A) Echocardiography parasternal long-axis view showed prolapse of the noncoronary cusp in the diastolic phase. (B) M-mode at the level of the aortic valve showed incomplete coaptation of right and noncoronary cusps. (C) Parasternal short-axis view showed incomplete coaptation of right, left, and noncoronary cusps. (D) Severe aortic valve regurgitation (asterisk).

The patient underwent intubation for impending respiratory failure, and a cardiovascular surgeon was consulted. Cardiac catheterization revealed a normal coronary angiogram and left ventricular systolic function, as well as severe AR without aortic dissection. Chest computed tomography showed aneurysmal dilatation of the ascending aorta with aortic valve calcifications and moderate bilateral pleural effusions.

The patient's family consented to aortic valve replacement. At surgery, mild atherosclerotic changes of the aortic cusps and prolapse of the left coronary cusp (LCC) and NCC (Fig. 2A), as well as a transverse crack measuring approximately 0.8 cm in length over the LCC–NCC commissure (Fig. 2B) were noted. In addition, concentric LVH and a fusiform aneurysm of the ascending aorta (with a maximal diameter of 4.8 cm) were noted. Aortic valve replacement was performed with a Hancock-II 25 mm porcine valve and longitudinal aortoplasty.

Histopathology examination of the aortic valve showed myxoid degeneration, fibrosis, and calcification (Fig. 3A and B). Examination of the ascending aorta revealed myxoid degeneration and fragmentation of elastic fibers (Fig. 3C and D). Postoperatively, the patient was transferred to the intensive care unit with ventilator and inotropic (dobutamine) support and continuous intravenous dormicum sedation. The patient was eventually successfully extubated, and the dobutamine was tapered. He was then transferred to a general ward and discharged in good condition. He was classified as New York Heart Association Class I when last seen 4 years after the surgery.

3. Discussion

Acute AR can be secondary to trauma, infective endocarditis, type A aortic dissection, annuloaortic ectasia, rheumatic fever, syphilis, bicuspid aortic valve, iatrogenic secondary to a procedure (such as aortic balloon valvotomy), or a failed surgical valve repair.[2] Nontraumatic aortic valve commissure avulsion is rare, but should be considered in a case of AR.

The diagnosis of avulsion of the aortic commissure is difficult prior to surgery on the basis of imaging alone. Transthoracic or transesophageal echocardiography may demonstrate aortic valve prolapse with severe AR,[1,4] which is a nonspecific finding. A more specific finding is the presence of 2 prolapsed aortic cusps, and a limited intimal tear on the commissure may be observed just above the valves.[5] In addition to avulsion of a commissure, the possible etiologies of aortic valve prolapse include floppy valve, fraying valve, excessive tissue within a congenital bicuspid valve, ruptured cusp, laceration of the aorta, and ventricular septal defect with displacement of the aortic root.[1] As prolapse of the cusps may be observed on echocardiography, the commissural tear may mimic an intimal tear in a type A aortic dissection,[5] or a mass-like lesion.[6] In such a case, trans-

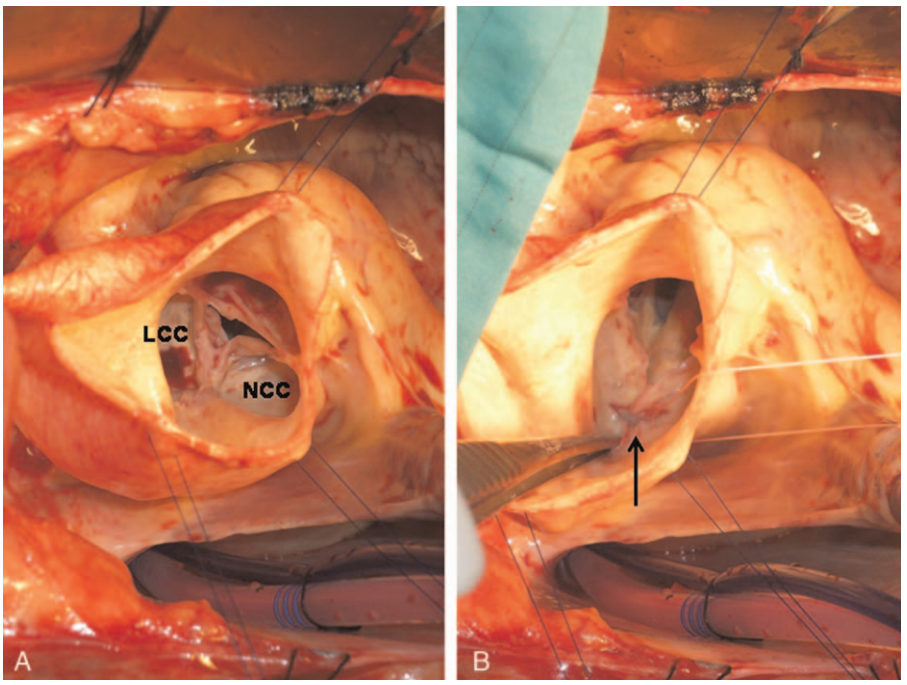

Figure 2. (A) Intraoperative photograph showing mild atherosclerotic changes of the aortic cusps, and prolapse of left and noncoronary cusps. (B) After removal of the aortic cusps, local avulsion of the aortic commissure between left and noncoronary cusps was observed (arrow).

esophageal echocardiography may demonstrate a normal aortic valve with a dissection flap in the proximal ascending aorta just above the aortic valve, but without the intimal tear extending to the aortic root. This is different from a type A aortic dissection, in which an intimal tear may be noted in the ascending aorta. In a case of AR without a history of trauma, infection, or valvotomy, when 2 prolapsed aortic cusps are observed in the absence of an intimal tear of the ascending aorta, an avulsion of the aortic commissure should be suspected. Three-dimensional echocardiography may provide imaging information for diagnosis.[7]

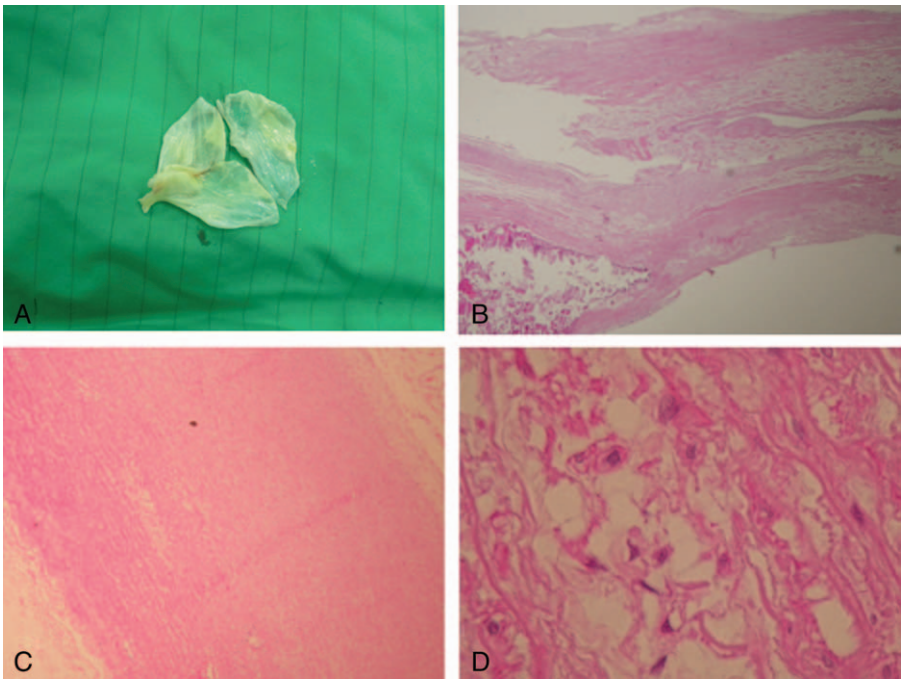

Figure 3. (A) The 3 aortic cusps that were surgically removed exhibited white and firm deformed valvular tissue. (B) Histopathological examination of the aortic valve showed myxoid degeneration, fibrosis, and calcification (×40). (C) Examination of the ascending aorta revealed myxoid degeneration and fragmentation of elastic fibers (×40). (D) Examination of the ascending aorta revealed myxoid degeneration and fragmentation of elastic fibers (×400).

Table 1

Published cases of aortic commissure avulsion.

Case no.		Sex	Age	H/T	Site	Aortic pathology	Aortic valve pathology	ARD	Treatment	Outcome
1	Carter et al[1]	M	43	NA	N-R	Not evaluated		(−)	Medical	Late death
2	Silverman et al[17]	M	47	NA	N-L	Not evaluated		(+)	Medical	Death
3	Silverman et al[17]	F	73	NA	N-R	Cystic medial necrosis		(+)	Medical	Death
4	Satokawa et al[13]	M	25	NA	R-L	Granulation		(+)	AVR	Survive
5	Mok et al[11]	M	78	(−)	R-L	Atheroma		(−)	AVR	Death
6	Aoyagi et al[9]	F	55	(+)	N-R	Normal		(−)	AVR	Survive
7	Aoyagi et al[9]	M	50	(−	R-L, N-L	Normal		(−)	AVR	Survive
8	Sakakibara et al[17]	M	56	(+)	N-R	Medial necrosis		(−)	AVR	Survive
9	Hirata et al[13]	F	68	(−)	N-L	Pseudoxanthoma elasticum		(−)	Repair	Survive
10	Kupersmith et al[5]	M	52	(−)	R-L	Cystic medial necrosis		(+)	ARR	Survive
11	Newcomb et al[20]	M	74	(+)	N-L	Not evaluated		(−)	AVR	Survive
12	Akiyama et al[6]	M	79	(−)	N-R	Cystic medial necrosis	Myxomatous change	(+)	AVR	Survive
13	Fukui et al[10]	M	67	NA	R-L	Normal	Atheroma	(+)	ARR	Survive
14	Okamoto et al[4]	M	68	(+)	N-L	Myxomatous change	Atheroma	(−)	AVR	Survive
15	Nakamura et al[8]	M	62	(+)	R-L	Normal		(−)	AVR	Survive
16	Charokopos et al[19]	F	60	(+)	R-L	Not evaluated		(−)	Repair	Survive
17	Shimamoto et al[7,14]	NA	NA	(−)	N-R	Annuloaortic ectasia		NA	Repair	Survive
18	Shimamoto et al[7,14]	NA	NA	(−)	N-R	Annuloaortic ectasia		NA	Repair	Survive
19	Shimamoto et al[7,14]	NA	NA	(−)	N-L	Dilated sinotubular junction		NA	Repair	Survive
20	Ishikawa et al[12]	M	69	(+)	N-L	Fibrosis, hyalinosis	Mucoid degeneration	(−)	AVR	Survive
21	Cunha et al[16]	M	66	(+)	N-L	Myxomatous change		(−)	AVR	Survive
22	Munakata et al[18]	M	74	(−)	R-N	Elastic fibers tear		(−)	AVR	Survive
23	Yamasaki et al[15]	M	20	(−)	R-N	Granulation		(−)	ARR	Survive
24	Ishii et al[18]	F	54	(+)	R-L	Not evaluated		(−)	ARR	Survive
25	Chang et al, 2016	M	80	(+)	N-L	Atherosclerosis	Myxomatous change	(+)	AVR	Survive

ARD = aortic root dilatation, ARR = aortic root replacement, AVR = aortic valve replacement, F = female, H/T = hypertension, L = left coronary cusp, M = male, N = noncoronary cusp, NA = not available, R = right coronary cusp.

Hypertension plays an important role in the formation of a nontraumatic aortic valve commissure avulsion. Using "avulsion" and "dehiscence" combining with "aortic valve commissure" as key words to search literature indexed in PubMed, we found 5 and 10 cases of nontraumatic aortic valve commissure avulsion, respectively, and we identified 9 more cases from the references of the reports retrieved (Table 1). Including our case, of the 20 cases in which information regarding hypertension was available, 10 had hypertension. Other potential risk factors included male sex (17/22) and older age (13/22 at 60 years or older). The possible pathogenesis of avulsion of the aortic commissure includes high blood pressure, especially high diastolic pressure which avulses a weakened commissure that may be normal[8–10] or have underlying atherosclerosis,[11] fibrosis,[12] hyalinosis,[12] cystic medial necrosis,[5,6,13] pseudoxanthoma elasticum,[14] annuloaortic ectasia,[15] granulation,[16,17] elastic fiber tear[18] or myxomatous changes.[4,19] The aortic valve cusps may be normal,[7] or have mucoid degeneration,[12] atherosclerosis,[4,10] or myxomatous changes.[6]

Without exception, avulsion of the aortic commissure results in progressive congestive heart failure. Medical therapy failed in 3 early cases of avulsion of the aortic commissure, and the patients died.[1,20] Surgery aimed at either repair or replacement of the aortic valve or aortic root is lifesaving, with only 1 death out of 22 cases. In the presence of aortic medial disease, connective tissue disease, atherosclerosis, or myxomatous degeneration of aortic valve, aortic valve replacement is advised to prevent recurrent avulsion. If avulsion of the commissure is accompanied by aortic root dilation or avulsion of the aortic cusp, aortic root replacement is indicated.[5,10,16] Aortic root replacement was performed in 1 case of 2 dehiscences of the aortic valve commissure and cusp in order to reinforce the aortic wall around the left coronary artery ostium.[21]

Aortic valve repair was performed successfully in 5 cases.[14,15,22] Direct suturing of the degenerated and avulsed commissure to the diseased aortic root usually leads to a poor long-term prognosis. Hirata et al[14] sutured the detached commissure to the aortic wall in a patient with pseudoxanthoma elasticum, but the long-term results were unknown. Newcomb et al[23] initially used Teflon pledgeted polypropylene sutures to suspend the commissure to its previous position in a case with a normal aortic valve, but changed to aortic valve replacement due to persistent mild to moderate aortic regurgitation. Charokopos et al[22] suspended the detached commissure to its proper position using 3 Teflon pledgeted 2–0 sutures combined with BioGlue to reinforce the site of repair which was sutured in 2 layers. Shimamoto et al[15] reported a novel method for repair of commissural detachment in 2 cases with annuloaortic ectasia and 1 case with a dilated sinotubular junction. He reimplanted the root in a Valsalva graft, and used 2–0 polyester sutures for the first row and 4–0 polypropylene sutures for the second row. The 9- to 45-month follow-up results were fair.[14] In our case, atherosclerotic changes within the commissure and aortic root were found at surgery. Therefore, we chose aortic valve replacement using a porcine valve, which achieved a good outcome at 4 years after surgery.

In conclusion, avulsion of the aortic valve commissure is a rare cause of acute aortic regurgitation. Transthoracic and transesophageal echocardiography may demonstrate 2 prolapsed aortic cusps with or without a limited intimal tear on the commissure just above the valves. Three-dimensional echocardiography may provide imaging information for diagnosis. Hypertension plays an important role in the formation of an avulsion in patients with atherosclerosis, medial necrosis, myxomatous changes, granulation, fibrosis, hyalinosis, or fibrosis. Surgery is the only reliable method available to treat this condition, which includes aortic valve repair, replacement, and aortic root replacement.

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