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Case Report

Implications of serial magnetic resonance imaging in the management of a newborn with vein of Galen aneurysmal malformation and a review of the relevant literature

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ABSTRACT

Background. Despite advanced endovascular methods and comprehensive intensive care in the neonatal vein of Galen aneurysmal malformation, overall mortality ranges between 37-63% in treated patients with 37-50% of survivors possessing poor neurologic outcomes. These findings stress the need for more accurate and timely recognition of the patients who may and may not benefit from aggressive intervention.

Case. This case report presents a newborn with a vein of Galen aneurysmal malformation whom antenatal and postnatal follow-up included serial magnetic resonance imaging (MRI) including diffusion-weighted series.

Conclusions. Given the experience from our current case and in light of the relevant literature, it is plausible that diffusion-weighted imaging studies may widen our perspective on dynamic ischemia and progressive injury occurring within the developing central nervous system of such patients. Meticulous identification of patients may favorably influence the clinical and parental decision on early delivery and prompt endovascular treatment versus aiding avoidance of further futile interventions both antenatally and postnatally.

Key words: vein of Galen aneurysmal malformation, magnetic resonance imaging, diffusion-weighted imaging, newborn.

Vein of Galen aneurysmal malformation (VGAM) is a relatively rare (1:25000 live births) congenital arteriovenous malformation (AVM) of the central nervous system (CNS) associated with high mortality rates in the neonatal period.[1,2] Endovascular treatment (ET) is the therapy of choice in symptomatic newborns and aims to relieve life threatening high-flow heart failure.[1,2] Despite recent improvements and comprehensive follow-up, survival rates as well as overall clinical outcomes are relatively poor, with neurodevelopmental disorders frequently seen among survivors, raising questions regarding our understanding of the disease.[1-4] This case report highlights the importance of individualized antenatal and postnatal magnetic resonance imaging (MRI) in particular, diffusion-weighted imaging (DWI) in newborns with VGAM, while addressing its possible contribution to clinical and parental decision making both antenatally and postnatally.

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Case Report

A singleton male neonate weighing 2530 grams (50-75th percentile) with a prenatally diagnosed VGAM was born to a 29-year-old mother by cesarean section at 34+5 weeks of gestation. The neonate was admitted to the neonatal intensive care unit (NICU) for further evaluation immediately after endotracheal intubation and stabilization of vital signs.

Antenatal diagnosis of VGAM was made incidentally during routine antenatal ultrasound screening at 23 weeks of gestation at another institution, where early termination of pregnancy was not considered. The patient was further evaluated at a later stage during pregnancy at 31 weeks of gestation in our institution with fetal MRI (Fig. 1), which revealed a significantly dilated median prosencephalic vein in continuum with a large choroidal type VGAM. DWI studies showed restricted diffusion with corresponding low signal areas on apparent diffusion coefficient (ADC) maps consistent with acute ischemia of the left occipital and the left posterior parietal regions.

The first examination showed a lethargic appearing baby with general hypotonicity and hypoactive newborn reflexes, an occipitofrontal circumference of 33 cm (75-90th percentile), a tense anterior fontanelle with a continuous murmur and hyperactive precordium with 2/6 systolic murmur on auscultation. Tachycardia (heart rate: 180/minute) and tachypnea (respiratory rate: 70/minute) with sub- and intercostal retractions were remarkable. Chest X-ray revealed cardiomegaly and ruled out concomitant severe respiratory parenchymal disease. Bedside echocardiography demonstrated supra-systemic pulmonary hypertension (right ventricle systolic pressure: 75 mmHg) in concurrence with dilated right-sided cavities of the heart, a wide patent ductus arteriosus (PDA) with right-to-left shunt and a retrograde aortic flow. Bedside cranial Doppler ultrasound imaging confirmed the diagnosis of VGAM.

Time-of-Flight MRI performed on the 2nd day of life showed the choroidal type VGAM located to the left of the midline, with a venous pouch of 4.4 cm in craniocaudal direction and multiple arterial feeders predominantly from the left posterior cerebral artery (PCA) and left middle cerebral artery (MCA) (Fig. 2A and 2B). When compared to fetal MRI findings, the venous pouch and feeding vessels had increased in

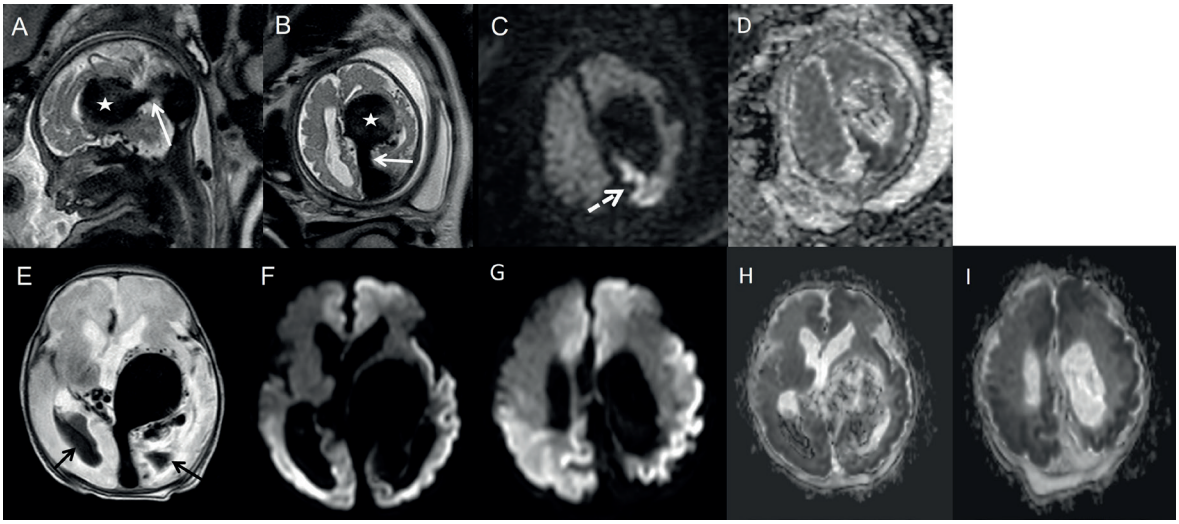

Fig. 1. Cranial MRI studies of the patient performed in-utero **(A-D)** and following endovascular treatment at 7 days of age **(E-I)**. Enormously enlarged Torcula and median prosencephalic vein (arrow) ending in a venous aneurysm (asterix) are displayed on sagittal **(A)** and axial **(B)** T2-weighted images. Restricted diffusion on average diffusion-weighted image **(C)** and corresponding low signal on ADC map **(D)** suggestive of cytotoxic edema due to acute ischemia of the left medial parietal cortex (dashed arrow) is observed. Following endovascular treatment, there is an intraventricular hemorrhage layering in the occipital horns of the lateral ventricles (black arrows) and accompanying hydrocephalus are shown on axial T2-weighted image **(E)** as well as bilateral extensive hemispheric cortical diffusion restriction **(F-I)** on both average diffusion-weighted image and ADC maps this time.

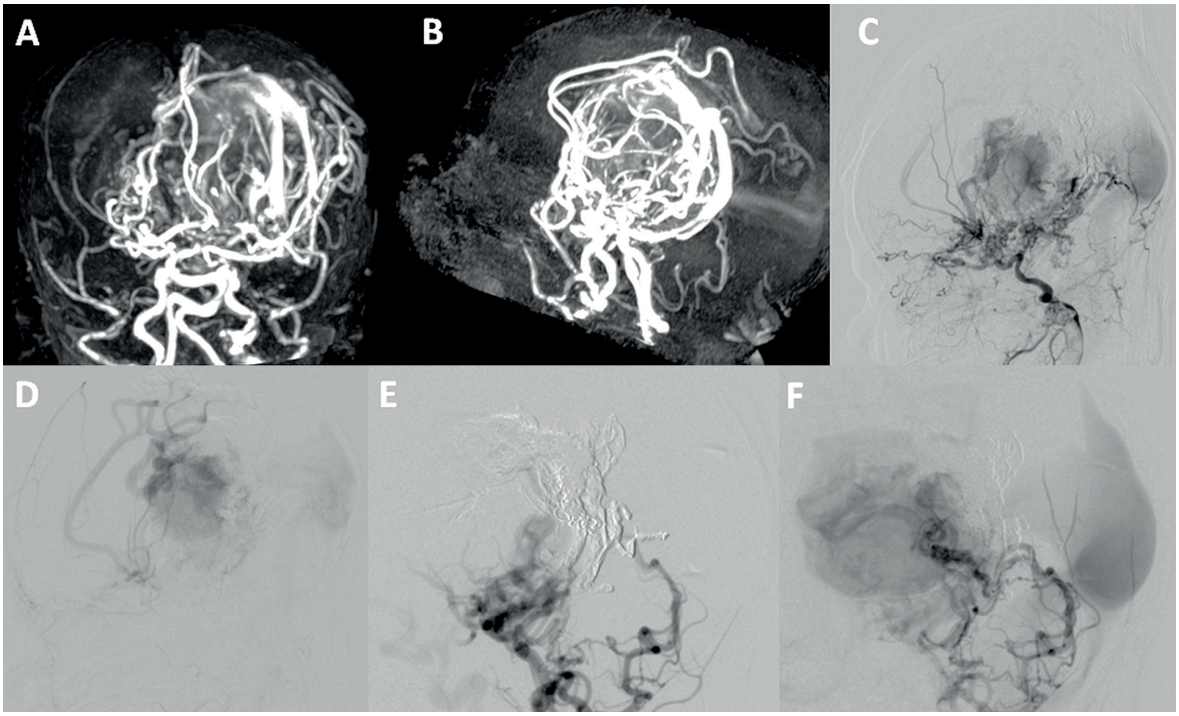

Fig. 2. Pre-embolization reconstructions (maximum intensity projection) of time-of-flight cranial MR angiogram in anteroposterior **(A)** and lateral **(B)** projections show a diffuse conglomerate of arteries and drastic enlargement of a venous pouch. Both anterior cerebral, posterior cerebral arteries and all choroidal arteries supply the arteriovenous malformation. On DSA obtained after the third session of embolization, on the lateral views of the left **(C)** and right **(D)** internal carotid artery injections as well as early **(E)** and late **(F)** phases of left vertebral arteriograms, there is still a significantly large residual arteriovenous malformation.

size, resulting in compression of the left lateral ventricle and left mesencephalon with a 1-cm subfalcine shift from the midline to the right. Both lateral ventricles were dilated.

The child continued to deteriorate and developed signs of circulatory collapse despite the administration of maximum cardiopulmonary support including dopamine, adrenaline and high frequency oscillatory ventilation. Given the inefficiency of medical treatment, a multidisciplinary consensus was reached to commence with ET. On the 3rd, 7th and 10th days of life, three sessions of ET were performed under general anesthesia and fluoroscopic guidance with full cardiorespiratory support were given by both the neonatology and anesthesia teams. In all sessions, the umbilical artery was used for arterial vascular access. Arterial feeders arising from the left PCA, left internal carotid artery (ICA) and left anterior cerebral artery (ACA) were super-selectively catheterized in the 1st, 2nd and 3rd procedures, respectively (Fig. 2). Embolization was achieved with coils, n-butyl cyanoacrylate/lipiodol mixture (Glue) and Squid (Emboflu, Switzerland). Angiograms were performed with a water-soluble nonionic iodine-based contrast material with a maximum dose limited to 7 ml per kilogram for each session.

Cranial MRI was performed after the 2nd session of ET for post-treatment follow-up imaging and for guidance regarding further endovascular intervention. It showed the decreased maximum size of the venous pouch with a caudocranial length of 3.2 cm, a small intraventricular extravasation and persistence of ventricular dilatation as well as midline shift. DWI studies and ADC maps showed increased extension of diffusion restriction of the hemispheric cortices bilaterally (Fig. 1) suggesting the

futility of the 2 previous treatments. As DWI restrictions symmetrically involved the main cortical areas, and as these findings were in part present in the prenatal imaging and finally since such restrictions were also present in the right anterior circulation which had not been accessed during any of the interventions, it was concluded that the MRI findings represented a severe and progressive arterial insufficiency involving the cerebral hemispheres. Although neuroimaging findings suggested undesirable neurologic outcomes, the 3rd session of ET was pursued upon request of the family who understood the poor prognosis of the child and also in accordance with the current legislation demanding that all that could be done unless brain death was officially announced.

The third neuroradiological intervention and meticulous neonatal intensive care failed an improvement in neurological findings. Serial echocardiographic examinations after each ET session showed only partial but no significant improvement in heart failure. Respiratory distress followed a refractory course, precluding extubation. The neonate was lost due to cardiorespiratory failure on the 14th day of life.

Discussion

Vein of Galen aneurysmal malformation (VGAM) is the most commonly seen congenital AVM in the neonatal period with morbidity and mortality reaching almost 100% in the absence of accurate management.[1,2,5] Despite advanced endovascular methods and thorough intensive care; overall mortality ranges between 37-63% (13-35% with successful ET).[1-7] Furthermore, 37-50% of survivors possess poor neurologic outcomes, while ones with presumed good outcome experience some neurocognitive or functional deficits in the later stages of their lives.[1-4,6,7] These findings indicate the need for a more accurate identification of the VGAM patients who will and will not benefit from aggressive intervention. While further comparative research is needed, it is plausible that antenatal and postnatal conventional MRI combined with DWI may potentially allow us to ascertain the dynamic progression and foresee the prognosis in patients with VGAM.

Despite being the first line imaging modality, antenatal ultrasound imaging may be insufficient to detect ischemic injury due to VGAM of the fetal brain, while this can be unequivocally demonstrated by antenatal conventional MRI.[4,8,9] Antenatal DWI studies in particular may stand out to have a potential diagnostic and prognostic importance in the evaluation of VGAM to detect impending or recently established brain ischemia of the fetal brain, where even the conventional MRI is still not yet informative.[9] Fetal or neonatal MRI lesions among patients with VGAM are reported to be predominantly supratentorial in location and usually bilateral.[10] Presence of diffusion restriction is most commonly but not exclusively due to underlying ischemia, neuronal migration disorders are also included in the differential diagnosis.[11] Venous congestion and venous hypertension seem to be the main underlying pathogenesis for the development of cerebral ischemia in patients with VGAM. In some cases, venous hypertension and subsequent ischemic changes can be so severe that widespread white matter destruction and cerebral atrophy ensue.[12] Several other factors related to ET can play role in the progression of cerebral ischemic lesions: Connection between the deep venous system and the venous pouch, inadvertent embolization of the venous pouch with resultant venous hypertension and both venous ischemic and hemorrhagic complications or rapid subsequent expansion of the thrombosis through the venous pouch after successful total embolization of the fistula especially when performed in a single session.[13,14]

Since the findings of a severe brain injury is a predictor of poor neurological outcome in the later stages of life, antenatal conventional MRI in addition to DWI studies may serve to understand the gravity of acutely presenting, progressively worsening or already irreversibly pronounced ischemic fetal brain damage.[4,7-9] This may allow a more precise and timely

interpretation of the prognosis and possible overall outcome of the developing brain, thereby favorably influencing the clinical and parental decision on early delivery and prompt intervention to prevent further injury versus the termination of the pregnancy which may be applicable to prevent unnecessary aggressive invasive interventions.[4,7-9]

Many referral centers do not offer ET in patients with widespread and irreversible brain damage detected by postnatal conventional MRI (massive brain infarct, diffuse loss of parenchymal volume, encephalomalacia) due to expectation of a poor neurologic outcome, although possible future modulating effects of neuroplasticity is taken into consideration intensively.[3,4,6-9] On the other hand, withholding further interventions may not be an option in cases without concrete radiologic evidence of progressive worsening unresponsive to therapy, given the fact that legislations of many countries (including Turkey) demand not to withdraw medical care in such patients as long as the family prefers the continuation of treatment. Serial postnatal DWI studies suggesting progressively worsening and expanding brain ischemia beyond a level that cannot be ameliorated by neuroplasticity may add weight to findings of conventional MRI, providing convenience in decision making and aiding avoidance of further futile interventions.[6]

In conclusion, given the experience from our current case and in the light of literature, conventional MRI strengthened with DWI studies may widen our perspective in the understanding of the dynamic processes involved in the developing CNS of patients with VGAM while aiding clinical and parental decision antenatally and postnatally. Further reports and studies in the field are also required for accurate and safe guidance.

Ethical approval

Informed consent has been obtained from the mother of the baby.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: AA, AA, ŞY; data collection: AA, SB, HKKO; analysis and interpretation of results: HKKO, AA, ŞY; draft manuscript preparation: AA, SB, HKKO, AA, ŞY. All authors reviewed the results and approved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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