Korean Society of Maternal Fetal Medicine
Vol. 7 No. 4 December 2011

산전 진단된 심부전을 동반한 Vein of Galen Aneurysmal Malformation의 내혈관치료 1예

성균관대학교 의과대학 삼성서울병원 산부인과[1], 영상의학과[2]

조세은[1] · 윤 건[1] · 차현화[1] · 최석주[1] · 오수영[1] · 박병관[2] · 노정래[1] · 김종화[1]

A Case of Endovascular Treatment for Prenatally Diagnosed Vein of Galen Aneurysmal Malformation Complicated with Heart Failure

Se-Eun Cho, M.D.[1], Gun Yoon, M.D.[1], Hyun-Hwa Cha, M.D.[1], Suk-Joo Choi, M.D.[1], Soo-Young Oh, M.D.[1], Byung Kwan Park, M.D.[2], Cheong-Rae Roh, M.D.[1], Jong-Hwa Kim, M.D.[1]

Departments of [1]Obstetrics and Gynecology, [2]Radiology and Center for Imaging Science, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

The Vein of Galen aneurysmal malformation (VGAM) is a rare congenital malformation that represents less than 1% of the cerebral arteriovenous malformations of neonates. Prenatal diagnosis of VGAM is possible by identifying enlarged veins and arteriovenous shunts with a turbulent flow in cranial ultrasound scan. Furthermore, fetal MRI can provide correct diagnosis and additional useful information about secondary intracranial pathology and other anomalies associated with poor prognosis. A large prenatally detected VGAM is likely to result in a heart failure either during the antenatal period or soon after birth. In particular, the cessation of blood flow to a low-resistance placenta after birth increases the amount of vascular shunt through VGAM, causing rapid hemodynamic decompensation, and eventually death. In this report, we present the prenatally diagnosed case of a vein of Galen aneurysm with a severe heart failure, which was aggravated after birth, showed a dismal outcome despite endovascular coiling. Clinical considerations are also discussed.

Key words: Vein of Galen aneurysmal malformation, Heart failure, Prenatal diagnosis, Endovascular embolization, Multidisciplinary approach

INTRODUCTION

The vein of Galen aneurysmal malformation (VGAM) in a neonate is a rare intracranial vascular malformation with a prevalence of less than 1% of all neonatal intravascular vascular malformations.[1] It is thought to be resulted from the development of an arteriovenous connection between primitive choroidal vessels and the median prosencephalic vein of Markowski. The natural history and the treatment are still unclear and the overall clinical outcome is reported to be poor.[2,3]

Prior to the advent of ultrasonography, prenatal diagnosis of cerebral vascular malformation is reported to be rare, therefore the diagnosis of this condition was made at postmortem examination in approximately 85% of the cases.[4] With the advancement of diagnostic technique, prenatal ultrasound provides a rapid and a noninvasive method for diagnosing the condition. Recently, fetal MRI has been used to identify structural cerebral damage and to offer detailed information about the intracranial vascular structure. Early detection of prognostic

투고일 : 2011. 10. 13. 수정일 : 2011. 12. 15. 확정일 : 2011. 12. 20.
주관책임자 : 오수영
E-mail : ohsymd@skku.edu

factors including the presence of fetal cardiac insufficiency and brain damage are known to be important to predict clinical outcome and subsequent counseling for the further treatment. Recently, the introduction of endovascular intervention has significantly improved the prognosis, both in terms of mortality and morbidity.[5,6] However, in severe case of VGAM with severe cardiac decompensation or known parenchymal damage in fetal brain, even early and sufficient endovascular embolization is unlikely to alter the fetal outcome.[7,8] In this report, we present the case of a large VGAM diagnosed prenatally, which was complicated by severe progressive cardiomegaly in utero and in early neonatal period and showed a dismal outcome despite endovascular coiling in neonatal period.

CASE REPORT

A 33-year-old pregnant women (37^{+4} weeks' gestation, Parity; 0-0-0-0) was referred to Samsung Medical Center for a fetal

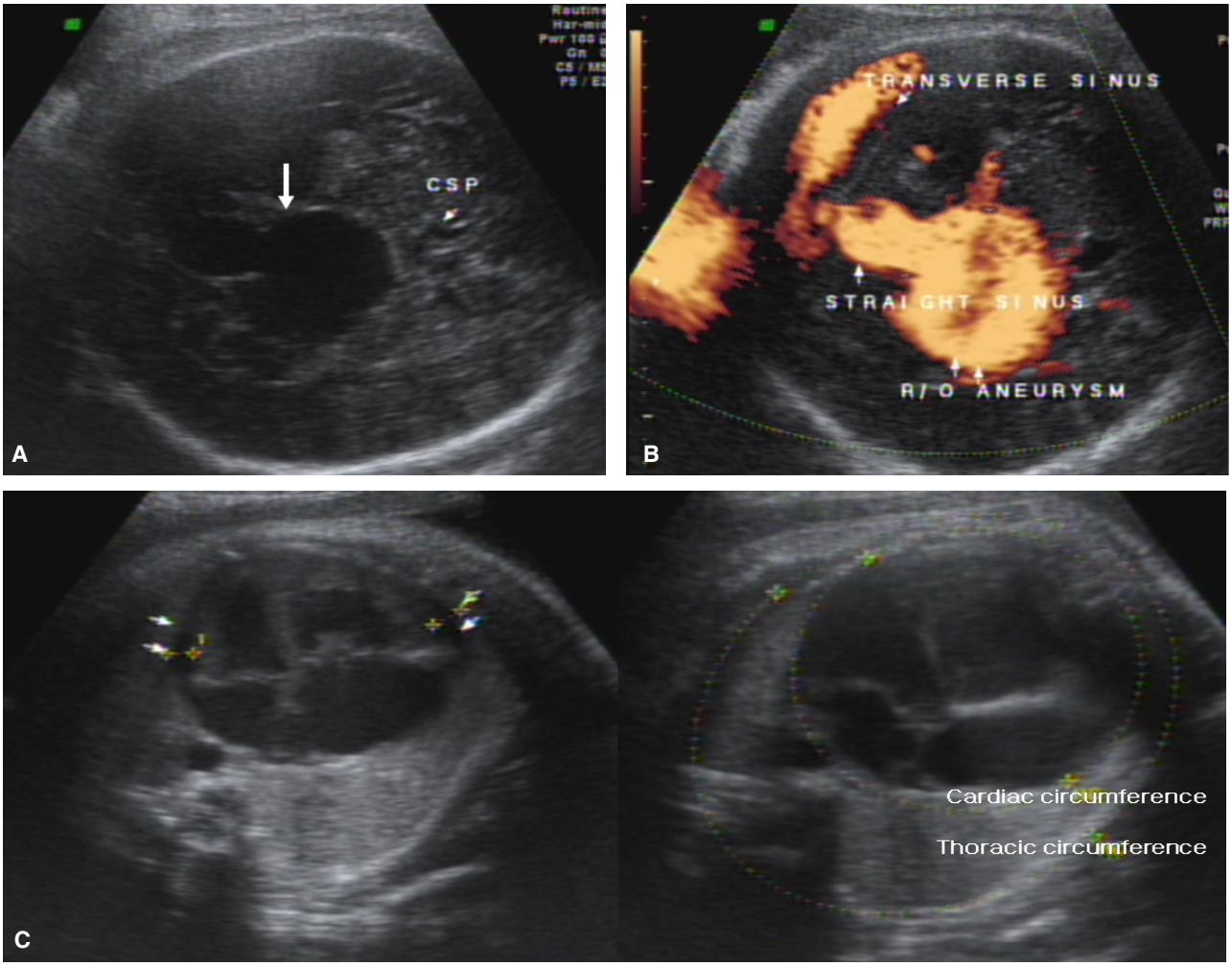

Fig. 1. Two-dimensional Ultrasound images at 37^{+4} weeks of gestation. (A) Intracranial vascular mass measuring in 3.2×3.2 cm. (B) Doppler imaging showing a turbulent flow and a blood draining into an enlarged straight sinus, suggesting a vein of Galen aneurysm. (C) Severe cardiomegaly with pericardial effusion, reflecting an advanced fetal congestive heart failure (left). Cardiac circumference/thoracic circumference ratio (cardiothoracic ratio) of 73% (right).

evaluation of severe cardiomegaly and an associated intracranial vascular malformation. There were no abnormal findings on ultrasound until 36 weeks of gestation during her routine prenatal check. At 37^{+0} weeks, severe cardiomegaly, pericardial effusion, and fetal intracranial vascular malformation were detected on an ultrasound. On a conventional ultrasound (Fig. 1) at our institution, a midline tubular anechoic structure (3.2 cm in longest diameter) located above the thalamus was identified. The Color Doppler imaging showed a turbulent flow and the blood draining into an enlarged straight sinus, suggesting a vein of Galen aneurysm. Mild ventriculomegaly was also suspected (atrial diameter of 11.46 mm). Cardiomegaly with pericardial effusion was found (cardiac circumference/thoracic circumference ratio 73%), reflecting an advanced fetal congestive heart failure.

Before transfer complementary fetal MRI (Fig. 2) was performed for a further evaluation. It showed a grossly dilated vein of Galen (3.2 cm in longest diameter) with mild ventriculomegaly (atrial diameter of 12 mm) and significant dilatation of a straight sinus, confluence sinuses, superior sagittal sinus and transverse sinus, suggesting a large shunt volume. From T2 weighted images of fetal MRI, a high signal intensity was observed around periventricular area and the vein of Galen slightly displaced the 3rd ventricle, but there was no structural brain damage. Due to the high- flow feature of the VGAM and the marked cardiomegaly, the prognosis was considered to be poor.

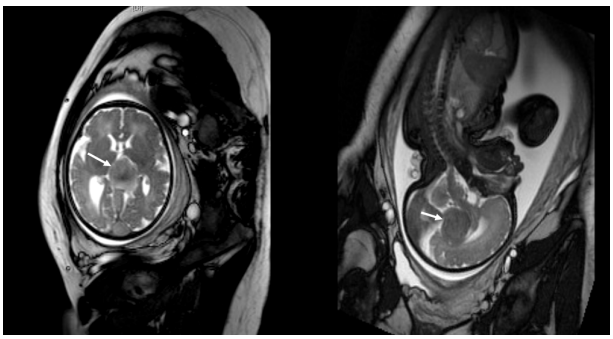

Fig. 2. Fetal MRI showing grossly dilated vein of Galen with mild ventriculomegaly and significant dilatation of sinuses.

At 37^{+5} weeks' gestation, a male 3.28 Kg infant was delivered by an emergent cesarean section due to non-reassuring fetal heart rate. Apgar score of 1 and 5 minutes was 8 and 9 respectively. The neonate was admitted to the neonatal intensive care unit. On the day of birth, a brain ultrasound was performed, re-confirming the prenatal findings: 3.6×3.4×3.0 cm sized VGAM with a turbulent flow by Doppler. The postnatal MRI was followed to assess the degree of VGAM and any structural brain damage compared to prenatal MRI. As a result, the size of the vein of Galen malformation was revealed larger (3.8 cm in longest diameter) combined with a secondary ventriculomegaly (atrial diameter of 14.25 mm) and it also showed low signal on the margin of ventricle, strongly suspicious for intraventricular hemorrhage (Fig. 3). Additionally, there were diffuse excessive white matter change and a scalp hemorrhage around occipital area. 2D echocardiography showed a high output heart failure (with severe TR: pressure gradient =55 mmHg, EF=61.2 %) without cardiac structural malformation. Consultation with neonatologists and pediatric neurologists was conducted and parents were informed of the poor prognosis as a result of a brain ischemia and a heart failure.

During the neonatal care, acidosis was exacerbated from the 3rd day of the birth. Despite the medical treatment including

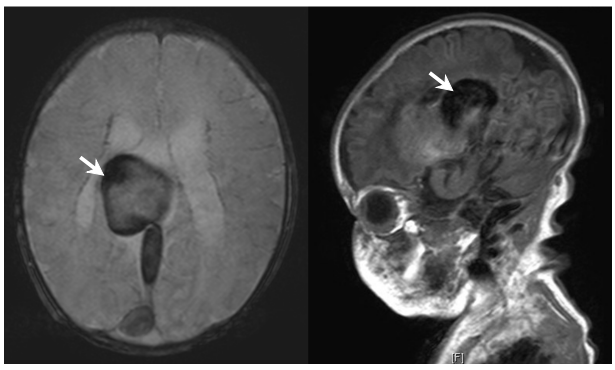

Fig. 3. Postnatal brain MRI of a vein of Galen aneurysmal malformation (right) combined with a secondary ventriculomegaly and showing low signals on the margin of ventricle, strongly suspicious for intraventricular hemorrhage (left).

Table 1. Literature review on prenatally diagnosed vein of Galen aneurysmal malformation

	GAD	Prenatal heart failure	Fetal MRI	Apgar score	Postnatal heart failure	Intervention	Survival
Karadeniz et al[19]	35 wks	Cardiomegaly	VGAM	9,9	Cardiomegaly, TR, pulmonary hypertension	Embolization	Doing well (4 years) Normal neurologic development
Nuutila et al[3]	33 wks	Cardiomegaly	VGAM, ventriculomegaly, cystic leukomalacia	6	Not mentioned	Palliative treatment	Expired (3 days)
Santo et al[8] (two cases)	32 wks	Mild cardiomegaly	VGAM	9, 10	Intractable CHF	Embolization	Expired (9 days)
	32 wks	Mild cardiomegaly, TR	VGAM	6,10	Intractable CHF	Embolization	Expired (16 days)
Koroglu et al[20]	34 wks	Not mentioned	Not performed	NA	Intractable CHF	Embolization	Delay in neurological development (20 months)
Gailloud et al[21]	35 wks	Mildly decreased right ventricular function	Not performed	6,7	Severe RV hypertension on day 2	Embolization	Doing well (4 years)
Allahdin et al[22]	36 wks	Not mentioned	Not performed	0,1,5	Intractable CHF	Palliative treatment	Expired (5 days)
Sepulveda et al[23]	36 wks	Mild cardiomegaly	VGAM	9,9	No sign of CHF	Awaiting embolization	Doing well (3 months)
Messori et al[24]	38 wks	Cardiomegaly	VGAM	9,9	No sign of CHF	Embolization	Doing well (7 years)
Has et al[25]	34 wks	Cardiomegaly	VGAM	6,8	Intractable CHF	Palliative treatment	Expired (3 days)
Maheshwariet al[7]	28 wks	Cardiac failure	VGAM	NA	Intractable CHF	Glue embolization	Expired (29 hours)
Heling et al[1]	33 wks	Mild TR, cardiomegaly	VGAM	NA	Cardiac decompensation	Coil angiography	Expired (5months)
Yuval et al[6] (two cases)	38 wks	Mild cardiomegaly	Not performed	9,10	Mild TR, cardiomegaly	VP shunt	Doing well (19 months)
	33 wks	Cardiomegaly	Not performed	NA	Intractable CHF	Palliative treatment	Expired (2days)

GAD: gestational age at diagnosis, wks: weeks, TR: tricuspid regurgitation, VGAM: vein of Galen aneurysmal malformation, NA:not available, CHF: congestive heart failure, VP shunt: ventriculoperitoneal shunt.

diuretics, the urine output had decreased and feeding intolerance had increased. After multidisciplinary discussion among neonatologists, neurosurgeons and neurointerventionalists, an emergent endovascular coiling was performed. However, brain hemorrhage and intraventricular hemorrhage were aggravated. High output heart failure assessed by 2D echocardiography was more aggravated thereafter. The prognosis of the neonate was considered to be very poor, and after discussion with the parents, palliative care was provided and the infant expired on the 10th day after birth.

DISCUSSION

The VGAM is a rare congenital vascular malformation composed of multiple arterial feeder establishing direct or indirect connections with a large median venous collector. The latter is not the vein of Galen *per se*, but a persistent embryonic channel that normally participates in a formation of the vein of Galen, the median prosencephalic vein of Markowski. The first reported case of VGAM was published in 1937.[9] Although rare in neonates, it had been estimated that VGAM represents approximately 30% of the pediatric vascular malformations.[10]

VGAM can cause severe morbidity and mortality in the early neonatal period and also later (in infant period). In utero, the arteriovenous shunt maintains a low flow rate as the low resistance of the placental vascular bed diminishes the flow through the low resistance aneurysm. However, after birth, the cessation of blood flow to a low-resistance territory such as the placenta suddenly increases the vascular shunt at the level of the head (70% of cardiac output is directed), causing rapid hemodynamic decompensation, heart failure, and eventually death.[11] High output congestive heart failure is the most common clinical manifestation in neonates and may further result in multi-organ failure and death. Other clinical manifestations include macrocephaly, seizures, and occasional intracranial hemorrhage or a focal neurologic deficit in older children and adults.

A prenatal diagnosis of VGAM and heart failure is possible by an ultrasound. During the past decades, prenatal diagnosis of vein of Galen aneurismal malformation has increased, because antenatal ultrasound use became more popular.[6,12] The most of the diagnosis of arteriovenous malformations has been made during the 3rd trimester, with two thirds of the cases diagnosed at>34 weeks of gestation.[11] The earliest reported diagnosis was made at 25 weeks of gestation.[13] The prognosis of our patient was poor because of late detection or late development of disease with rapid progression. On an ultrasound, VGAM appears to have an anechoic tubular intracranial lesion in the midline, situated superior to the cerebellum with high turbulence flow identified by a color Doppler ultrasound. The differential diagnosis includes arachnoid cysts, enlarged third ventricle, brain arteriovenous malformation that drains into the vein of Galen, or an interhemispheric cyst associated with agenesis of the corpus callosum and holoprosencephaly.[14] However, an ultrasound alone cannot show a parenchymal pathology. A clear appreciation of the degree of irreversible cerebral damage by image studies is essential for both therapeutic decision-making and prognosis evaluation. In this situation, MRI can additionally provide valuable information about other anomalies, heart failure and secondary brain pathology, all associated with poor prognosis.[3,12,15] So, we summarized the prenatal MRI finding, the presence of heart failure in prenatal and postnatal period and the clinical course of VGAM (Table 1).

In our patient, brain ultrasound on the day of birth showed findings of progression of disease compared with prenatal images. Technically, sedation of neonate is required for taking MRI, which might be undesirable for unstable neonate. So we conducted prenatal MRI for evaluation at first and conducted postnatal MRI additionally for identifying brain damage after consulting pediatric neurosurgeon. Recently, it was reported that 3D ultrasound can identify both the aneurysm and its vascular connections in spatial orientation in neonate, which is believed to be useful in angiographic embolization af-

ter birth.

Physiopathologic consequences of the VGAM are high flow left to right shunt, vascular steal in the brain and cerebral venous hypertension. Therefore, the severity of a congestive heart failure, which depends on the volume of arteriovenous shunting and the brain injury are known as primary prognostic factors[15] and it should be the main focus of the antenatal imaging. In their absence, the prognosis is difficult to assess. Early identification of these factors is critical, as it allows early medical treatment of heart failure and reduces the need for complex diagnostic neuroradiological modalities in critically ill patients.

The proper management of VGAM is still controversial and considered dismal in severe cases. The result of neurosurgery was reported to be poor because of the complexity of the vascular malformation. The interventional endovascular embolization has improved the outcome and has currently emerged as the primary treatment.[16] In general, patients with severe cardiogenic shock, multisystem organ failure, and/or irreversible brain damage have a very poor prognosis and are not candidates for endovascular intervention and patients with mild cardiac overload can be medically managed until embolization is performed at 4 to 5 months of age.[17] However, excessive delay may lead to permanent impairment of the cerebrospinal fluid hydrodynamic, after which even successful correction of the shunts would not result in hydrocephalus regression. Recently, Lasjaunias and colleagues proposed a 21-point score, score is based on cardiac function, cerebral function, respiratory function, hepatic function, and renal function to help guide management of VGAM in neonates.[18]

With this background, antenatal counseling is still difficult and identifying those patients with improved outcome after embolization remains a challenge. In the clinical point of view, whatever the treatment modality is, a close cooperation between neurointerventionalists, neurosurgeons, neonatologists, anesthesiologists, and obstetricians is strongly required to treat this challenging disease. Additionally, developments of screening tools that can prenatally predict the prognosis of VGAM associated with a heart failure seem to be essential for an early multidisciplinary management and a parent counseling.

참고문헌

1. Heling KS, Chaoui R, Bollmann R. Prenatal diagnosis of an aneurysm of the vein of Galen with three-dimensional color power angiography. Ultrasound Obstet Gynecol 2000; 15: 333-6.
2. Heuer GG, Gabel B, Beslow LA, Stiefel MF, Schwartz ES, Storm PB, et al. Diagnosis and treatment of vein of Galen aneurysmal malformations. Childs Nerv Syst 2010; 26: 879-87.
3. Nuutila M, Saisto T. Prenatal diagnosis of vein of Galen malformation: a multidisciplinary challenge. Am J Perinatol 2008; 25: 225-7.
4. Holden AM, Fyler DC, Shillito J Jr, Nadas AS. Congestive heart failure from intracranial arteriovenous fistula in infancy. Clinical and physiologic considerations in eight patients. Pediatrics 1972; 49: 30-9.
5. Mitchell PJ, Rosenfeld JV, Dargaville P, Loughnan P, Ditchfield MR, Frawley G, et al. Endovascular management of vein of Galen aneurysmal malformations presenting in the neonatal period. AJNR Am J Neuroradiol 2001; 22: 1403-9.
6. Yuval Y, Lerner A, Lipitz S, Rotstein Z, Hegesh J, Achiron R. Prenatal diagnosis of vein of Galen aneurysmal malformation: report of two cases with proposal for prognostic indices. Prenat Diagn 1997; 17: 972-7.
7. Maheshwari PR, Pungavkar SA, Narkhede P, Patkar DP. Image in radiology. Vein of Galen aneurysmal malformation: antenatal MRI. J Postqrad Med 2003; 49: 350-1.
8. Santo S, Pinto L, Clode N, Cardoso E, Marques JP, Melo A, et al. Prenatal ultrasonographic diagnosis of vein of Galen aneurysms-report of two cases. J Matern Fetal Neonatal Med 2008; 21: 209-11.
9. Jaeger R, Forbes RP, Dandy WE. Bilateral congenital cerebral arteriovenous communication aneurysm. Trans Am Neurol Ass 1937; 63: 173-6.
10. Long DM, Seljeskog EL, Chou SN, French LA. Giant arteriovenous malformations of infancy and childhood. J Neurosurg 1974; 40: 304-12.
11. Paternoster DM, Manganelli F, Moroder W, Nicolini U. Prenatal diagnosis of vein of Galen aneurysmal malformations. Fetal Diagn Ther 2003; 18: 408-11.
12. Campi A, Scotti G, Filippi M, Gerevini S, Strigimi F, Lasjaunias P. Antenatal diagnosis of vein of Galen aneurysmal malformation: MR study of fetal brain and postnatal follow-up. Neuroradiology 1996; 38: 87-90.
13. Ballester MJ, Raga F, Serra-Serra V, Bonilla-Musoles F. Early prenatal diagnosis of an ominous aneurysm of the vein of Galen by color Doppler ultrasound. Acta Obstet Gynecol Scand 1994; 73: 592-5.
14. Epelman M, Daneman A, Blaser SI, Ortiz-Neira C, Konen O, Jarrin J, et al. Differential diagnosis of intracranial cystic lesions

at head US: correlation with CT and MR imaging. Radiographics 2006; 26: 173-96.
15. Brunelle F. Brain vascular malformations in the fetus: diagnosis and prognosis. Childs Nerv Syst 2003; 19: 524-8.
16. Friedman DM, Verma R, Madrid M, Wisoff JH, Berenstein A. Recent improvement in outcome using transcatheter embolization techniques for neonatal aneurysmal malformations of the vein of Galen.Pediatrics 1993; 91: 583-6.
17. Recinos PF, Rahmathulla G, Pearl M, Recinos VR, Jallo GI, Gailloud P, et al. Vein of Galen Malformation: Epidemiology, Clinical Presentation, Management. Neurosurg Clin N Am 2012; 23: 165-77.
18. Lasjaunias PL, Chng SM, Sachet M, Alavarez H, Rodesch G, Garcia-Monaco R. The management of vein of Galen aneurysmal malformations. Neurosurgery 2006; 59: S184-94.
19. Karadeniz L, Coban A, Sencer S, Has R, Ince Z, Can G. Vein of Galen aneurysmal malformation: prenatal diagnosis and early endovascular management. J Chin Med Assoc. 2011; 74(3): 134-7.
20. Koroglu M, Cil B, Yesildag A, Baykal B, Cekirge S, Oyar O. Prenatal diagnosis of intracranial pial arteriovenous fistula and endovascular treatment during the neonatal period. Diagn Interv Radiol 2006; 12: 64-7.
21. Gailloud P, O'Riordan D P, Burger I, Lehmann CU. Confirmation of communication between deep venous drainage and the vein of galen after treatment of a vein of Galen aneurysmal malformation in an infant presenting with severe pulmonary hypertension. AJNR Am J Neuroradiol 2006; 27: 317-20.
22. Allahdin S, Reid B, MacDonald I. Pregnancy complicated by a fetus with aneurysm of the vein of Galen and microcephaly. J Obstet Gynaecol 2005; 25: 396-7.
23. Sepulveda W, Vanderheyden T, Pather J, Pasquini L. Vein of galen malformation: prenatal evaluation with three-dimensional power Doppler angiography. J Ultrasound Med 2003; 22: 1395-8.
24. Messori A, Polonara G, Salvolini U. Prenatal diagnosis of a vein of Galen aneurysmal malformation with fetal MR imaging study. AJNR Am J Neuroradiol 2003; 24: 1923-5.
25. Has R, Gunay S, Ibrahimoglu L. Prenatal diagnosis of a vein of galen aneurysm. Fetal Diagn Ther 2003; 18: 36-40.

「국문초록」

Vein of Galen aneurysmal malformation (VGAM)은 신생아 뇌혈관 기형의 1% 이하로 드문 기형이다. VGAM은 초음파상 태아의 심비대, 확장된 정맥과 뇌동정맥 shunt로 진단할 수 있다. Fetal MRI는 VGAM의 산전 진단시 이차적으로 발생한 뇌병변과 예후와 관련된 동반 기형을 확인하는 데 도움이 된다. VGAM은 산전 또는 출생 직후에 심부전에 빠지게 되며, 특히 출생 후 저항이 낮은 태반으로 흐르는 혈류가 중단된 후 VGAM을 통한 혈류량이 늘어나면서 사망률이 증가된다. 저자들은 산전에 진단된 VGAM 태아로 중증의 심부전을 동반하여 출생 후 사망 한 증례를 보고하는 바이다.

중심 단어: Vein of Galen aneurysmal malformaltion (VGAM), 심부전, 산전 진단, 내혈관치료, Multidisciplinary approach