Ascending aortic aneurysm and supravalvular aortic membrane in a case with Marfan's syndrome and Hodgkin lymphoma diagnosed in the postoperative period

Ameliyat sonrası dönemde Hodgkin lenfoma tanısı konulan Marfan sendromlu bir olguda çıkan aort anevrizması ve supravalvüler aortik membran

Azman Ateş,[1] Bilgehan Erkut,[1] Yekta Gürlertop,[2] İbrahim Yekeler[3]

Departments of [1]Cardiovascular Surgery, [2]Cardiology, Medicine Faculty of Atatürk University, Erzurum; [3]Department of Cardiovascular Surgery, Siyami Ersek Training and Research Hospital, İstanbul

The presence of Hodgkin's lymphomas concurrently with cardiovascular diseases is generally a rare condition. In this article we present a 26-years-old male case with Marfan's syndrome in whom we detected an aneurysm of ascending aorta, and supraaortic valvular membrane and Hodgkin lymphoma that was diagnosed in the postoperative period. Echocardiographic examination of the patient revealed the presence of an ascending aortic aneurysm and a supravalvular aortic ring. A computed tomography of the chest revealed the presence of an aneurysm of approximately 9 cm in the ascending aorta. Emergency ascending aortic and root replacement was performed on the patient using the modified flanged Bentall technique. No problems were detected during the postoperative monitoring. Due to the presence of supraclavicular lymphadenopathy, lymphoid tissue biopsy was taken from supraclavicular region and the patient was diagnosed with nodular sclerosing Hodgkin's lymphoma. The patient has been receiving chemotherapy for three-years since the surgery and in the three-year postoperative evaluation, he had no problems or symptoms.

Key words: Ascending aortic aneurysm; Hodgkin's lymphoma; supravalvular aortic membrane.

Kardiyovasküler hastalıklarla birlikte Hodgkin lenfoma varlığı genellikle nadir görülen bir durumdur. Bu yazıda çıkan aort anevrizması ve supravalvüler aortik membran tespit ettiğimiz Marfan sendromlu ve ameliyat sonrası dönemde Hodgkin lenfoma tanısı konulan 26 yaşındaki bir erkek olgu sunuldu. Hastaya yapılan ekokardiyografi incelemesinde bir çıkan aort anevrizması ve supravalvüler aortik ring varlığı saptandı. Bilgisayarlı tomografide çıkan aortta yaklaşık 9 cm'lik bir anevrizma tespit edildi. Hastaya etekli modifiye Bentall tekniği kullanılarak acil çıkan aort ve kök replasmanı yapıldı. Ameliyat sonrası izlem süresince herhangi bir sorun saptanmadı. Supraklaviküler lenfadenopati varlığı nedeniyle supraklaviküler bölgeden lenfoid doku biyopsisi alındı ve hastaya nodüler sklerozan tip Hodgkin lenfoma tanısı konuldu. Hasta üç yıldır kemoterapi görmektedir ve ameliyattan üç yıl sonra yapılan değerlendirmede herhangi bir sorunu ve hastalık belirtisi olmadığı görüldü.

Anahtar sözcükler: Çıkan aort anevrizması; Hodgkin lenfoma; supravalvüler aortik membran.

Although supravalvular aortic stenosis is an uncommon congenital cardiac anomaly characterized by varying degrees of left ventricular outflow tract obstruction and congenital narrowing of the ascending aorta above the level of the coronary arteries,[1] our case had neither left ventricular outflow pathology nor narrowing of the ascending aorta. This patient with Marfan's syndrome (MS) only had a dilated ascending aorta above the level of the coronary arteries and supravalvular membrane. The patient was operated by performing the Bentall procedure. Postoperatively, Hodgkin lymphoma (HL) was diagnosed and he received chemotherapy for two-years.

We report this extremely rare pathological combination, which has not been reported previously and suggest that the flanged technique may be suitable in the treatment of an ascending aorta aneurysm in a patient with MS.

Received: November 8, 2006 *Accepted:* February 9, 2007

Correspondence: Bilgehan Erkut, M.D. Atatürk Üniversitesi Tıp Fakültesi Kalp ve Damar Cerrahisi Anabilim Dalı, 25240 Erzurum, Turkey.
Tel: +90 442 - 316 63 33 e-mail: bilgehanerkut@yahoo.com

CASE REPORT

A 26-year-old male patient with Marfanoid manifestations was admitted to our clinic for chest and back pain. Widened pulse pressures, bounding pulses, and a parasternal systolo-diastolic murmur were determined on physical examination. There was a right supraclavicular mass (3x4 cm). He was in functional class 2-3 according to New York Heart Association.

Telecardiogram showed mediastinal enlargement. Echocardiography revealed ascending aortic aneurysm (approximate 9 cm) combined with severe aortic regurgitation, and there was a circumferential membrane 1 cm above the coronary ostia on the intimal face of the ascending aorta (Fig. 1a). Computed tomography scan demonstrated that the aortic sinuses and ascending aorta were widened up to approximately 9-10 cm.

The patient was taken to the operating room for emergency ascending aorta and root replacement. After median sternotomy, extracorporeal circulation was established through the left femoral artery and the right atrium with cannulation of the superior and inferior vena cavae. Cold oxygenated hyperkalemic blood cardioplegia was used for myocardial protection. Cardioplegic solution was administered by means of direct cannulation of the coronary ostium. The ascending aorta was very large (Fig. 1b) and cross-clamped at the origin of the innominate artery without total circulatory arrest and the ascending aorta was incised longitudinally distally 2 cm from the cross-clamp. The supraaortic vessels were not involved.

The membrane was resected above the coronary sinuses (Fig. 2). Because of the large aortic root and clear aortic regurgitation, we decided to perform root and ascending aortic replacement. Therefore, we used the flanged technique which was described by Yakut[2] We used a 28 mm Dacron vascular graft (Sulzek Vascutek, Scotland) and bileaflet no. 25 prosthetic valve (Sorin Biomedica, ART25LNF, Italy) to build a flanged composite graft. After the surgical procedure, extracorporeal circulation was weaned smoothly. Total cardiopulmonary bypass time was 255 minutes, aortic cross-clamp time was 57 minutes. The patient was extubated on the first postoperative day and the postoperative course was uneventful. The patient remained in the intensive care unit for 52 hours after surgery.

On the 5th postoperative day, a supraclavicular lymph node biopsy was performed on the supraclavicular mass. The specimens measuring 4x2x1 cm were examined in paraffin sections stained with either hematoxylin and eosin or Wright-Giemsa. Histologically, they showed effacement of their primitive architecture, which was substituted by lymphoid nodules, partly bordered by collagen bands and composed of mature lymphocytes intermingled with eosinophils, histiocytes, and lacunar-type Reed-Sternberg (RS) cells. Histopathologic examination was interpreted as nodular sclerotic type Hodgkin lymphoma (Fig. 3).

The patient was discharged on the 20th postoperative day. At the patient's two-year postoperative evaluation, he had fully recovered and was asymptomatic. He is still undergoing chemotherapy for HL.

DISCUSSION

The most important manifestations of this case with MS in "classic HL" are its combination with ascending aortic aneurysm, and supraaortic membrane. The MS is characterised by highly variable clinical manifestations in primarily skeletal, ocular and cardiovascular organ systems. The most common cardiovascular

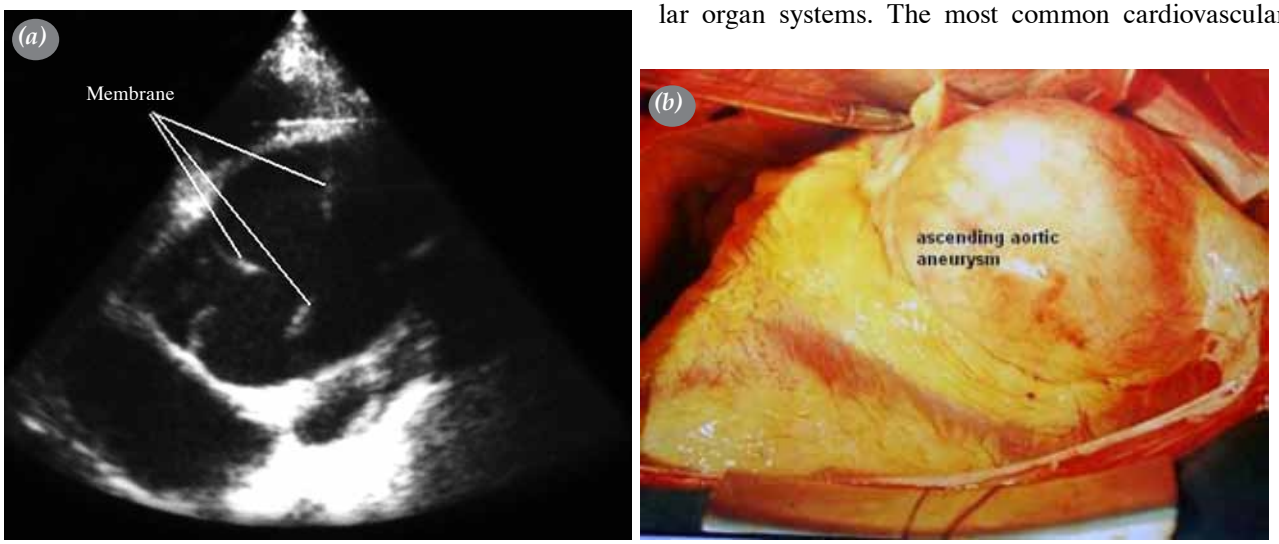

Fig. 1. (a) Transthoracic echocardiography view depicts the membrane in the ascending aorta. **(b)** The preoperative view shows dilated ascending aorta.

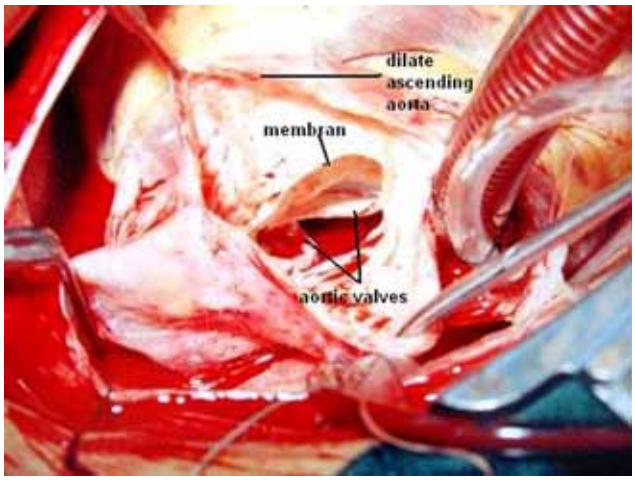

Fig. 2. The picture obtained during surgery shows the normal aortic valve leaflets and supravalvular membrane.

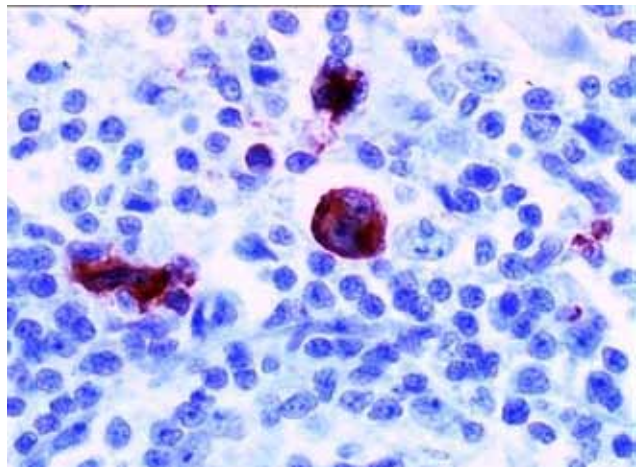

Fig. 3. Detail of the supraclavicular lymph node specimen with an occasional Reed-Sternberg cell noted among lymphoid cells (H-E x 250).

manifestations of MS include mitral valve prolapse and regurgitation, but aortic dilatation, especially of the aortic root, is the most common cause of morbidity and mortality.[3] Prognosis is mainly determined by progressive dilatation of the aorta, potentially leading to aortic dissection and death at a young age.[4]

Early identification of patients with MS is therefore of considerable importance. Without surgery most patients with MS die in the third decade of their lives from complications of aortic root aneurysm, such as aortic rupture, aortic dissection, and aortic insufficiency. Aortic root replacement dramatically improves the survival of these patients. In a recent report by Gott et al.[5] on the results of aortic root replacement in 271 patients with MS, there was no operative mortality among 235 patients operated on electively, and operative mortality was only 5.6% among 36 patients operated on urgently.

Over the past 30 years, composite valve grafting has become a low risk operation and a very durable one for MS. For those patients wishing to avoid anticoagulation therapy two types of valve-sparing operations have been introduced in the early 1990s: reimplantation and remodelling of the aortic root.[6]

The results of aortic valve-sparing operations in patients with MS have been excellent during the first decade. The long-term results of valve-sparing aortic root replacement, and the overall incidence of all valve-related and aorta-related complications in large numbers of patients with MS are still unknown. Because our patients had severely aortic insufficiency, we performed aortic root replacement with flanged technique without valve-sparing treatment.

Supravalvular aortic membrane is a rare congenital anomaly and it can occur alone or in association with other cardiac anomalies. Cardiac pathologies like coronary artery aneurysm, intracardiac defects or other types of left ventricular outflow tract obstructions can be observed.[1] Our patient had a supravalvular membrane, but he had no ascending aortic narrowing and outflow tract obstruction. It is possible that aortic root enlargement might have occurred due to both the supravalvular aortic membrane and the ascending aortic aneurysm associated with MS.

Several diseases are described in association with HL. There have been also some reports of simultaneous occurrence of HL and pulmonary tuberculosis, multiple myeloma, including one of a case with eosinophilic granuloma and after transplantation. We encountered no report of simultaneous cardiac disease and malignant lymphoma in the literature. To our knowledge no cases with ascending aortic aneurysm, supravalvular aortic membrane, MS and HL have been reported. In addition, known and unknown factors could have a role in the pathogenesis of several processes. Radiation can affect all the structures in the heart, including the pericardium, the myocardium, the valves and the conduction system. Radiation therapy and cancer chemotherapy are associated with a wide range of vascular toxicities, which may be related to endothelial cell damage by these agents. Although some patients have developed aortic dissection after HL therapy,[7] in our case there was no prior history of irradiation or chemotherapy.

Finally, we consider that the flanged technique is feasible for treatment of MS. Although the ascending aortic dilatation may be associated with Marfan's syndrome and supraaortic membrane, this unusual constellation including four pathologies might be mere coincidence.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Brown JW, Ruzmetov M, Vijay P, Turrentine MW. Surgical repair of congenital supravalvular aortic stenosis in children. Eur J Cardiothorac Surg 2002;21:50-6.
2. Yakut C. A new modified Bentall procedure: the flanged technique. Ann Thorac Surg 2001;71:2050-2.
3. Gürbüz A, Tetik Ö, Lafçı B, Özsöyler İ. Mitral annular plication and valve replacement in a patient with Marfan's syndrome: a case report. T Klin Kalp-Damar Cerrahisi Dergisi 2002;3:174-6.
4. Fabricius AM, Autschbach R, Doll N, Mohr W. Acute aortic dissection during pregnancy. Thorac Cardiovasc Surg 2001;49:56-7.
5. Gott VL, Greene PS, Alejo DE, Cameron DE, Naftel DC, Miller DC, et al. Replacement of the aortic root in patients with Marfan's syndrome. N Engl J Med 1999;340:1307-13.
6. David TE, Feindel CM. An aortic valve-sparing operation for patients with aortic incompetence and aneurysm of the ascending aorta. J Thorac Cardiovasc Surg 1992;103:617-21.
7. Golden MA, Vaughn DJ, Crooks GW, Holland GA, Bavaria JE. Aortic dissection in a patient receiving chemotherapy for Hodgkin's disease-a case report. Angiology 1997;48:1063-5.