Obstructive membrane in arch of aorta in a case of Shone’s complex

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ABSTRACT

Shone’s complex is a rare congenital heart disease consisting of multisite obstruction on the left side of the heart. The obstructive membrane in the arch of aorta is never described among these obstructions. We report echocardiographic findings in a patient with Shone’s complex with the obstructive membrane in the arch of aorta.

Key words: Echocardiography; Obstructive membrane; Shone’s complex

INTRODUCTION

A 30-day-old female child was referred to our center with complaints of fast breathing and feeding difficulty. On examination, she was underweight and had respiratory distress. Femoral and pedal pulses were feeble while upper limb pulses were of good volume. Her arterial oxygen saturation was 99%. Her chest X-ray showed levocardia, the cardiomegaly-cardiothoracic ratio of 0.6 with left ventricular type apex and normal pulmonary vascularity. The echocardiogram showed levocardia, situs solitus, and incomplete Shone’s complex. There was parachute mitral valve with mild mitral stenosis, bicuspid aortic valve with mild aortic stenosis, severe coarctation of the aorta, severe pulmonary artery hypertension, left ventricular dysfunction. In addition, an obstructive echogenic membrane was noted in the arch of aorta, just after the origin of innominate artery (best seen in subcostal coronal view and in suprasternal long axis view) [Figure 1]. The membrane was causing mild obstruction with a peak gradient of 20 mmHg with mild diastolic spillage. The patient was posted for off-pump surgical coarctoplasty with resection and end to end anastomosis from the left posterolateral thoracotomy, but membrane could not be approached due to its unique location at the surgical aortic clamp site. The postoperatively membrane was in same status with a gradient of 16 mmHg with mild diastolic spillage [Figure 2].

DISCUSSION

Shone’s complex is a rare congenital heart disease described by Shone et al. initially, in 1963. It typically consists of four obstructive lesions of the left side of heart and circulation namely parachute mitral valve, supravalvar mitral ring, subaortic stenosis, and coarctation of the aorta.[1] There is a complete form of Shone’s complex wherein all the four lesions are present; however, incomplete forms with two or three lesions are also described.[1] Other coexisting mitral valve anomalies have been reported such as fused chordae, single papillary muscle, and “typical” (Ruckman and Van Praagh) congenital mitral stenosis.[2] The left ventricular outflow tract obstruction features may include subaortic stenosis, valvar aortic stenosis, bicuspid aortic valve, and coarctation of the aorta.[2] Our patient had incomplete Shone’s complex as she was not having supramitral stenosis. After reviewing
available published literature on this topic, we came to know that obstructive membrane in the arch of aorta has not been described. This finding in our patient may be a part of Shone’s complex, or it may be just an associated finding. This makes this case as first of its kind in available literature.

We had planned off-pump coarctoplasty with minimum cross clamping time. After discussing with our surgeon, we had decided that we will look for the membrane and try to excise, if possible off-pump only. As our surgeon felt that the membrane was exactly at the clamp site, the membrane could not be approached, so only coarctoplasty was carried out. The membrane was examined after the surgery; it was at the same site with the same severity of obstruction.

**CONCLUSIONS**

Shone’s complex is a combination of parachute mitral valve, supramitral ring, subaortic stenosis, and coarctation of the aorta. The obstructive membrane of this kind has not been described till now; it may be a part of Shone’s complex and has to be looked in case of Shone’s complex for proper understanding of this kind of membrane.

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**Conflict of interest**
There are no conflict of interest.

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