Aortic Chordae Tendineae Strands with Significant Aortic Regurgitation: A Case Report and Review of the Literature

Shuai Yuan, MD, Rong Mou, MD, Xi Sun, MD and Yun Mou, MD

Summary
The aortic chordae tendineae strands are suggested to be embryonic remnants of the cusp formation process. We herein describe a 70-year-old male who was admitted to our hospital for shortness of breath and chest tightness. During echocardiographic examination, severe aortic regurgitation with a ruptured fibrous strand was detected. Moreover, another fibrous strand was found by three-dimensional transesophageal echocardiography (TEE). To our knowledge, this is the first literature review of aortic chordae tendineae strands, including diagnosis, management, and mechanisms of aortic regurgitation due to such informal strands.

Key words: Aortic valve, Aortic valve replacement, Echocardiography, Abnormal fibrous strand

CASE REPORT

The aortic chordae tendineae strands, also referred to as “fibrous strands” in the literature, are suggested to be embryonic remnants of the cusp formation process. These strands are a rare cause of aortic regurgitation (AR) and have been previously described on both the bicuspid and tricuspid aortic valves. We herein report a case involving a patient with severe AR caused by rupture of an aortic chordae tendineae strand. We also present a review of the related literature. Three-dimensional TEE revealed another fibrous strand anchored between the free rim of the non-coronary cusp and the sinus of Valsalva. To our knowledge, this is the first literature review of aortic chordae tendineae strands to include the diagnosis, management, and mechanisms of AR caused by such strands.

Case Report
A 70-year-old male was admitted to our hospital for shortness of breath and chest tightness. Physical examination revealed a diastolic murmur without obvious wet or dry rales. An electrocardiogram was normal. No fever or obvious inflammatory syndrome was present. Transthoracic echocardiography in our hospital suggested severe AR with suspected mobile linear echo and mild dilatation of the aortic root (diameter at sinus of Valsalva: 41 mm) with a normal annulus diameter (19 mm). Further TEE (FigureA-C) revealed commissural detachment between the right coronary and non-coronary cusp of the aortic valve and severe eccentric AR due to right coronary cusp prolapse with a mobile fibrous strand connected to the free edge of the right coronary cusp. In addition, three-dimensional TEE showed another fibrous strand anchored between the free rim of the non-coronary cusp and the sinus of Valsalva.

Because of the symptomatic severe AR, the patient underwent surgical replacement of the aortic valve. Intraoperative examination revealed an enlarged left ventricle and a normal aortic annulus. A ruptured fibrous strand was found on the free edge of the right coronary cusp, and another fibrous strand was found between the free rim of the non-coronary cusp and Valsalva aortic root. The postoperative pathological examination showed fibrous tissue hyperplasia with myxomatous degeneration of the aortic valves and no pathological findings of infective endocarditis (FigureD).

Discussion
The aortic chordae tendineae strands, also referred to as “fibrous strands” in the literature, are thought to be embryonic remnants of the cusp formation process at an early stage of aortic valve development. Several cases have been reported in the English-language literature. Fifteen case reports of aortic chordae tendineae strands were published from 1984 to 2019, including the present case (Table). The patients comprised 12 men and 3 women ranging from 10 to 76 years (median, 56 years) of age. The location of these fibrous strands was averagely distributed among the three cusps, and ruptured strands accounted for 60% of cases (9/15). These fibrous strands of aortic valve tissue have been previously reported in Asian
countries, especially in Japan (10/15). They are a rare cause of AR, and they cause AR by two main mechanisms: strand rupture between the aortic cusp and the aortic wall and restricted closure of the aortic valves by the strands. To the best of our knowledge, most reported cases of aortic chordae tendineae strands were associated with the first mechanism; fewer than five cases were associated with the latter mechanism.2,7,10) The present case might be an example of involvement of both of these mechanisms. These fibrous strands are supportive tissues that maintain aortic valve coaptation. If balance is effectively maintained by the strands, no valve dysfunction with significant regurgitation will occur. Conversely, ruptured strands may lead to poor valve coaptation. In the present case, one of the two strands ruptured, causing severe AR because of commissural detachment between the right coronary and non-coronary cusps of the aortic valve. Coaptation of the aortic valve can be prevented by dilation of the ascending aorta because attachment of fibrous strands restricts closure of the aortic valves. In the present patient, the diameter of the sinus of Valsalva was enlarged to 41 mm, and the maximal diameter of the proximal ascending aorta was 36 mm. Our review of the literature indicated that most cases of AR caused by congenital aortic chordae tendineae strands occur in patients of advanced age. Rupture of fibrous strands can be caused by dilation of the ascending aorta and valve degeneration secondary to growth and aging.12) In addition, older people often have hypertension, and a spontaneous increase in blood pressure may cause rupture of aortic chordae tendineae strands.19)

In terms of diagnosis, patients with unexplained AR must be carefully evaluated for fibrous strands, which can be more easily detected by TEE than transthoracic echocardiography. In addition, three-dimensional TEE plays an important supplemental role in the diagnosis.15) However, fibrous strands should be differentiated from Lambl's excrescences and infectious and noninfectious neoplasms when abnormal lines are detected at the aortic valves. First, Lambl's excrescences16) are slender, filiform, or papillary structures located on the peripheries of valves and are always found in patients of advanced age. They are more commonly seen on the mitral valve than on the aortic valve, typically near the closure line of the mitral valve. They are mostly diagnosed using TEE as a potential etiological factor of stroke. Pathological examination shows a collagen fiber core covering a monolayer of endothelial cells.17) However, valve dysfunction is rarely caused by Lambl's excrescences, such as significant regurgitation. Therefore, our case does not support this diagnosis. Second, an infectious neoplasm18) refers to inflammation caused by bacteria, fungi, or other microorganisms. The basic pathological change in the heart valve is vegetation composed of platelets, fibrin, red blood cells, white blood cells, and pathogens. Echocardiography also plays an important role in the diagnosis, showing excrescence with rough edges and shapes on one or more leaflets. However, according to the modified Duke criteria,19) infectious neoplasm is not an initial consideration in the accepted criteria for diagnosis of infective endocarditis. Finally, noninfectious neoplasms20) should also be differentiated from chordate tendineae strands, which are commonly present in patients with rheumatic endocarditis, antiphospholipid syndrome, extramedullary proliferative disease, and solid tumors. In patients with autoimmune disor-
complicated reoperation. The main disadvantages are a need for chronic anticoagulation, preservation of a near placement of low thromboembolic risk, which avoids the necessity of aortic valve replacement. Aortic valve repair is an option only at centers with suitable anatomy (aortic dilation without aortic root remodeling).

According to the AHA/ACC valve guideline, most patients with chronic severe AR will require aortic valve replacement. Aortic valve repair is an option only at centers with established expertise and experience and for patients with suitable anatomy (aortic dilation without a thickened, deformed, or calcified valve). Therefore, surgical aortic valve replacement is the main treatment for AR caused by aortic chordae tendineae strands. Aortic valve repair has major advantages compared to aortic valve replacement of low thromboembolic risk, which avoids the need for chronic anticoagulation, preservation of a near normal aortic valve orifice, and the potential for a less complicated reoperation. The main disadvantages are a higher rate of reoperation and less widespread surgical experience. Valve repair has more often been performed in young patients. Because of elderly age of the patient, the mortality rate of reoperation is also relatively higher. Aortic valve replacement may be more beneficial to our patients and can be performed safely with low rates of aortic valve reoperation and acceptable rates of recurrent aortic insufficiency, further research is required to evaluate the clinical efficiency and related complications.

**Conclusion**

The aortic chordae tendineae strands are suggested to be embryonic remnants of the cusp formation process. They are a rare cause of AR and are mostly observed in advanced-age patients with bicuspid aortic valves. TEE has an important role in the diagnosis of aortic chordae tendineae strands, and three-dimensional TEE plays a supplemental role in the diagnosis. However, these abnormal strands should be differentiated from Lamb’s excrescences and infectious and noninfectious neoplasms. At present, the treatment of aortic chordae tendineae strands with significant AR mainly involves aortic valve replacement. Although aortic valve repair is feasible in such patients, further research is required to evaluate the clinical efficacy and related complications.

**Acknowledgment**

We thank Angela Morben, DVM, ELS, from Liwen Bianji, Edanz Editing China (www.liwenbianji.cn/ac), for their excellent linguistic assistance during the preparation of this manuscript.
editing the English text of a draft of this manuscript.

Disclosure

Conflicts of interest: The authors declare that there is no conflict of interest regarding the publication of this manuscript.

Authors’ contributions: SY drafted the manuscript and conceived the study. RM searched relevant references. XS edited the images. YM contributed to the development of the methodology. All authors read and approved the final manuscript.

Consent for publication: Written informed consent of clinical detail and image publication was obtained from the patient.

References

1. Hashimoto R, Miyamura H, Eguchi S. Congenital aortic regurgitation in a child with a tricuspid non-stenotic aortic valve. Br Heart J 1984; 51: 358-60.
2. Yavuz S, Türk T, Celkan MA, Koca V, Ata Y, Ozdemir IA. Congenital aortic insufficiency due to aortic cusp stretching: ‘kite anomaly’. J Heart Valve Dis 1999; 8: 284-6.
3. Nakajima M, Tsuchiya K, Naito Y, Hibino N, Inoue H. Aortic regurgitation caused by rupture of a well-balanced fibrous strand suspending a degenerative tricuspid aortic valve. J Thorac Cardiovasc Surg 2002; 124: 843-4.
4. Mishima T, Yamamoto K, Sugimoto T, Sakakibara K, Uehara A, Yoshii S. Severe aortic regurgitation resulting from a downward displacement of anterior aortic annulus and fibrous strands in the bicuspid aortic valve. Ann Thorac Cardiovasc Surg 2010; 16: 57-9.
5. Minami H, Asada T, Gan K, Yamada A, Sato M. Aortic regurgitation caused by rupture of the abnormal fibrous band between the aortic valve and aortic wall. Gen Thorac Cardiovasc Surg 2011; 59: 488-90.
6. Akasaka K, Saito E, Higuchi T, et al. Aortic regurgitation caused by fibrous strand rupture in a fenestrated aortic valve. J Echocardiogr 2012; 10: 151-3.
7. Bouchachi AA, Folliguet T, Hebert JL, et al. A severe restrictive aortic regurgitation resulting from valve tenting by unusual aortic chordae tendineae strands. Circulation 2012; 126: e139-41.
8. Ishige A, Uejima T, Kannatsue K, Endo M. Giant fenestration and fibrous strand rupture of aortic valve without massive regurgitation. J Cardiol 2012; 5: e163-5.
9. Irisawa Y, Ilatani K, Kitamura T, et al. Aortic regurgitation due to fibrous strand rupture in the fenestrated left coronary cusp of the tricuspid aortic valve. Int Heart J 2014; 55: 550-1.
10. Esteve-Ruiz I, López-Pardo F, Lagos-Degrande Ó, López-Haldón JE, Urbano-Moral JA. Severe restrictive aortic regurgitation due to aortic fibrous strand. Eur Heart J Cardiovasc Imaging 2015; 16: 465.
11. Abdelaziz MM, Martinelli G, Luckraz H. Chordae tendineae of the aortic valve. J Card Surg 2016; 31: 328-9.
12. Ogawa S, Sawada K, Goto Y, Koyama Y, Okawa Y. Abnormal mobile structure in the aortic valve due to fibrous strand rupture. Asian Cardiovasc Thorac Ann 2017; 25: 304-6.
13. Matsukuma S, Inoue T, Tanigawa K, et al. Valve-sparing aortic root re-implantation for commissional detachment with fibrous strand. Gen Thorac Cardiovasc Surg 2018; 66: 54-6.
14. Nishida H, Suenaga E, Ishii K. Acute aortic regurgitation following fibrous strand rupture of aortic valve successfully diagnosed by transesophageal echocardiography. Echocardiography 2018; 35: 753-4.
15. Karia N, Adlam D, Dhanookchand R, Sabharwal N. Three-dimensional transesophageal echocardiography of an aberrant chordae tendineae causing aortic valvular regurgitation. Eur Heart J 2011; 32: 1038.
16. Chu A, Aung TT, Sahalon H, Choksi V, Feiz H. Lambli’s excescence associated with cryptogenic stroke: A case report and literature review. Am J Case Rep 2015; 16: 876-81.
17. Nakahira J, Sawai T, Minami T. Pathologic examination of Lambli’s excescence. J Cardiothorac Vasc Anesth 2014; 28: e3-4.
18. Baddour LM, Wilson WR, Bayer AS, et al. Infective Endocarditis in Adults: Diagnosis, Antimicrobial Therapy, and Management of Complications: A Scientific Statement for Healthcare Professionals from the American Heart Association. Circulation 2015; 132: 1435-86.
19. Durack DT, Lukes AS, Bright DK. New criteria for diagnosis of infective endocarditis: Utilization of specific echocardiographic findings. Duke Endocarditis Service. Am J Med 1994; 96: 200-9.
20. Reinsner SA, Brenner B, Hain M, Edoute Y, Markiewicz W. Echocardiography in nonbacterial thrombotic endocarditis: From autopsy to clinical entity. J Am Soc Echocardiogr 2000; 13: 876-81.
21. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Thorac Cardiovasc Surg 2014; 148: e1-132.
22. Wong CHM, Chan JSK, Sanli D, Rahimt R, Harky A. Aortic valve repair or replacement in patients with aortic regurgitation: A systematic review and meta-analysis. J Card Surg 2019; 34: 377-84.
23. Boodhwani M, de Kerchove L, Watremez C, et al. Assessment and repair of aortic valve cusp prolapse: Implications for valve-sparing procedures. J Thorac Cardiovasc Surg 2011; 141: 917-25.