Unoperated tetralogy of Fallot in a 68-year-old patient

Altmış sekiz yaşında bir hastada düzeltilmemiş Fallot tetralojisi

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Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, generally treated with total correction within the first two years of life. Occasionally, some unoperated cases can reach older ages. A 68-year-old woman with diabetes mellitus presented with swelling in legs and abdomen, weakness, exertional dyspnea, and orthopnea. On physical examination, she had mild cyanosis with clubbing. Her blood pressure was 110/60 mmHg and pulse rate was 79 beat/min. She had a systolic ejection murmur and bilateral rales on basal lung areas. Massive edema was noted in both lower limbs. Electrocardiography showed atrial fibrillation with normal ventricular response. Chest radiography showed an increased cardiothoracic ratio and bilateral minimal pleural effusion. Echocardiography showed biatrial dilatation (right atrium 62 mm, left atrium 49 mm) and thickening of left ventricular walls. There was right ventricular hypertrophy with decreased systolic function. A very large ventricular septal defect and severe pulmonary stenosis were noted. The patient did not accept any interventional procedure. To our knowledge, this is the oldest unoperated TOF case reported from our country.

Key words: Aged; echocardiography; tetralogy of Fallot/diagnosis.

Fallot tetralojisi en sık siyanotik doğuştan kalp hastalığıdır. Genellikle bu olgulara yaşamın ilk iki yılında tam düzeltme ameliyatları uygulanmaktadır. Cerrahi uygulanmamış olguların erişkin yaşlara ulaşması nadirdir. Altmış sekiz yaşında bir diyabetik hasta bacaklarda ve karında şişlik, halsizlik ve nefes alamama yakınmalarıyla başvurdu. Fizik muayenede hafif siyanoz ve parmaklarda çomaklaşma vardı; kan basıncı 110/60 mmHg, nabzı 79 vuru/dk idi. Dinlemede sistolik ejeksiyon üfürümü ve bazal akciğer alanlarında iki taraflı raller duyuldu. Her iki bacağında yaygın ödem vardı. Elektrokardiyografide normal ventrikül yanıtlı atriyal fibrilasyon izlendi. Göğüs radyografisinde kardiyotorasik oran artmış bulundu ve iki taraflı hafif plevral efüzyon vardı. Ekokardiyografide iki taraflı atriyal genişleme (sağ atriyum 62 mm, sol atriyum 49 mm), sol ventrikül duvarında kalınlaşma, sağ ventrikül hipertrofisi ve sistolik fonksiyonunda azalma görüldü. Hastada çok büyük bir ventriküler septal defekt ve ciddi pulmoner darlık saptandı. Hasta herhangi bir girişimsel müdahaleyi kabul etmedi. Bildiğimiz kadarıyla, bu olgu ameliyat edilmemiş Fallot tetralojili olgular içinde ülkemizde bildirilen en yaşlı olgudur.

Anahtar sözcükler: Yaşlı; ekokardiyografi; Fallot tetralojisi/tanı.

Tetralogy of Fallot includes four major components: right ventricular outflow tract obstruction, outlet ventricular septal defect, overriding aorta, and right ventricular hypertrophy. Actually, it is the consequence of a single developmental abnormality, the misalignment of the supraventricular crest with associated underdevelopment of the infundibulum. This abnormality results in ventricular septal defect, anterior displacement of the aortic valve, right ventricular outflow tract obstruction, and right ventricular hypertrophy. Tetralogy of Fallot is among the most common cyanotic heart diseases in children and accounts for approximately 4% to 8% of all congenital cardiac lesions.[1] In its natural history, progressive hypoxia is expected at the beginning of the life. Survival to adult ages is extremely rare without palliative or corrective operations. The degree of pulmonary obstruction and pulmonary blood supply determines the progression of the disease.

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Survival rate of corrective operations is perfect, 25-year survival being 94%.[2] However, sudden cardiac death due to malignant ventricular arrhythmias originating from ventricular septal patches can occur in the long-term.[3] The longest survival among unoperated TOF cases was reported in a 86-year-old patient.[4]

In this case report, we presented an elderly unoperated patient who was at the age of 68 years at the time of presentation.

CASE REPORT

A 68-year-old woman was admitted to our hospital because of swelling in legs and abdomen, weakness, exertional dyspnea, and orthopnea. On physical examination, she had mild cyanosis with clubbing. Her blood pressure was 110/60 mmHg and pulse rate was 79 beat/min. She had a systolic ejection murmur. Bilateral rales were auscultated on basal lung areas. She had a distended abdomen and palpable liver. Massive edema was noted in both lower limbs. She had diabetes mellitus. She underwent surgery for cholelithiasis, of eight-year history. Electrocardiography showed atrial fibrillation with normal ventricular response, without right ventricular hypertrophy, or right bundle branch block with QRS duration of 90 msec. Chest radiography showed an increased cardiothoracic ratio and bilateral minimal pleural effusion.

Echocardiography showed biatrial dilatation (right atrium 62 mm, left atrium 49 mm), normal left ventricular dimensions and function, and thickening of left ventricular walls. There was right ventricular hypertrophy with decreased systolic function. We noted a very large ventricular septal defect overriding the aorta (Fig. 1a) and severe pulmonary stenosis (with a peak gradient of 69 mmHg) (Fig. 1b). In biochemical analyses, serum creatinine level was 0.9 mg/dl, aspartate aminotransferase and alanine aminotransferase levels were mildly high (42 mg/dl and 48 mg/dl, respectively). She also had hypothyroidism in serum analyses. She was treated with furosemide, spironolactone, warfarin, and L-thyroxine. Insulin was applied subcutaneously four times a day. Cardiac catheterization was recommended, but she did not accept further treatment.

DISCUSSION

The most important abnormality of TOF is obstruction of the right ventricular outflow tract. Although the majority of cases have the complex of four abnormalities described by Fallot, other cardiac and extracardiac abnormalities such as right-sided aortic arch, anomalous coronary anatomy, multiple ventricular septal defects, atrial septal defect, and persistent ductus arteriosus may also be observed in as many as 40% of patients.[1]

When resistance to flow in the right ventricular outflow tract exceeds that of the aorta, right-to-left shunting ensues, leading to cyanosis and diagnosis at an early age. If resistance in the right ventricular outflow tract is less than that of the aorta, there will be a left-to-right shunt across the ventricular septal defect without peripheral cyanosis. Although dynamic cyanosis may occur under extreme right ventricular loading conditions, patients with a restrictive ventricular septal defect are generally acyanotic and known as pink Fallot. The incidence of acyanotic TOF was reported as 5% among patients with TOF.[5]

Life expectancy of unoperated patients with TOF is approximately 10 years and only 3% can reach the fifth

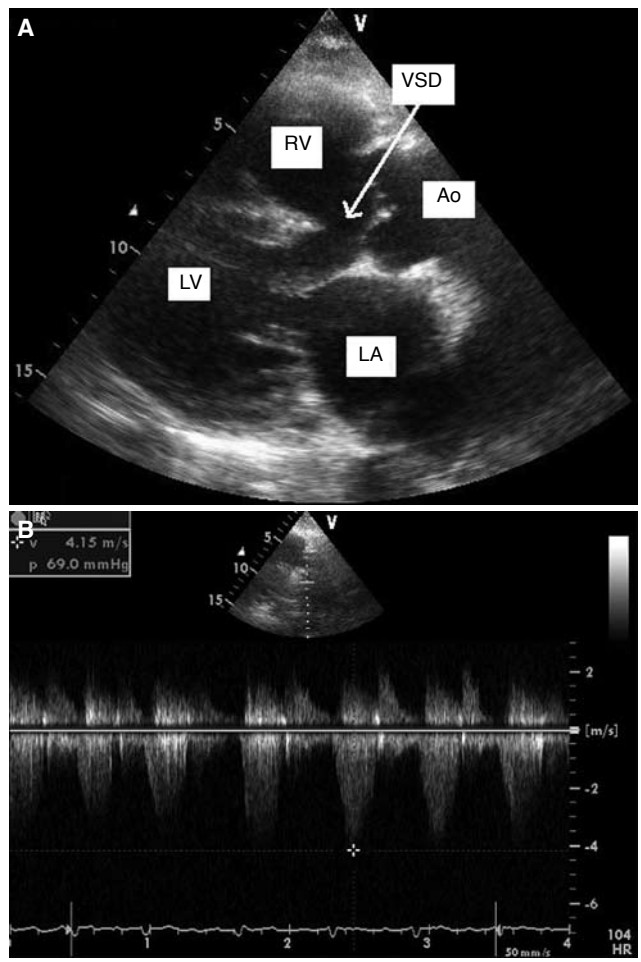

Figure 1. (A) Parasternal long axis view of tetrology of Fallot. VSD: Ventricular septal defect; RV: Right ventricle; Ao: Aorta; LV: Left ventricle; LA: Left atrium. **(B)** Continuous wave Doppler image of the pulmonary valve from the parasternal short axis view showing a pressure gradient of 69 mmHg.

decade.[6] Improved survival in unoperated patients has been attributed to the presence of an extracardiac shunt, like patent ductus arteriosus, which contributes to blood supply to pulmonary circulation.[7] Another reason is the development of left ventricular hypertrophy, which may act as a balancing factor against the occurrence of a right-to-left shunt.[8] In our case, echocardiography showed left ventricular hypertrophy, but no extracardiac shunt.

In a previous study in which 30 patients with TOF underwent total correction at the ages of 40 to 60 years, operative mortality was found as 3%, whereas survival was 92% at five years and 74% at 10 years.[9] We considered cardiac catheterization or surgery but the patient did not accept any interventional procedure.

In conclusion, albeit extremely rare, some patients with TOF, especially acyanotic cases, can reach older ages without operation. To the best of our knowledge, this is the oldest unoperated case reported from our country.

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