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A case of multiple aneurysms of the vein of Galen with heart failure due to persistent fetal circulation

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Introduction

Vein of Galen aneurysm (VGA), first mentioned in the literature in 1937 [1], has a high mortality rate in spite of the development of therapeutic strategies. Mortality rates as high as 91.4% have been reported for a total of 80 neonates [2]. VGA frequently accompanies congestive heart failure, which is the major cause of mortality and morbidity [3], and neonates with this disease present the most severe challenges to physicians. We report a patient with huge multiple aneurysms with persistent fetal circulation, which were treated by embolization with direct injection of coils into the aneurysms under craniotomy.

Case report

A 30-year-old pregnant woman was referred to Osaka University Hospital with a diagnosis of suspected fetal hydrocephalus. Color doppler ultrasonography revealed a fetal intracranial aneurysm and heart failure secondary to the aneurysm.

At 35 weeks' gestation, a male baby was delivered by cesarean section. The body weight was 2787 g, and Apgar scores were 5 at 1 min and 7 at 5 min. Because of evident respiratory distress and impaired arterial oxygenation as monitored by pulse oximetry, he was intubated and ventilated mechanically soon after the delivery. A few minutes after intubation, he had seizures and diazepam was administered intravenously. His arterial blood gas data showed severe metabolic acidosis with hypoxemia. The infant was then admitted to the intensive-care unit. The electrocardiogram showed sinus tachycardia with a rate of 177 · min^{-1} and a respiratory rate of 47 · min^{-1}. Blood pressure was 44/24 mmHg. Arterial blood gas analysis showed 88%–90% of preductal SaO_2 and 95%–98% of postductal SaO_2. Cranial color doppler ultrasonography revealed multiple large aneurysms with turbulent blood flow in the left hemisphere. Two-dimensional and doppler echocardiography revealed dilatation of the right side of the heart, over-systemic pulmonary hypertension with wide patent ductus arteriosus and foramen ovale, complicated by a large amount of right-to-left shunt flow, mainly through the former sites. Head helical computed tomography was performed, and the three-dimensional image of the brain was reconstructed to form a computer graphic image to aid in understanding the location of each aneurysm. The three-dimensional image revealed severe dilatation of the vein of Galen, surrounded by multiple aneurysmal vessels (Fig. 1). The number of aneurysms was estimated as approximately 100.

Treatment with intravenous administration of inotropic drugs and diuretics and inhalation of nitric oxide at 8 ppm was immediately started for congestive heart failure and persistent fetal circulation. The patient was ventilated with high-frequency oscillation (HFO) because conventional mechanical ventilation did not improve the arterial blood oxygenation, and HFO was thought to be more useful for improving the pulmonary hypertension. Because of the size and number of the aneurysms, we did not choose percutaneous endovascular approaches, but rather embolization by direct injection of coils into the drainage veins under craniotomy. To prevent brain edema and hemorrhage due to normal perfusion pressure breakthrough after the obliteration

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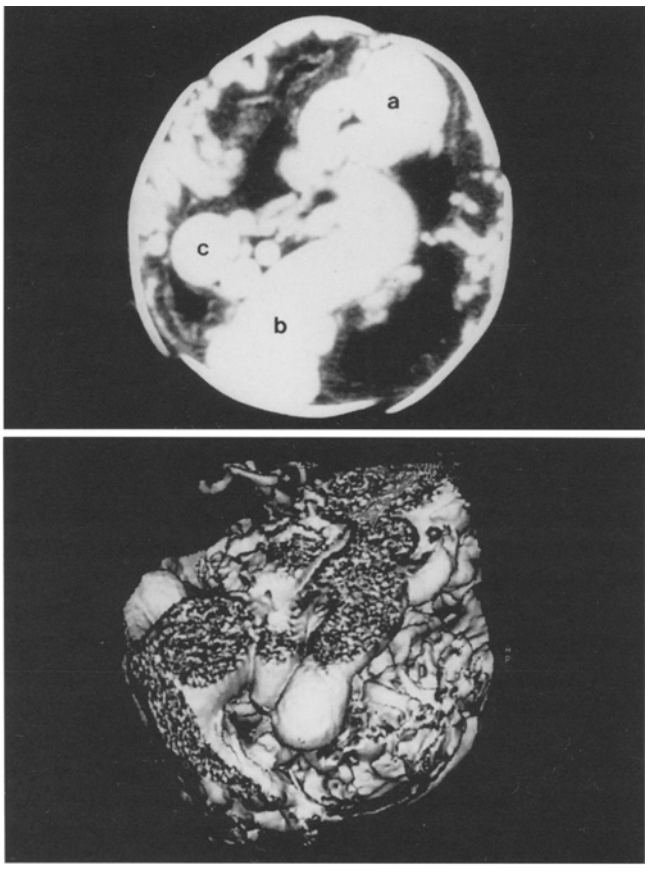

Fig. 1. Two-dimensional **a** and three-dimensional **b** computed tomographic images. *a*, *b*, and *c* represent three large aneurysms, and numerous aneurysms are seen in the images

of some of the aneurysms, staged embolization of aneurysms was scheduled, and embolization of half of the aneurysms using more than 50 coils was performed under general anesthesia according to the neurolept anesthesia method. We confirmed the location of the coils following a plain radiograph of the skull.

During the period following the embolization, ultrasonic study revealed that there was a slight improvement in the volume overload of the right heart and in the right-to-left shunt through the ductus arteriosus. Subsequently, the heart rate increased and blood pressure decreased. We started peritoneal dialysis because of anuria and increased the inotropic support. Cranial doppler ultrasonography then revealed a persistently large amount of blood flow around the aneurysms and the persistence and worsening of fetal circulation, even showing retrograde flow through the aortic arch in the systolic phase. Electroencephalography showed flat waves, and the infant showed no reaction to stimuli, even long after the administration of sedative drug was stopped. He died at the age of 5 days of refractory circulatory decompression.

Discussion

The mortality and morbidity from VGA in neonates remains high despite developments in therapeutic strategy. Less than 10% of neonates with this anomaly survive for long [2].

The most striking manifestation of VGA is congestive heart failure. In severe cases, as much as 80% of left ventricular output may be delivered to the head due to low vascular resistance within the malformation. Since this output returns directly to the right ventricle, right heart failure due to volume overload is always a possibility. In addition, myocardial ischemia caused by the reduction in diastolic pressure may occur [4,5], which also worsens the cardiovascular distress. Consequently, the primary indication for the treatment of VGA is for congestive heart failure refractory to medical treatment. The end point of treatment is not necessarily a complete occlusion of the shunt, but rather improvement in cardiac function [3]. Persistent fetal circulation, which is due to remarkably decreased systemic vascular resistance and not to elevation of pulmonary vascular resistance, makes the cardiovascular distress more complex and severe. In some reports, persistent fetal circulation is the first sign of cerebral arteriovenous malformation in neonates [6]. In the case reported here, the elevation of pulmonary resistance, due to the increased left-to-right shunt around the VGA, combined with low systemic blood pressure caused by the massive shunt, induced persistent fetal circulation. Insufficient elevation of oxygen tension around the ductus left the neonate trapped in a vicious circle. Metabolic acidosis occurred became of an insufficient supply of oxygen and might have caused the impaired response to inotropic support. After diagnosing persistent fetal circulation with doppler ultrasonography, we began to administer inhaled nitric oxide to selectively reduce pulmonary vascular resistance [7]. Although nitric oxide was effective for several hours, the benefit was limited.

Ever since Jaeger et al. [1] reported on carotid ligation as one measure to control the degree of blood flow through the aneurysmal malformation [1], various therapeutic techniques have been introduced into practice. Advances in catheter design and embolization materials have allowed the therapeutic option of neuroradiologic endovascular approaches to the treatment of VGA. As surgical procedures pose a higher risk in neonates than in older patients, endovascular techniques are preferable in neonates or infants [3]. Various techniques of approach for the obliteration of VGA have been described, including transarterial, transvenous, and transtorcular embolization [5,8,9]. In this case, there were too many aneurysms to try any of these conventional endovascular approaches. We chose instead a more invasive embolization procedure, the

direct injection of coils into the drainage vessels under craniotomy. Although the technique was invasive, we considered it an acceptable therapy to try to rescue the neonate, because of the size and multiplicity of the aneurysms and the presence of an emergent and life-threatening condition caused not only by the stolen output of the left ventricle, but also by the persistent fetal circulation. After failing to improve the condition of the neonate by embolization, we considered alternative methods, such as patent ductus arteriosus ligation and foramen ovale closure, but we thought that they were not likely to be effective because, in addition to the risk of operation, such procedures would increase the pulmonary blood flow, which would make the respiratory condition worse. Extracorporeal membrane oxygenation (ECMO) might have been effective, but we did not select it because ECMO would give no more than temporary relief, and anticoagulation therapy during ECMO would limit our options, including additional embolization.

Ultrasonography, computed tomography, magnetic resonance imaging, and angiography are useful means of diagnosing VGA [3]. Ultrasonography is a noninvasive and repeatable means of examination that quickly provides interior imaging, but it does not offer a three-dimensional view. Angiography does, but it is invasive and may sometimes even be life-threatening to neonates in severe cardiovascular distress. Consequently, helical computed tomography and the reconstruction of a three-dimensional image from two-dimensional image data is very useful, because it is not invasive and presents comprehensive information concerning aneurysms, which, we found, could successfully guide embolization procedures.

To conclude, we have presented the clinical course of a case of a neonate with VGA. Invasive therapy consisting of the injection of coils under craniotomy was attempted. Inhalation of nitric oxide provided temporary relief, but ultimately we were unable to stabilize the circulatory system. VGA is a rare condition, but when it does occur, it presents extreme challenges to neurosurgeons and to intensive-care physicians.

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