Type II aortopulmonary window coexisting with a ventricular septal defect in a 23-year-old man

Yirmi üç yaşındaki erkek olguda tip II aortopulmoner pencere ile birlikte ventriküler septal defekt

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Aortopulmonary window is a rare congenital cardiac anomaly. We herein report a case of an aortopulmonary window coexisting with a ventricular septal defect in a 23-year-old man. He had a history of a systolic murmur detected when he had been seven years old, after which he had been followed-up only with medications. Two-dimensional and Doppler echocardiography showed a type II aortopulmonary window and an inlet type ventricular septal defect. The diagnosis was confirmed by cardiac catheterization and angiography. Surgical correction was performed using the transaortic approach. After a follow-up of 16 months, the patient was in New York Heart Association class I.

Key words: Aorta, thoracic; aortopulmonary septal defect/surgery; cardiopulmonary bypass; heart septal defects, ventricular/surgery; pulmonary artery/abnormalities.

Aortopulmoner pencere nadir bir doğumsal kardiyak anomalisidir. Yirmi-üç yaşındaki bir erkek hastada aortopulmoner pencere ve buna eşlik eden ventriküler septal defekt saptandı. Hastanın öyküsünden, yedi yaşında iken sistolik üfürüm saptandığı ve o tarihten itibaren sadece medikal tedavi ile izlendiği öğrenildi. İkiboyutlu ve Doppler ekokardiyografik incelemede tip II aortopulmoner pencere ve ventriküler septal defekt görüldü. Tanı kardiyak kateterizasyon ve anjiyografi ile doğrulandı. Hastaya transaortik yaklaşımla cerrahi düzeltme uygulandı. On altı aylık izlem sonunda hastanın durumu New York Heart Association sınıf I'e uymaktaydı.

Anahtar sözcükler: Aort, torasik; aortopulmoner septal defekt/cerrahi; kardiyopulmoner bypass; kalp septal defekti, ventriküler/cerrahi; pulmoner arter/anormallik.

Aortopulmonary window (APW) is a communication between the main pulmonary artery and the ascending aorta in the presence of two separate semilunar valves. It results from incomplete fusions of the conotruncal ridges during the fifth week of the fetal development. It is found in 0.2% of patients with congenital heart disease.[1] Severe pulmonary hypertension and heart failure lead to poor outcome if it is not corrected very early in life. Although it is often associated with other cardiac defects, coexistence with a ventricular septal defect (VSD) is rare. The current classification of APW was made by Jacobs et al.[2]

In this report, a case of APW accompanied by a simple VSD is presented.

CASE REPORT

A 23-year-old man was admitted to the hospital for evaluation of a cardiac murmur. He had a history of a systolic murmur detected when he had been seven years old. Because of social and economic factors, he had been followed-up only with medications without any diagnostic intervention until the admission date to our hospital. On physical examination, there was a holosystolic murmur at the left sternal border. A chest roentgenogram showed cardiomegaly with a cardiothoracic ratio of 0.6 and increased pulmonary vascularity. His electrocardiogram showed normal sinus rhythm, biventricular hypertrophy, and nonspecific changes in the ST segment. Two-dimensional and Doppler echocardiography showed a type II APW, an inlet type VSD, and a persistent left superior vena cava draining to the right atrium via an enlarged coronary sinus. Cardiac catheterization was performed to confirm the diagnosis, detect associated lesions, and obtain hemodynamic information. Hemodynamic and oximetric measurements revealed a markedly elevated pulmonary arterial pressure (102/55 mmHg; mean 75 mmHg) with a large pulmonary flow (Qp/Qs=2.94). Pulmonary vas-

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cular resistance was 0.5 (Rp/Rs). Right heart catheterization revealed left-to-right shunting at ventricular and arterial levels. On aortography, the ascending aorta, the main pulmonary trunk, and the pulmonary arteries were simultaneously opacified (Fig. 1a), which confirmed the diagnosis of an APW. No atrial septal defect or patent ductus arteriosus were found. The coronary arteries were in normal position.

The operation was performed via a median sternotomy. Arterial cannulation was performed in the distal ascending aortic artery. Two venous cannulae were placed separately in the persistent left superior vena cava, one for the hypoplastic superior vena cava and one for the inferior vena cava. After cannulation, cardiopulmonary bypass was initiated following occlusion of the right and left main pulmonary arteries with snares. The patient was cooled to 28 °C and a vent was placed in the right superior pulmonary vein. The ascending aorta was clamped cautiously just above the localization of the APW and cold blood cardioplegic solution was injected into the aortic root. A transvertical aortotomy was carried out for exposure of the defect and origins of the coronary arteries. A type II APW defect (15x12 mm) was identified between just above the sinus of Valsalva and the main pulmonary artery. The origins of the coronary arteries were normal in position. The APW defect was closed with Dacron patch using 3-0 polypropylene and interrupted pledgeted sutures. After closure of the APW, the inlet type VSD was repaired by Dacron patch using interrupted pledgeted sutures via an oblique right atriotomy. The anterior aortotomy and right atriotomy were closed with running polypropylene sutures. The heart was de-aired and the patient was warmed and weaned from cardiopulmonary bypass. The postoperative course was uneventful. The patient was discharged on the sixth postoperative day. An aortography obtained in the postoperative second month, showed that both the APW and VSD were successfully closed without any residual leakage (Fig. 1b). After a follow-up of 16 months, the patient was in New York Heart Association class I, and on treatment with an ACE inhibitor and an oral diuretic twice a week.

DISCUSSION

Aortopulmonary window results from failure of septation of the aorticopulmonary trunk during the fifth to eight weeks of development. The more posterior the septum forms the closer the window encroaches on the pulmonary artery (type II) until it finally passes the right pulmonary artery, giving the appearance of emerging from the descending aorta (type III).[3] Aortopulmonary window has also been defined as simple and complex types. Simple APW is associated with insignificant anomalies such as a patent ductus arteriosus, atrial septal defect, or a patent foramen ovale, whereas the complex type is associated with more complex anomalies such as an interrupted aortic arch, transposition of great arteries, Fallot's tetralogy, or anomalous coronary arteries.

Aortopulmonary window is a very uncommon congenital anomaly and there are very few surgical series of more than 20 patients in the literature.[4-6] Our literature search revealed only 30 cases of APW that were accompanied by a VSD, and of them, only four reports consisted of three or more cases.[5-8] At our clinic, we encountered only two patients with an APW among

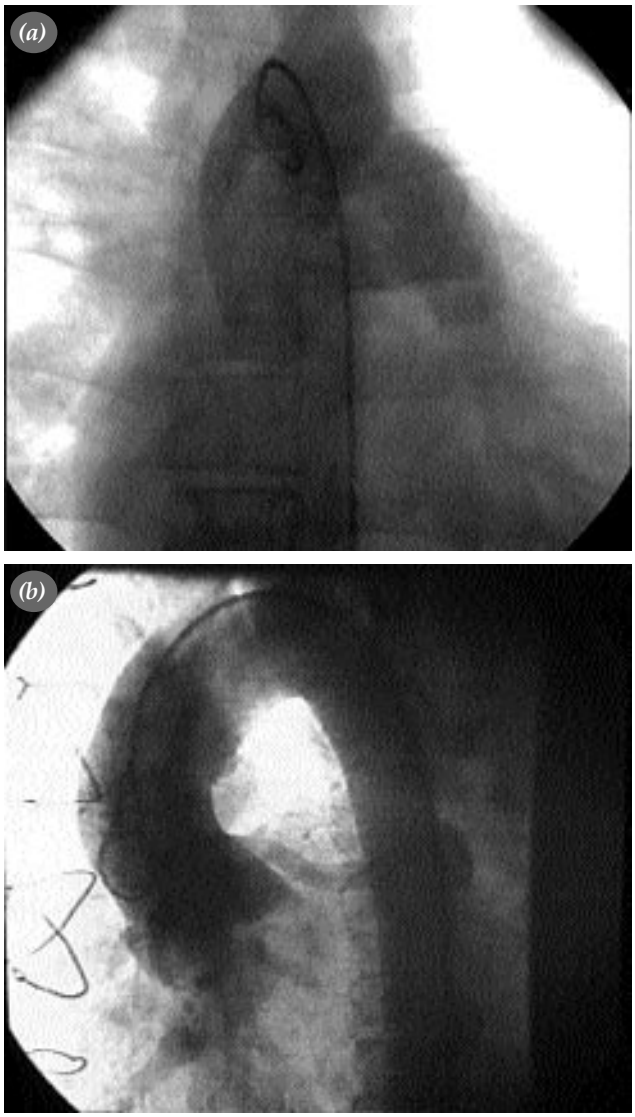

Fig. 1. (a) The preoperative angiographic appearance of the type II aortopulmonary window between the inner curvature of the ascending aorta and the pulmonary trunk. **(b)** Postoperative aortography showing successful closure of the aortopulmonary window and ventricular septal defect without any residual leakage.

1882 congenital cases from February 1985 to October 2003. The other case was a 15-year-old boy who had a simple type I APW, whose defect was successfully repaired using total circulatory arrest in 1988.

Clinically, APW presents with similar symptoms to those of a patent ductus arteriosus and a VSD with pulmonary hypertension. The magnitude of the shunt is mainly related to the size of the defect and pulmonary vascular resistance. Commonly, the defect is large and a large left-to-right shunt is present, resulting in congestive heart failure and pulmonary vascular obstructive disease. Cyanosis is usually absent unless severe pulmonary vascular disease has developed. Since elevation of the pulmonary resistance is rapid in these patients, a prompt diagnosis and treatment are mandatory.

An accurate diagnosis of APW based on clinical symptoms is extremely difficult when it coexists with a VSD. Our patient had been followed-up with a diagnosis of just a simple VSD until the admission to our hospital. This was probably because of the difficulty in differentiating between an APW and a simple VSD. Although two-dimensional echocardiography is important in the diagnosis of this defect,[9] the best method of confirming a suspected APW is to obtain selective angiograms. When there is a coexisting VSD, the diagnosis of APW can be missed unless an aortogram is obtained.

Surgical closure is indicated in all patients with an APW, except for asymptomatic patients with small defects. In most patients, closure should be undertaken at the time of diagnosis because of the risk for pulmonary vascular disease. We used the transaortic approach because of such advantages as a good visualization and preservation of the coronary orifices, the aortic leaflets, and the right and left pulmonary artery orifices.[10]

The morbidity and mortality rates are favorable in the surgical correction of APWs even when other cardiac anomalies exist. In the absence of associated anomalies, the late results of surgical correction are excellent; however, in patients with complex anomalies, the prognosis is largely determined by the presence of these anomalies. In older patients, the outcome will largely depend on the pulmonary vascular resistance at the time of repair. Backer and Mavroudis reported 22 cases of APW in a 40-year period with no mortality; they used transaortic patch closure in the most recent six patients.[6]

In conclusion, APW is a rare, but well identifiable anomaly, and surgical closure is indicated as soon as the diagnosis is established, regardless of the patient's age.

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