Discrete Subvalvular Aortic Membrane With Double Aortic Arch

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Abstract

Double aortic arch with discrete subvalvular aortic membrane is an uncommon association. We present in this study a case that diagnosed discrete subvalvular aortic membrane with double aortic arch.

INTRODUCTION

Vascular rings are rare, however, often present with common symptoms (1). Double aortic arch is a common form of complete vascular ring, encircling both the trachea and esophagus, resulting in noncardiac morbidity (2). Congenital vascular rings may often cause unexplained respiratory symptoms in infants and young children. Their diagnosis and treatment are often delayed (3).

CASE PRESENTATION

Our case was an 11-year-old boy. He was suffering from easy fatigability for a year. Investigations revealed severe aortic stenosis and he was referred to our clinic for operation. Transthoracic echocardiography showed valvular stenosis with a peak gradient of 95 mm Hg. Thereafter, cardiac catheterization was carried out identifying a valvular aortic stenosis with a peak gradient of 45 mm Hg and a separate subvalvular aortic stenosis with a peak gradient of 80 mm Hg. Moreover, a double aortic arch anomaly was revealed, where RCA and LCA were originating from superior arch and LCSA and RCSA from inferior arch (Figures 1, 2 & 3).
No pathological finding or anamnestic characteristic specific to the respiratory system was identified.

With these findings, we brought our patient into the operating room. We carefully performed median sternotomy and routine cannulation. Arrest was achieved with moderate hypothermia of 30°C, and incompressive retrograde isothermic potassiumed blood cardioplegia. Following aortotomy we explored; a normal aortic valve. On the other hand; discrete subvalvular aortic membrane located in a circular manner was explored (Figure 4).

This circular discrete fibrous membrane was then resected (Figure 5).

No insufficiency was identified in the native valve after the procedure, where there already wasn’t any stenosis at valvular level. A Hegar dilator of 18 mm could easily pass through the valve (Figures 6&7).
No additional problem was seen postoperatively. Patient was discharged on 7th postoperative day with total recovery. In the first postoperative month a control transthoracic echocardiography was carried out showing an insignificant aortic regurgitation.

**DISCUSSION**

Abnormalities of brachiocephalic arterial branching and arch laterality are common in patients with a cervical aortic arch. In addition, structural anomalies of the arch such as obstruction, aneurysms, and tortuosity are found in a significant number of cases (4).

In the setting of normal cardiac situs, a right-sided aortic arch is uncommon. When a right arch does occur, it is typically in conjunction with other congenital cardiovascular anomalies. The rarity of such lesions among patients with right aortic arch may be explained in part by the fact that the fetal hemodynamic conditions associated with persistence of a right arch do not facilitate flow-related arch obstruction (5). Such complex cardiac lesions may complicate an otherwise normal surgical procedure.

Such abnormalities may be the result of hemodynamic conditions and/or abnormal vascular tissue related either to the cervical position of the arch or its embryologic precursors. Given the highly variable anatomy of patients with a complicated cervical aortic arch, surgical considerations will vary in kind (4).

In conclusion; complete resection of discrete subvalvular aortic membrane associated with double aortic arch is feasible. Complex cardiac anatomy presents no additional risk for this procedure. It yields excellent surgical results and good outcome.

**References**

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