CASE REPORT
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Subaortic Membrane Late after Surgical Correction of Tetralogy of Fallot

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We herein report a rare case of subaortic stenosis in association with a previous tetralogy of Fallot (TOF) surgical repair, which was not taken into account as a differential diagnosis. Echocardiography plays a pivotal role in identification of this rare combination. Therefore, echocardiography should be performed periodically during follow-up of patients with surgically corrected TOF. Given the clinical complications that can result from subaortic stenosis (i.e., aortic regurgitation and infective endocarditis), early and aggressive management of this rare combination should be performed.

Keywords: Tetralogy of Fallot; Ventricular outflow obstruction; Echocardiography

INTRODUCTION

Tetralogy of Fallot (TOF) accounts for approximately 5% to 7% of all cases of congenital heart disease [1]. Complications related to surgical repair of TOF are clinically relevant and include pulmonary regurgitation, residual obstruction of the right ventricular (RV) outflow tract, RV systolic dysfunction, and arrhythmias [2,3]. Among these, residual RV outflow tract obstruction with accompanying pulmonary regurgitation is the most frequently reported sequela after initial surgical correction of TOF [3]. Therefore, a systolic murmur that is audible over the base of the heart is frequently interpreted as a residual RV outflow tract lesion. However, this is not always true. We herein report a rare case of subaortic stenosis in association with a previous TOF surgical repair that was not considered to be a differential diagnosis. Discrete subaortic stenosis is generally thought of as a disease of children or young adults (< 25 years of age) and is thus rarely reported in subjects older than 25 years, particularly in association with TOF [4,5].

CASE REPORT

A 29-year-old woman presented for a routine check-up of surgically corrected congenital heart disease. At 7 years of age, she had undergone surgical repair of TOF with excision of the infundibular stenosis and closure of a 10-mm ventricular septal defect. She had been asymptomatic until her first decade of life, and only a 5-year

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clinical follow-up had been performed after the operation. Approximately 6 months prior to presentation, she began to feel exertional dyspnea that was progressively aggravated. On physical examination, she was alert and well-oriented. Her blood pressure was 100/60 mmHg, pulse 60 beats/min, respiration rate 18/min, and body temperature 36.6°C. The left ventricular (LV) impulse was almost normal in location. On auscultation, a grade 3 of 6 mid- to late-systolic murmur was audible over the base of the heart and heard clearly on both sides of the sternum. Routine blood examinations, including a complete blood count, chemistry panel, coagulation panel, and C-reactive protein level, were all within normal ranges. An electrocardiogram showed a sinus rhythm with nonspecific ST-T changes. Transthoracic and transesophageal echocardiograms both demonstrated severe subaortic stenosis due to a subvalvular membrane, on which a mobile mass-like structure that was suspicious of a vegetation was noted (Fig. 1). The peak pressure gradient across the subaortic membrane was 95.3 mmHg (Fig. 2). The aortic valve was trileaflet with associated mild aortic regurgitation. There was a mild degree of tricuspid regurgitation, but no clinically significant obstruction of the RV outflow tract. The maximum velocity measured across the tricuspid valve was 2.8 m/sec, and thus the maximal pressure gradient was estimated to be 31.4 mmHg using the simplified Bernoulli equation. Due to the up to 20 mmHg pressure gradient across the pulmonary valve, clinically relevant pulmonary hypertension was not expected. Her RV was not dilated, and its systolic function was good. Due to the suspicion of infective endocarditis despite the absence of clinical evidence, blood cultures were performed three times; however, no pathogen was cultured. The patient underwent resection of the subaortic membrane and myomectomy of the LV outflow tract along with aortic valve commissurotomy. Fig. 3 shows the subaortic membrane from the surgeon's view. The membrane, with the exception of the noncoronary cusp area, was located beneath the aortic annulus (i.e., inverted U shape). The mobile mass-like structure described on transesophageal echocardiography was proven to be loose tissue attached to the subaortic membrane. The patient remains asymptomatic 18 months after surgery.

DISCUSSION

Although subaortic stenosis is the second most-common form of LV outflow tract obstruction, it is predominantly found in children or adolescents [4-8]. Subaortic stenosis is strongly associated with other congenital heart defects [6,8]; however, subaortic stenosis in association with TOF, especially in women, is extremely rare. According to a Medline database review of the last 30 years, only 11 patients were reported to have had both TOF and subaortic stenosis. Furthermore, all were diagnosed in childhood or adolescence (< 20 years of age) with two exceptions [5]. The present case is peculiar in that subaortic stenosis manifested more than 20 years after the initial TOF surgical correction.

The cause of subaortic stenosis in the present case

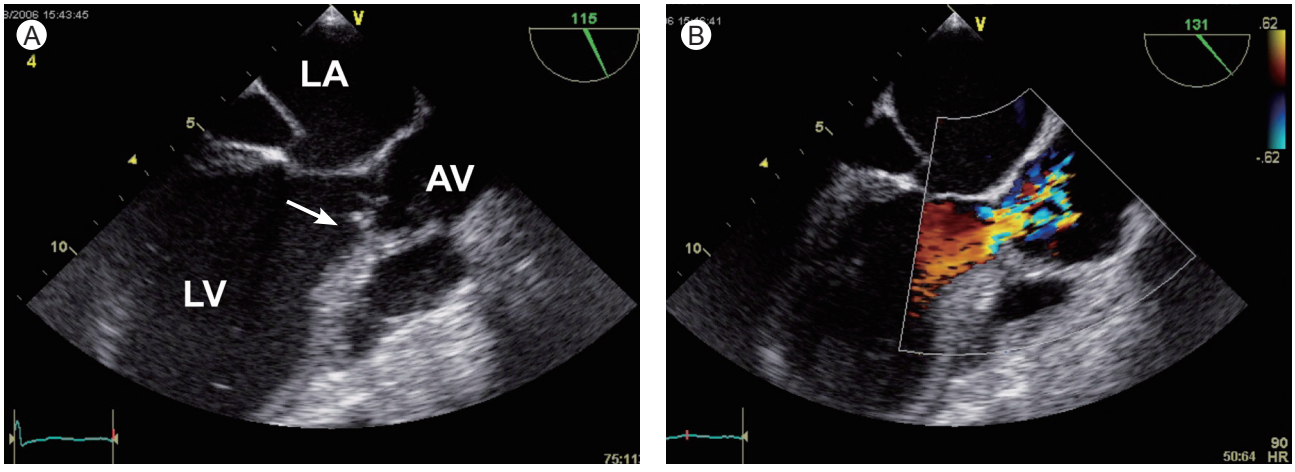

Figure 1. Two-dimensional (A) and Doppler (B) transesophageal echocardiogram in the longitudinal plane. Solid arrow indicates the subaortic membrane. AV, aortic valve; LA, left atrium; LV, left ventricle.

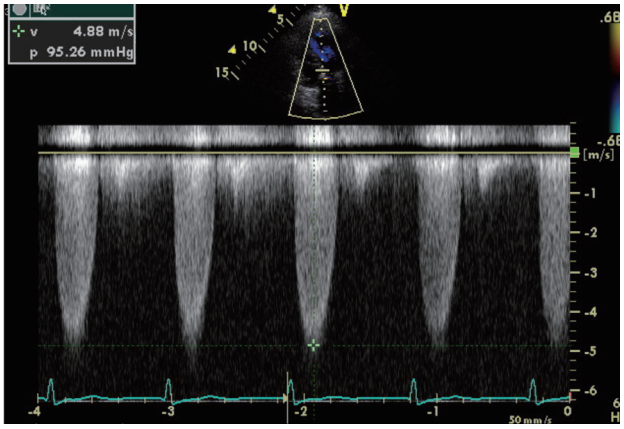

Figure 2. Maximal pressure gradient across the subaortic membrane was estimated to be 95 mmHg.

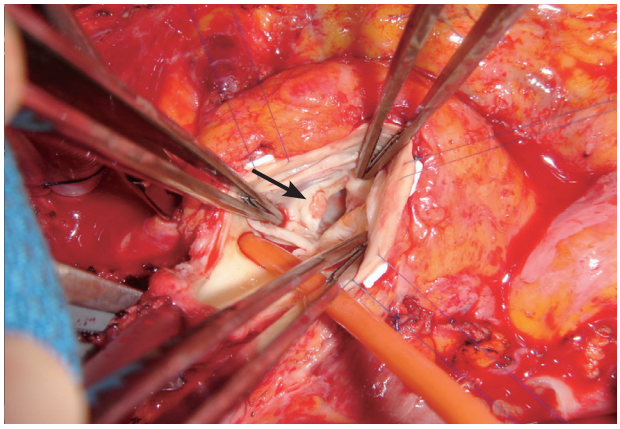

Figure 3. Operative findings. Solid arrow indicates the subaortic membrane.

remains uncertain. However, both congenital and acquired forms were suggested. Although we could not definitively say that the subaortic stenosis in the present case was acquired, we consider it an acquired or progressive form given the long interval (> 20 years) before echocardiographic confirmation. Thomas and Foster [5] suggested that acquired subaortic stenosis can develop gradually due to hemodynamic disturbances derived from coexisting lesions or after surgical correction. Moreover, the mass-like mobile structure observed on transesophageal echocardiography could be regarded as a degenerative byproduct of flow convergence induced by the subaortic membrane, further supporting the concept of an acquired or progressive form of the disease [9,10].

Discrete subaortic stenosis is well-known to be linked to important complications, such as bacterial endocarditis and significant aortic regurgitation. Flow turbulence distal to the obstructing subaortic membrane plays a crucial role in progressive damage to the aortic valve with resultant thickening of its leaflets and regurgitation. The secondary jet lesion also predisposes the patient to bacterial endocarditis [4,9]. In this context, early detection of subaortic stenosis is of clinical relevance and should be pursued aggressively. In particular, patients with surgically corrected TOF tend to be misinterpreted as having only residual RV outflow obstruction without consideration of the LV outflow tract lesion. Transthoracic and transesophageal echocardiography can assist differentiation of the RV from the LV outflow tract obstruction. Therefore, periodic performance of echocardiography should be kept in mind during follow-up of patients with surgically corrected TOF.

Conflict of interest

No potential conflict of interest relevant to this article is reported.

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