Neonatal ascending aorta thrombosis: A rare and lethal entity

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ABSTRACT

Ascending aorta thrombosis (AAT) in a neonate is a rare and lethal event. To date, AAT has been reported in around 25 patients, out of whom only eight patients had concomitant arch involvement. We report a case of one such patient with ascending aorta and arch thrombosis and present a brief review of the available literature.

Keywords: Ascending aorta thrombosis, neonatal aortic thrombosis, surgical thrombectomy

INTRODUCTION

Ascending aorta thrombosis (AAT) in a neonate is a rare and lethal event. Most of the published literature comprises case studies, and the overall mortality is high. Thrombosis is found more commonly in the abdominal aorta than the thoracic aorta, in which case umbilical artery catheterization is the most common identified cause. AAT has been reported in 25 patients to date, of whom only eight patients had concomitant arch involvement.\(^1\)\(^-\)\(^5\) Surgical thrombectomy is the preferred management option. We report a case of one such patient with ascending aorta and arch thrombosis. We also present a brief review of the available literature.

CASE REPORT

A 24-year-old, 36 weeks pregnant previously unbooked patient presented to the department of obstetrics with a normal antenatal history. On fetal echocardiography, the fetus was diagnosed to have interrupted aortic arch with dilated left ventricle, pericardial effusion, and heart failure, which prompted an emergent cesarean section. A male child weighing 3.36 kg was born, who cried immediately after birth, but started developing cyanosis refractory to oxygen therapy. Apgar score was 4 and 8 at 5 and 10 min. The child had poor respiratory efforts and was intubated. Heart rate was 120/min, respiratory rate was 65/min, and liver edge was 3 cm below the right costal margin. Postdelivery echocardiogram revealed type B interruption of the aorta with a clot in the ascending aorta [Figure 1], right-to-left flow across patent ductus arteriosus, severe left ventricular dysfunction, and pericardial effusion. Mother was negative for TORCH [Figure 1: Echocardiograph showing clot in the ascending aorta]
infections, COVID antigen, and antibody. Umbilical arterial catheterization was not performed.

The child was taken up for emergency surgery. Aortic and pulmonary artery cannulation was done along with single venous right atrial cannulation and right superior pulmonary venous vent. Right and left pulmonary arteries were snugged. On insertion of antegrade cardioplegia cannula, there was no flow. Transverse aortotomy was done to find total aortic occlusion by clots [Figure 2], extending to the interrupted segment between left common carotid and left subclavian artery. Direct ostial Del-nido cardioplegia was given and the patient was cooled to 18°C for total circulatory arrest. The clot was extending into right common carotid and right subclavian artery for which balloon thrombectomy was done. Clot was removed and interruption repaired by end-to-side anastomosis with pericardial patch augmentation. Total circulatory arrest time was 65 min with aortic clamp time of 98 min. The patient could not be weaned off bypass support.

**DISCUSSION**

AAT in neonates is a rare finding. Iliac, femoral, and cerebral arteries are the most commonly involved vessels in arterial thrombosis. Most of the documented cases of aortic thrombosis involve the descending aorta or the renal arteries, and umbilical artery catheterization is the most common identified cause.[6] Other factors, which can predispose, include polycythemia, dehydration, asphyxia, maternal diabetes, and coagulopathy.[7]

Most common manifestations include unequal or absent pulses, cyanosis of the lower body, and heart failure. Echocardiography is diagnostic. Guidelines by the American College of Chest Physicians advise that treatment should be individualized based on the urgency of the clinical situation and the extent of thrombosis, which includes anticoagulation, thrombolytic therapy, and surgery.[8] Streptokinase, urokinase, and heparin, which form the medical approach to therapy, have better results in thrombosis of descending aorta caused by iatrogenic umbilical artery catheterization.[7] Results of medical management in thrombosis of the ascending aorta are inconsistent, and it is of limited value, only to tide over the initial crisis. Successful surgical repair with survival beyond the immediate postoperative period has only been described in one patient of ascending aorta with arch thrombosis, although it has been documented in eight out of twelve surgically managed patients of AAT alone. The mortality is 100% in untreated patients. Comorbidities such as renal, brain, and bowel ischemia also affect survival.[2]

From 1953 to 2020, we were able to identify 25 patients with neonatal AAT.[1-5] Of these 25 patients, 14 (60%) died. The mortality rate of patients with multiple thrombosis was 84.6% (11/13) compared with 33.3% (4/12) for patients with isolated AAT. Only one patient out of eight who had combined ascending aorta and arch thrombosis survived. All three patients with central nervous system involvement died [Figure 3]. The patients who either had no treatment (5 patients) or underwent thrombolysis (two patients) died [Figure 4]. Overall mortality in patients undergoing surgical thrombectomy was (8/18) 44.4%.

The cause of AAT was documented in only (7/25) 28% of patients.[1] Three patients were identified to have factor V Leiden heterozygous mutation, two patients had protein C deficiency, and two patients had cytomegalovirus infection. Eighteen patients (72%) had no etiology identified [Figure 5]. Protein C deficiency and factor V Leiden mutation are the most common risk factors for aortic thrombosis as per the literature regarding the potential underlying thrombophilic risk factors.[1]
In summary, AAT in neonates is a rare but highly lethal condition of unknown origin, requiring immediate surgical management. Prognosis is dismal, especially if there is involvement of arch or other concomitant systems.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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