A rare cause of severe mitral regurgitation: mitral valve aneurysm

İleri mitral yetersizliğinin nadir bir nedeni: Mitral kapağı anevrizması

Ahmet Güler, M.D., Suzan Hatipoğlu, M.D., Can Yücel Karabay, M.D., Cevat Kırma, M.D.

Department of Cardiology, Kartal Koşuyolu Heart and Research Hospital, İstanbul

Summary – Perforation of a mitral valve aneurysm is a rare cause of acute mitral regurgitation, and valvular aneurysm formation and its rupture without infectious involvement are unusual. An 80-year-old man was admitted with acute onset and progressive dyspnea. He had no history of chest pain, palpitation, or fever. Laboratory findings did not suggest any signs of infection. Transthoracic echocardiography revealed an aneurysm of the mitral septal leaflet protruding into the left atrium during systole and color-flow Doppler ultrasonography showed severe mitral regurgitation. There was no aortic regurgitation nor evidence for rheumatic involvement of the valvular structures. Pulmonary artery systolic pressure estimated from the tricuspid regurgitation jet was 50 mmHg. Transesophageal echocardiography showed a saccular, thin-walled, mitral valve aneurysm on the atrial surface, expanding during systole and a small tissue defect on the aneurysmatic segment of the mitral leaflet. There were no signs of connective tissue disease. The patient was submitted to surgery. The aneurysmatic and perforated parts on the septal leaflet were resected and an annuloplasty ring was placed. The histopathological examination of the mitral valve tissue showed nonspecific degenerative changes. The postoperative period was uneventful and the patient was discharged on the fifth postoperative day.

Özet – Mitral kapağı anevrizmasının yırtılması akut mitral yetersizliğinin nadir bir nedenidir ve enfektif endokarditle ilişkili olmayan kapak anevrizmalarının gelişmesi ve yırtılmaları sıra dışı bir durumdur. Seksen yaşında erkek hasta, akut başlangıçlı ve ilerleyici nefes darlığı yakınmasıyla yatırıldı. Göğüs ağrısı, çarpıntı veya ateş öyküsü olmayan hastanın laboratuvar incelemelerinde enfeksiyonu akla getiren bir bulgu yoktu. Transtorasik ekokardiyografide, mitral septal yaprakçıkta, sistolde sol atriyuma doğru uzanan bir anevrizma görüldü. Renkli akım Doppler incelemesinde ise ileri derecede mitral yetersizliği izlendi. Aort yetersizliğine ya da kapak yapılarında romatizmal tutuluma rastlanmadı. Triküspit kaçağı jetinden hesaplanan pulmoner arter sistolik basıncı 50 mmHg idi. Transözofageal ekokardiyografi incelemesinde, mitral kapağın atriyal yüzeyinde, sakküler, ince duvarlı ve sistolde genişleyen bir anevrizma ve mitral yaprakçığın anevrizmatik segmenti üzerinde küçük doku defekti görüldü. Bağ dokusu hastalığı bulgusu yoktu. Hasta cerrahiye sevk edilerek, septal yaprakçıktaki anevrizmatik ve yırtık parçalar temizlendi ve anüloplasti halkası yerleştirildi. Mitral kapak dokusunun histopatolojik inceleme sonucu spesifik olmayan dejeneratif değişiklikler olarak bildirildi. Ameliyat sonrası dönemi sorunsuz geçiren hasta beşinci günde taburcu edildi.

Perforation of a mitral valve aneurysm is a rare cause of acute mitral regurgitation, and it is most commonly associated with aortic regurgitant jet in the setting of infective endocarditis. Thus, aneurysm formation and its rupture without infectious involvement are unusual.

We present a patient with a ruptured mitral valve aneurysm without evidence for previous or active endocarditis.

CASE REPORT

An 80-year-old man was admitted to our hospital with acute onset and progressive dyspnea of one-week duration. There was a 4/6 systolic murmur at the apex. The electrocardiogram showed normal sinus rhythm (during hospitalization atrial fibrillation attacks were observed). He had no history of chest pain, palpitation, or fever. Laboratory findings did not suggest any signs

Received: May 24, 2011 *Accepted:* September 27, 2011

Correspondence: Dr. Ahmet Güler. Kartal Koşuyolu Yüksek İhtisas Eğitim ve Araştırma Hastanesi, Kardiyoloji Kliniği, Denizer Cad., Cevizli Kavşağı, 34846 İstanbul, Turkey. Tel: +90 216 - 459 40 41 e-mail: ahmetguler01@yahoo.com.tr

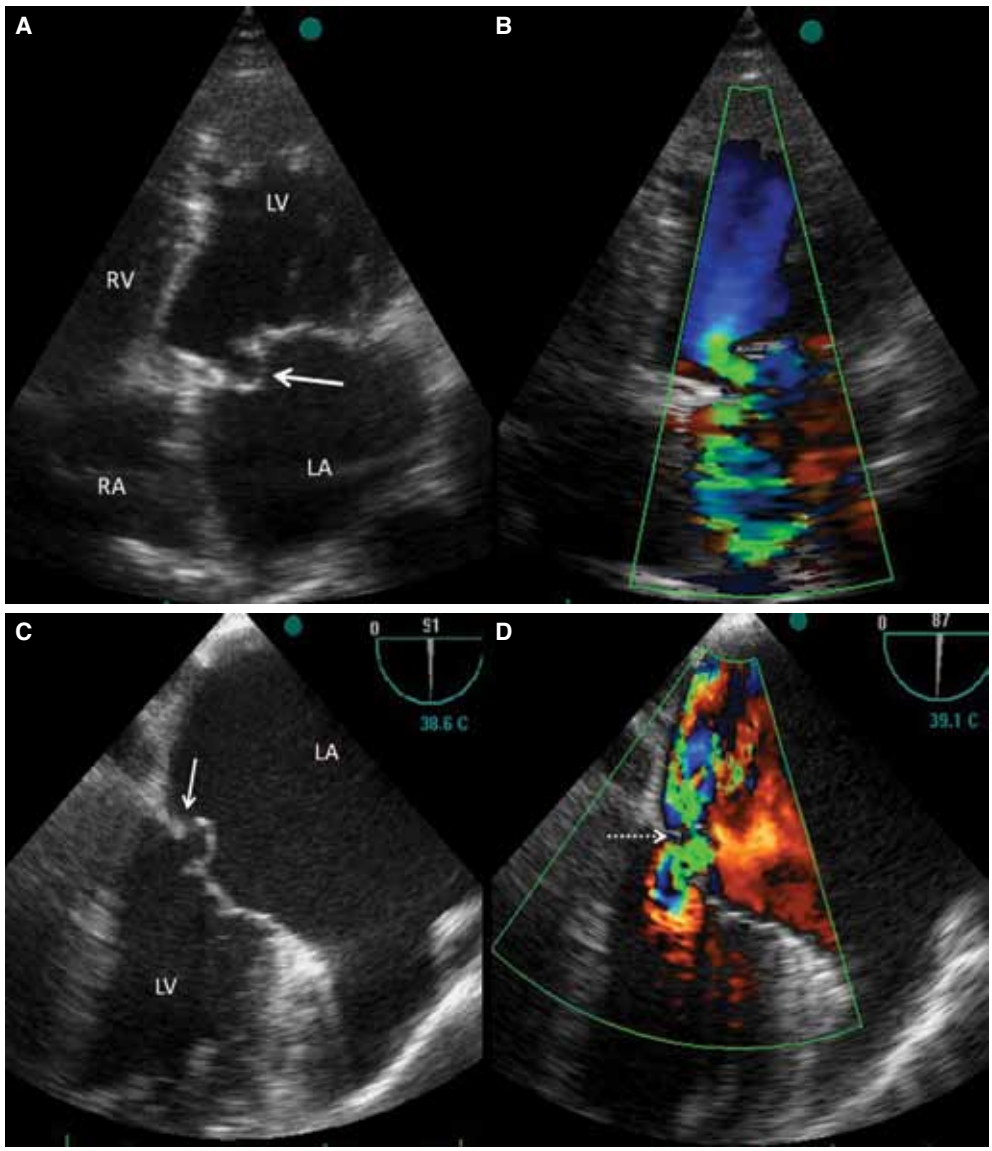

Figure 1. **(A)** Transthoracic apical 4-chamber view shows aneurysmatic mitral septal leaflet (arrow) and **(B)** color-flow Doppler image shows severe mitral regurgitation with an anterior eccentric jet. **(C)** Transesophageal 2-chamber view demonstrates a thin, saccular, perforated aneurysm of the mitral septal leaflet (arrow) and **(D)** color-flow Doppler image shows a turbulent regurgitant jet (dotted arrow) through the perforated aneurysm. RA: Right atrium; RV: Right ventricle; LA: Left atrium; LV: Left ventricle.

of infection. Transthoracic echocardiography revealed an aneurysm of the mitral septal leaflet protruding into the left atrium during ventricular systole and color-flow Doppler ultrasonography showed severe mitral regurgitation (Fig. 1a, b). The size and function of the left ventricle were normal, aortic and mitral valves were degenerative, the left atrium was enlarged, and there was no aortic regurgitation nor evidence for rheumatic involvement of the valvular structures. Pulmonary artery systolic pressure calculated from the tricuspid regurgitation jet was estimated to be 50 mmHg. Transesophageal echocardiography was performed for better delineation of the aneurysmal structure and for further assessment of the severity of mitral regurgitation. There was no vegetation on the mitral or aortic leaflets; however, a saccular, thin-walled, regularly contoured formation consistent with a mitral valve aneurysm was seen on the atrial surface of the A2 scallop, expanding during systole and decompressing during diastole. A small tissue defect was also visible on the aneurysmatic segment of the mitral leaflet. A turbulent jet through the aneurysm

resulted in severe eccentric mitral regurgitation (Fig. 1c, d). Additionally, there was no echocardiographic evidence for aortic root involvement suggestive of unrecognized connective tissue disease, and his history, physical examination, and laboratory findings did not provide any signs of connective tissue disease.

Surgery was scheduled for symptomatic severe mitral regurgitation and echocardiographic findings were confirmed by surgical exploration. Following the resection of the aneurysmatic and perforated part on the septal leaflet corresponding to the A2 scallop, an annuloplasty ring was placed. Intraoperative and postoperative echocardiographic examinations showed mild mitral regurgitation. The histopathological examination of the mitral valve tissue showed nonspecific degenerative changes. The postoperative period was uneventful and the patient was discharged on the fifth postoperative day.

DISCUSSION

Mitral valve aneurysms frequently accompany infective endocarditis. Due to the frequent association with aortic valve lesions, aortic regurgitation has been suggested to cause mitral valve aneurysms.[1,2] Several cases of mitral valve aneurysm not associated with infective endocarditis were reported, but these cases usually had connective tissue disorders including mitral valve prolapse, osteogenesis imperfecta, Marfan syndrome, pseudoxanthoma elasticum, or physical stress due to severe aortic regurgitation.[3-6] In our patient, none of these etiologies were identified. We thought that degenerative changes that occurred in the mitral valve with aging were responsible for the aneurysm formation and its perforation. The calcific nature of the mitral and aortic valves was apparent in our case and there was no other cardiac or systemic abnormality to explain the formation of the aneurysm.

The differential diagnosis of mitral valve aneurysms includes mitral valve prolapse, myxomatous degeneration of the mitral valve, flail mitral leaflet, papillary fibroelastoma, myxoma involving the mitral valve, and mitral valve cysts without endothelization. In this case, mitral valve aneurysm and severe mitral regurgitation were detected by transthoracic echocardiography; however, transesophageal echocardiography served as a valuable complement for the identification of a turbulent jet through the ruptured part of the aneurysm as the main mechanism of mitral regurgitation and selection of mitral valve repair as the surgical strategy. Furthermore, three-dimensional transesophageal echocardiography and cardiac magnetic resonance imaging may also provide valuable information in the evaluation of valves and valvular apparatus.

Recognition of this rare entity as the cause of mitral regurgitation will guide the surgeon in planning the operation and during the procedure. Early detection and surgical removal of these perforated aneurysms with subsequent repair of the mitral valve will prevent complications such as emboli or endocarditis.[7]

Conflict-of-interest issues regarding the authorship or article: None declared

REFERENCES

1. Gülmez Ö, Sade LE, Yıldırır A, Müderrisoğlu H. Mitral valve aneurysm associated with aortic valve regurgitation. Türk Kardiyol Dern Arş 2009;37:263-5.
2. Reid CL, Chandraratna AN, Harrison E, Kawanishi DT, Chandrasoma P, Nimalasuriya A, et al. Mitral valve aneurysm: clinical features, echocardiographic-pathologic correlations. J Am Coll Cardiol 1983;2:460-4.
3. Rückel A, Erbel R, Henkel B, Krämer G, Meyer J. Mitral valve aneurysm revealed by cross-sectional echocardiography in a patient with mitral valve prolapse. Int J Cardiol 1984;6:633-7.
4. Edynak GM, Rawson AJ. Ruptured aneurysm of the mitral valve in a Marfan-like syndrome. Am J Cardiol 1963;11:674-7.
5. Lebwohl MG, Distefano D, Prioleau PG, Uram M, Yannuzzi LA, Fleischmajer R. Pseudoxanthoma elasticum and mitral-valve prolapse. N Engl J Med 1982;307:228-31.
6. Özeke Ö, İlkay E. Cyst-like anterior mitral valve aneurysm. Türk Kardiyol Dern Arş 2009;37:359.
7. Vilacosta I, San Román JA, Sarriá C, Iturralde E, Graupner C, Batlle E, et al. Clinical, anatomic, and echocardiographic characteristics of aneurysms of the mitral valve. Am J Cardiol 1999;84:110-3, A9.

Key words: Echocardiography, transesophageal; heart aneurysm/surgery; mitral valve insufficiency/etiology.

Anahtar sözcükler: Ekokardiyografi, transözofageal; kalp anevrizması/cerrahi; mitral kapağı yetersizliği/etyoloji.