Prenatal MR findings in a case of aneurysm of the vein of Galen

Abstract We describe a case of aneurysm of the vein of Galen (AVG), which was diagnosed by intrauterine US, MRI and MRA. The baby girl was born at 35 weeks’ gestation. She had severe clinical symptoms at birth and died at 29 h of age from intractable congestive heart failure. Intrauterine US detected an intracranial aneurysm and cardiomegaly due to excessive arteriovenous shunting. Intrauterine MRI (SSFSE) confirmed the diagnosis of AVG, and intrauterine MRA (2D-TOF) successfully demonstrated the anatomical structure of the AVG. MRA may be a useful additional sequence to evaluate AVG, and 2D-TOF is thought to be an appropriate technique for scanning fetal AVG.

Introduction Aneurysm of the vein of Galen (AVG) is a rare vascular anomaly in children. The clinical manifestations of AVG depend on the volume of arteriovenous shunting, ranging from a severe fetal-neonatal form to a mild childhood form [1]. In the severest form, congestive heart failure (CHF) has already progressed in utero and the mortality is high. The huge volume of shunting causes hypoperfusion of intact brain parenchyma, the ‘steal’ phenomenon, and the prognosis is also poor when brain injury is already present.

Recently, SSFSE (single-shot fast-spin-echo) has been widely used for detecting fetal anomalies, and the diagnosis of AVG has been made by both ultrasound (and/or CT) and SSFSE before birth [1, 2]. Intrauterine MRA
was thought to be useful for revealing the detailed vascular structure of AVG, but some technical problems remained. In this study, 2D-TOF (two-dimensional time-of-flight) MRA was used for scanning the fetal AVG and successfully demonstrated its vascular structure.

Case report

A 28-year-old woman, P0 G0, was referred to our hospital because of fetal cardiomegaly at 35 weeks’ gestation. US demonstrated severe fetal cardiomegaly, with enlargement of both right atrium and superior vena cava, and an intracranial aneurysm. Intrauterine MRI (SSFSE: TE, 93 ms; 1/2 NEX; acquisition matrix, 256 × 192; field of view, 34 cm; slice thickness, 5–7 mm; interslice gap, 2–3 mm; acquisition time, 2 s/slice; torso array coil) and MRA (2D-TOF: TR/TE, 12/4.3 ms; flip angle, 30°; NEX 1; field of view, 40 cm; acquisition matrix, 256 × 160; acquisition time, 2 s/image; 33 slices; slice thickness of original image, 3 mm; torso array coil) were performed. MRI demonstrated a large intracranial aneurysm, confirmed as AVG from its vascular structure and location (Figs. 1, 2). MRA demonstrated four feeding arteries (mainly from the posterior choroidal artery), the aneurysm itself and a falcor sinus draining to the superior sagittal sinus (Figs. 3, 4). No obvious brain injury was detected by the MRI. The fetus showed diminished movements and echocardiography revealed poor ejection fraction.

The following day, a baby girl was born by caesarean section because of fetal distress due to CHF. Apgar score was 2 at 5 min. She showed cyanosis, systemic hypotension, tachycardia and metabolic acidosis. In spite of all supportive therapy her clinical condition progressively deteriorated and she died at 29 h of age. Although catheter embolisation via the posterior cerebral artery was planned, this was impossible because of her poor clinical condition. Autopsy demonstrated that the vascular structure of the AVG was identical to the images obtained by intrauterine MRA. The straight sinus was absent. There were some micro-infarctions in the periventricular brain parenchyma.

Discussion

AVG is rare, but is one of the major arteriovenous anomalies in children [1]. In the fetal-neonatal form with severe clinical symptoms of CHF, as in the case reported here, the prognosis is extremely poor. There are two major factors that affect the prognosis of AVG. One is the severity of CHF, which depends on the volume of arteriovenous shunting [3]; the other is the brain injury resulting from the ‘steal’ phenomenon [4]. Intrauterine MRI and US (including Doppler) can confirm the diagnosis of AVG and assess the severity of CHF and brain injury. As physical supportive therapies are limited, catheter embolisation is the first choice for surgical treatment of AVG. In order to improve the prognosis of the severe form of AVG, it is desirable that embolisation is performed early to prevent irreversible CHF and brain injury. In the present case there was no opportunity to treat the baby because she was referred late to our hospital and already showed irreversible CHF, even in utero.

Accurate information about the vascular anatomy of the AVG is a prerequisite for catheter embolisation. SSFSE MRI is a fast imaging technique, which is suitable for imaging fetal anatomy because it can ‘freeze’ fetal movements. However, it is also necessary to demonstrate the feeding arteries and the venous drainage of fetal AVG. Non-invasive MRA is used postnatally for demonstrating intracranial vascular disease, in preference to digital subtraction angiography, but there is no report of intrauterine MRA detecting fetal AVG, probably because intrauterine MRA has some technical problems, which remain to be solved. Nevertheless, intrauterine MRA may be an additional sequence, which