Perioperative Management of a Neonatal Aneurysm of the Vein Galen with Heart Failure

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Aneurysm of the vein of Galen is an uncommon disease defined as an aneurysmal dilation of the vein of Galen located between the great cerebral veins and the straight sinus¹. Johnston et al.² reported on 245 cases of the aneurysm collected from the literatures since the 1940's. The mortality rate of 80 neonatal cases in the report exceeded 90% and only three intact survivors were confirmed. We report on a neonatal aneurysm with congestive heart failure treated successfully.

Case Report

The male neonate had an uneventful 40-week-gestation and weighed 4,110g at birth. Two days after the delivery, tachypnea, tachycardia and cyanosis appeared. As congenital heart anomaly was suspected, he was transported to our hospital at 7 days old, where he showed cyanosis and retractive respiration on admission. Chest X-ray showed cardiomegaly of 78% CTR. (fig. 1) Echocardiography revealed severe pulmonary hypertension with right to left shunt through the patent ductus arteriosus (PDA) as well

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as through the foramen ovale. Retrograde aortogram through the right radial artery failed to identify the exact lesion of cardiac failure, however this maneuver worsened heart failure with dyspnea enlarging CTR up to 87%. Immediate artificial ventilation started to maintain Pa_{O_2} above 90 mmHg and Pa_{CO_2} below 35 mmHg. Ten $\mu g \cdot k g^{-1} \cdot min^{-1}$ dopamine was administrated. When cardiac catheterization was performed two days after the first examination, abnormal high oxygen saturation of superior vena caval blood gave a clue for diagnosis, which was confirmed by CT scan later (fig. 2).

The operation was performed two days after the examination. Anesthesia was induced by 10 $\mu g \cdot k g^{-1}$ fentanyl and maintained additional 20 μ g fentanyl and 55 to 60% nitrous oxide with the aide of pancronium. Blood pressure was monitored directly with an indwelling catheter of the right radial artery. A large bore catheter was inserted to the superior vena cava (SVC) from the axillar vein for monitoring saturation of SVC as well as for massive blood transfusion. At the beginning of the operation, hemodynamics was stable except moderate tachycardia and low diastolic pressure (heart rate: 150/min blood pressure: 70/38 mmHg). When the right feeding artery to the aneurysm was occluded, systolic pressure rose up from 70 to 100 mmHg and the heart rate fell to 100/min. The increased blood pressure was attenu-



Fig. 1. A chest X-ray film on admission.

ated by sodium nitroprusside (SNP) infused at the rate of 4 $\mu g \cdot k g^{-1} \cdot min^{-1}$. Following the transient increase of blood pressure, despite the interruption of the SNP infusion, blood pressure gradually went down and heart rate increased, accompanied with a narrowing of the pulse pressure. Since intraoperatively performed echocardiography revealed a worsened cardiac wall motion compared to the preoperative state, clipping of the other feeding artery was postponed. Forty ml of concentrated red cell (CRC) was transfused against 50g blood loss during the operation. Transfontanellar doppler echography confirmed reduction of the shunt flow and the still significant influx through the unclamped feeding artery. The residual feeding artery was clipped 7 days after the first operation. Anesthesia was induced and maintained by fentanyl as the same dose as the first operation. Despite an expected increase of blood pressure and bradycardia at clipping, no significant changes of heart rate nor blood pressure were observed this time.

Thereafter, the neonate was received other operations for coagulation of secondary growing small feeding arteries and a ventricular-peritoneal shunt for secondary hydrocephalus. Although cardiomegaly with pulmonary hypertension and PDA persist,



Fig. 2. A preoperative CT scan of the aneurysm.

his psycomotor development is presently within the normal range of a 10 months old child.

Discussion

The symptom of most concerned in aneurysm of the vein of Galen in the neonate is congestive heart failure due to high cardiac output. In the neonatal period, increased venous return through the aneurysm causes pulmonary hypertension accompanied with right to left shunt through the re-opened ductus arteriosus and the foramen $ovale^{3,4}$. Systemic hypoxia due to right to left shunt and hypercapnea worsen the pulmonary hypertension further. Thus, in addition to standard catecholamine therapy, ventilatory support plays an important role in attenuating pulmonary hypertension. Care should be taken with invasive procedures, which might fatally deteriorate the heart failure⁵.

Fentanyl was selected as an anesthetic with the least cardiovascular suppression. The combination of fentanyl and nitrous oxide offered stable hemodynamics at induction as well as during maintenance for the first two operations. As regards the precipitous increase of afterload at the occlusion of the feeding artery, reducing excessive circulating blood volume is recommended preoperatively⁶. However, McLeod et al.⁷ suggested the neonate with aneurysm kept in hypovolemia risks of inadequate myocardial perfusion which may lead to cardiac arrest when exposed to even small amounts of blood loss. Thus, we kept the neonate nearly euvolemia by replacing blood loss with CRC.

The decision to leave one of the main feeding arteries unoccluded increased the risk of gradually progressing heart failure after the operation. An occlusion of all feeding arteries has been preferred, however, our neonate could have been intolerant of a precipitous increase of afterload. The advantage of a stepped operation is that the leaving some feeding arteries may work as a vent of the left ventricle. Even partial reduction of the shunt flow can reduce cardiac work to allow the neonate to adjust the peripheral vascular resistance to the new hemodynamic circumstances within a few days. The fact that no significant hemodynamic change was observed at the second operation in the case indicates that this adaptation occurred. Embolization of the aneurysm has been tried in order to reduce shunt flow on neonates with serious cardiac failure⁸. Although further discussion is necessary regarding those condition which would indicate and interval between operations, a stepped procedure including embolization before surgical correction should be considered valid.

In conclusion, preoperative heart failure due to aneurysm of the vein of Galen should be treated with catecholamines and also with ventilatory support. Post-clipping hypertension and heart failure could be overcome by a staged operation. During operation, euvolemic condition would be advantageous to the management in neonatal cases.

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