Case report

Repair of aortic arch atresia with diffuse hypoplasia of the descending thoracic aorta

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Abstract

We report successful repair of a rare combination of aortic arch atresia and diffuse long-segment hypoplasia of the descending aorta in a 2-month-old infant with PHACES association. Intraoperative findings and surgical techniques are discussed.

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1. Introduction

Aortic arch atresia is a rare but well-known pathological entity distinct from aortic arch interruption (IAA). We are reporting an interesting presentation of aortic arch atresia with a severe hypoplasia of the descending thoracic aorta in 2-month-old infant with PHACES syndrome.

2. Case report

A 2-month-old baby girl presented to our hospital in shock with multi-organ dysfunction. She was intubated and resuscitated with intravenous fluids and inotropes. Trans-thoracic echocardiography showed an IAA with suspicion of hypoplasia of the descending aorta (DAo), left ventricular dysfunction (fraction shortening = 14%), and moderate mitral regurgitation. To clarify the aortic anatomy we proceeded with a computerized tomography (CT) angiography, which suggested IAA type B with very severe diffuse long-segment hypoplasia of the DAo down to 2 cm above the diaphragm (Fig. 1A). Clinically, the patient continued to demonstrate progressive improvement of her renal and hepatic functions with inotropic support and mechanical ventilation. She also had a left facial hemangioma extending to the retro-orbital space and intra-cranial posterior fossa. The collection of these findings was consistent with the genetic disorder called PHACES syndrome. Because of the long-segment DAo hypoplasia, we decided to perform full repair of the aortic arch and DAo through a left thoracotomy without cardiopulmonary bypass. In addition, we preferred to avoid cardiopulmonary bypass and cardiac arrest in the setting of the severe left ventricular dysfunction.

3. Surgical technique

Using a single skin incision and muscle-sparing thoracotomy, we entered the left chest through the third and seventh intercostal spaces. This provided an excellent exposure to the entire thoracic aorta. To our surprise, there was an extensive dense fibrosis of the posterior mediastinum with inflamed lymph nodes surrounding the aortic arch and the origins of the great vessels (Fig. 1B). This inflammation was probably part of the pathological process, although it has not been described before. The intraoperative anatomy was consistent with aortic arch atresia instead of interruption. The atretic segment of the arch was thickened and fibrosed with no lumen and extended from the left common carotid artery (LCCA) to the proximal DAo for a distance of 4 cm above the diaphragm (Fig. 1A). Clinically, the patient continued to demonstrate progressive improvement of her renal and hepatic functions with inotropic support and mechanical ventilation. She also had a left facial hemangioma extending to the retro-orbital space and intra-cranial posterior fossa. The collection of these findings was consistent with the genetic disorder called PHACES syndrome. Because of the long-segment DAo hypoplasia, we decided to perform full repair of the aortic arch and DAo through a left thoracotomy without cardiopulmonary bypass. In addition, we preferred to avoid cardiopulmonary bypass and cardiac arrest in the setting of the severe left ventricular dysfunction.

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repair was unachievable despite extensive mobilization due to the long atretic segment and severe thickening and fibrosis of the posterior mediastinal structures. We allowed the patient temperature to drift to 34 °C, and then we clamped the DAo distally at the diaphragm, ligated the small patent ductus arteriosus and controlled all the intercostal branches with fine aneurysm micro-clips. The DAo was opened longitudinally down to the level of the diaphragm. We used a patch of bovine pericardium to perform angioplasty of the DAo. After reconstructing the DAo, we clamped it proximally and released the distal clamp and the micro-clips to perfuse the intercostal branches via collaterals. The distal aortic arch was constructed using an 8 mm PTFE tube graft extending from the proximal arch opposite to the innominate artery to the newly constructed DAo (Fig. 1C and D). After removal of the clamps, aortic antegrade flow was restored with equal pressure readings from the arterial lines in the right radial and femoral arteries. The patient had no neurological complications and uneventful postoperative course with quick weaning of inotropic support and extubation. The patient was discharged home 2 weeks postoperatively in good condition. Histological examination of specimens taken from the aortic wall and posterior mediastinal lymph nodes showed non-specific inflammation and fibrosis. One year later, the patient is asymptomatic with no gradient across the arch or the descending aorta as seen by Doppler echocardiography.

4. Comment

In 1996 Frieden et al. proposed the acronym PHACES to define a syndrome including posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, eye abnormalities, and sternal malformations or malformations of the abdominal midline [1]. Up to 70% of patients have only one extra-cutaneous diagnosis and this syndrome occurs predominantly in girls [2]. Congenital cardiac anomalies are reported in about one third of patients with PHACES syndrome and the most common anomaly is aortic coarctation [1,2]. Arterial anomalies such as aberrant subclavian artery, stenosis or atresia of the origin of an aortic arch branch, and others have also been commonly reported [1—3].

Several other authors have reported successful repair of DAo hypoplasia [4—6]. To our knowledge this is the first report of distal aortic arch atresia and diffuse long-segment DAo hypoplasia, which requires surgical reconstruction of the thoracic aorta from the mid-arch down to the distal DAo. This can be achieved either by anatomical or extra-anatomical graft bypass or by combination of short interposition graft to reconstruct the gap in the aortic arch and patch angioplasty of the DAo. We opted for the latter option for the following reasons: (1) the estimated length of a PTFE tube graft to connect the ascending aorta to the distal DAo would have been about 15 cm. We believe that such a relatively long and narrow graft will likely cause a pressure gradient and an early need for re-intervention. (2) The lumen of the DAo was very small, although it was giving multiple large branches. Retrograde blood flow might not be adequate for these branches. (3) There is potential for growth of the retained part of the DAo wall after restoring antegrade flow. Considering the severe inflammation and fibrosis surrounding the aortic arch and DAo, the use of extra-anatomical ascending to descending aortic bypass through a median sternotomy will be an attractive option in the future when this patient develops a pressure gradient across the PTFE graft used for arch reconstruction due to somatic growth. In the Congenital Heart Surgeons Society study on risk factors associated with repair of IAA in neonates, McCrindle and colleagues reported that when repair was accomplished by placement of a PTFE interposition graft, at 16 years, there was a 54% chance of survival to arch reintervention, a 30% chance of death, and a 16% chance of survival without reintervention [7].

References

