Mediastinal lymphoma causing extrinsic pulmonary stenosis

Ekstrensek pulmoner darlığa neden olan mediyastinal lenfoma

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Acquired pulmonary stenosis is rare in adults and may be missed unless a high index of suspicion is present. Extrinsic pulmonic stenosis is even rarer and predominantly caused by external thoracic masses creating non-dynamic obstruction of the right ventricular outflow tract. A 20-year-old female was referred to our center with a cystic mass detected by transthoracic echocardiography and thoracic computed tomography, in the superoanterior mediastinum, 5.5x5.5x7 cm in size, causing main pulmonary trunk compression. Repeat transthoracic echocardiography demonstrated a mass causing compression of the main pulmonary artery immediately after the pulmonary valve. Continuous wave Doppler showed a peak systolic gradient of 65 mmHg and a mean gradient of 37 mmHg in the pulmonary artery at the site of compression. She underwent an open thoracotomy via a midline sternotomy. The mass was firmly attached to the pericardium. Its largest diameter was 15 cm; it surrounded the left phrenic nerve completely and invaded the outer wall of the pulmonary artery and aorta. The mass could only be partly dissected. The pathological diagnosis of the mass was stage IIa nodular sclerosing Hodgkin's lymphoma. The patient received postoperative chemotherapy and thoracal radiotherapy. She was in remission without any cardiac complaint.

Key words: Echocardiography; Hodgkin disease; lymphoma/complications; mediastinal neoplasms/complications/surgery; pulmonary artery/pathology; pulmonary valve stenosis/etiology.

Sonradan gelişen pulmoner darlık erişkinlerde nadir görülen ve şüphe edilmediği taktirde kolaylıkla atlanabilen bir durumdur. Daha da nadir olan ekstrensek pulmoner darlık, sıklıkla göğüs kitlelerinin sağ ventrikül çıkış yolu üzerinde yol açtıkları dinamik olmayan obstrüksiyon nedeniyle ortaya çıkmaktadır. Yirmi yaşında bir kadın hasta, başka bir merkezde transtorasik ekokardiyografi ve göğüs bilgisayarlı tomografisi ile saptanan ve ana pulmoner yapıda basıya yol açan, 5.5x5.5x7 cm büyüklüğünde bir mediyastinal kitle nedeniyle merkezimize sevk edildi. Transtorasik ekokardiyografi ile tekrar incelenen hastada, pulmoner kapağın hemen sonrasında ana pulmoner artere bası yapan bir kitle görüldü. Devamlı dalga Doppler ile incelemede, bası bölgesindeki pulmoner arterde tepe sistolik gradiyent 65 mmHg, ortalama gradiyent 37 mmHg ölçüldü. Hastaya orta hat sternotomi ile açık torakotomi uygulandı. Ameliyatta, en büyük çapı 15 cm ölçülen kitlenin perikarda sıkı bir şekilde tutunduğu, sol frenik siniri tamamen sardığı izlendi; pulmoner arter ve aortun dış duvarında invazyon vardı. Kitle sadece kısmi olarak çıkarılabildi ve histopatolojik tanı evre IIa sklerozan Hodgkin lenfoma olarak kondu. Ameliyat sonrasında kemoterapi ve radyoterapi uygulanan hastada remisyon bulguları görüldü; hastanın kardiyak yakınması yoktu.

Anahtar sözcükler: Ekokardiyografi; Hodgkin hastalığı; lenfoma/komplikasyon; mediastinal neoplaziler/komplikasyon/cerrahi; pulmoner arter/patoloji; pulmoner kapak darlığı/etyoloji.

Acquired pulmonary stenosis is rare in adults and may be missed unless a high index of suspicion is present. Mediastinal malignant neoplasms, mainly of hematogenous and lymphoid origin, may cause compression of cardiac chambers and great vessels. We present a young female with mediastinal Hodgkin's disease causing compression of the main pulmonary artery and a systolic pressure gradient.

CASE REPORT

A twenty-year-old female with complaints of chest pain, dyspnea, palpitations, and easy fatigability was

Received: January 1, 2009 *Accepted:* March 3, 2009

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referred to our hospital in September 2006. She first presented to another center with acute chest pain in July 2006 and was hospitalized. Transthoracic echocardiography obtained at that time showed normal left ventricular systolic performance and a moderate pericardial effusion with fibrillary bands. Cardiac catheterization was not performed. Ibuprofen therapy led to alleviation of symptoms together with regression of the pericardial fluid and the patient was discharged with complete cure. She presented to the same center in September 2006 with a complaint of back pain, of 20-day history, in the interscapular area. She also had complaints of fatigue and palpitations. A repeat transthoracic echocardiogram revealed an increased supravalvular pulmonary gradient of 80 mmHg and a cystic mass adjacent to the pulmonary valve and main pulmonary artery, causing compression of the latter. Thoracic computed tomography confirmed the presence of a cystic mass in the superoanterior mediastinum with an irregular border, measuring 5.5 x 5.5 x 7 cm and causing main pulmonary trunk compression (Fig. 1). Due to the lack of a thoracic surgery unit, the patient was referred to our center.

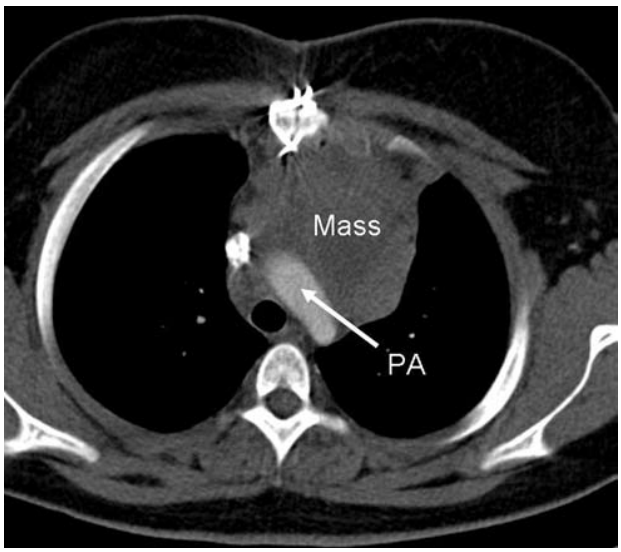

Figure 1. Computed tomography scan of the chest demonstrates an anterior mediastinal mass adjacent to the pulmonary artery (PA).

She was anxious and pale and had continuous back pain. Physical examination showed a pulmonary ejection click, a midsystolic murmur of grade 3/6 in the second left intercostal area, and a pansystolic murmur of grade 2/6 at the lower left sternal border. Her electrocardiogram was unremarkable and chest X-ray showed an enlarged mediastinum. A complete blood count revealed leukocytosis (WBC 17,600/mm^3), anemia (hemoglobin 9.7 g/dl), and a normal thrombocyte count. Erythrocyte sedimentation rate was 72/hr. Blood chemistry was normal. Transthoracic echocardiography demonstrated normal left ventricular and right ventricular systolic performance with normal wall thicknesses and chamber sizes. There were no signs of pericardial or pleural effusion. In the parasternal short-axis view, a mass was detected, causing compression of the main pulmonary artery immediately after the pulmonary valve (Fig. 2a). Continuous wave Doppler examination showed a peak systolic gradient of 65 mmHg and a mean gradient of 37 mmHg in the pulmonary artery at the site of compression (Fig. 2b). Following consultation with the thoracic surgery department, an open thoracotomy via a midline sternotomy was

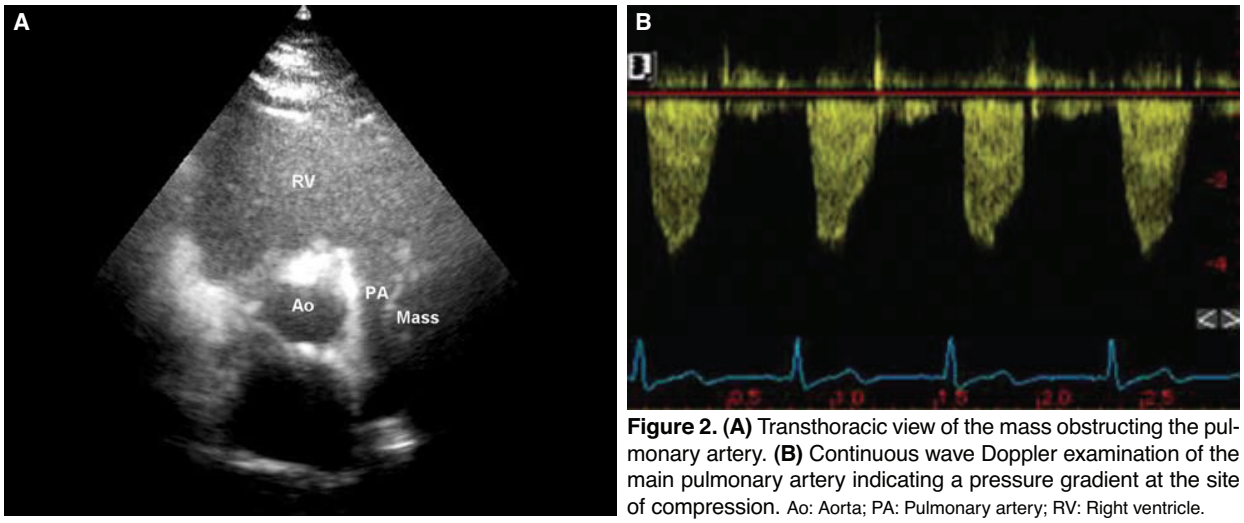

Figure 2. (A) Transthoracic view of the mass obstructing the pulmonary artery. **(B)** Continuous wave Doppler examination of the main pulmonary artery indicating a pressure gradient at the site of compression. Ao: Aorta; PA: Pulmonary artery; RV: Right ventricle.

performed. At surgery, the mass had a solid-cystic structure and was firmly attached to the pericardium. Its largest diameter was 15 cm; it surrounded the left phrenic nerve completely and invaded the outer wall of the pulmonary artery and aorta. A frozen-section analysis suggested the diagnosis of lymphoma. Because of its close association with the great vessels and pericardial sac, the mass could only be partly dissected and the rest was left *in situ*. The pathological diagnosis of the mass was nodular sclerosing Hodgkin's lymphoma. The patient was consulted with the medical oncology unit and a whole-body gallium scintigraphy was obtained, which showed low-intensity pathologic gallium uptake in the lower left cervical, left supraclavicular regions, and in the anterosuperior mediastinum in the aortico-pulmonary projection. Abdominal computed tomography showed no abdominal involvement. The disease was staged as IIa. The patient was discharged on the sixth postoperative day after an uneventful course and was scheduled to receive ABVD chemotherapy plus thoracal radiotherapy in medical oncology and radiation oncology departments on an outpatient basis. After six courses of ABVD chemotherapy, follow-up transthoracic echocardiography showed the right atrial and ventricular chambers in normal size and a 10-mmHg peak systolic pressure gradient across the pulmonary artery. The patient was in remission without any cardiac complaint.

DISCUSSION

Mediastinal Hodgkin's disease may cause compression of the vital organs and structures. Compression of the pulmonary artery and aorta has been reported.[1] Several malignant masses may cause compression of the pulmonary artery, including non-microcytic lung carcinoma,[2] non-Hodgkin's lymphoma,[3,4] thymoma,[5] teratoma,[6] and malignancies of the pericardium[7] or of unknown origin.[6] The most common complaints associated with malignant extrinsic pulmonary compression include chest pain and dyspnea.[8,9] Weight loss, fatigue, cough, and palpitations may also be seen.[9] On physical examination, a systolic ejection murmur is the most frequent physical finding,[9] and less frequently, a precordial lift or palpable thrill may be noted. On electrocardiography, normal sinus rhythm as well as a right axis deviation or right ventricular hypertrophy may be present. Finally, the most common appearance on the chest radiogram is enlargement of the mediastinum, followed by a normal chest radiograph and cardiomegaly.

Tesoro-Tess et al.[10] examined 36 patients with mediastinal lymphoma by chest magnetic resonance imaging and two-dimensional echocardiography. They reported that the most common cardiac finding associated with both Hodgkin's disease and non-Hodgkin's lymphomas was the contiguity of the mass with the parietal pericardium without obliteration of the pericardial contour. The prevalence of great vessel involvement was 22.7% for Hodgkin's disease, and 42.8% for non-Hodgkin lymphomas and other involvements were pericardial effusion, pleural effusion, and pericardial infiltration. They found that magnetic resonance imaging was more successful than transthoracic echocardiography in detecting great vessel involvement and pericardial contiguity.

Echocardiography enables dynamic investigation of cardiac and paracardiac structures and may better visualize the severity of great vessel compression, assessed by Doppler interrogations. Other diagnostic modalities to detect cardiac involvement by lymphomas include computed tomography, transesophageal echocardiography, and pericardial and endomyocardial biopsy. Therapy for low-stage lymphomas is mainly based on radiotherapy; however, constitutional symptoms, massive disease, and extranodal involvement require chemotherapy. Chemotherapy is usually necessary prior to radiotherapy to reduce the irradiation field.

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