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CASE REPORT

Ethylenevinylalcohol copolymer (Onyx-18) used in endovascular treatment of vein of Galen malformation

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Abstract
Background Vein of Galen malformations (VGM) are rare congenital arteriovenous fistulas that usually present with heart failure in the neonate. Endovascular treatment options in the past have utilized coils, balloons, and acrylics.
Case report We present, for the first time in the literature, a case of an infant with VGM treated initially with staged coil embolizations followed 1 year later by the transarterial and transvenous catheter based injection of Onyx-18 (ethylenevinylalcohol copolymer) in a single treatment session. The fistula was eliminated, and the infant's cardiopulmonary symptoms were improved.

Keywords Vein of Galen malformation · Onyx-18 (ethylenevinylalcohol copolymer) · Arteriovenous fistula Heart failure · Endovascular

Introduction

Vein of Galen malformation (VGM), a term used to refer to a group of vascular anomalies sharing a dilated vein of Galen or congenitally enlarged and fistulous median prosencephalic vein, often presents in the neonate with congestive heart failure (CHF) and a cranial bruit [3]. Heart failure because of low vascular resistance within the malformation and subsequent right heart failure and pulmonary hypertension is the major cause of morbidity and mortality. Treatment options are geared toward improving cardiac function by reducing flow through the arteriovenous fistula [8]. These include medical therapy, surgery, radiosurgery, and endovascular therapy. Previous endovascular techniques report transarterial and transvenous routes for embolization of the fistulous connections using embolic agents such as balloons, acrylics, and coils [2, 5–7]. We report for the first time in the literature the use of Onyx-18® (ethylenevinylalcohol copolymer, Micro Therapeutics, Plymouth, MN) via a transarterial and transvenous route in the treatment of an infant with a VGM.

Onyx, a liquid embolic material, received Food and Drug Administration approval in July, 2005. The system (ethylene–vinyl copolymer, dimethyl sulfoxide solvent, and micronized tantalum powder) is a bio-compatible liquid polymer that precipitates and solidifies upon contact with blood, thus forming a soft, spongy embolus. In the USA, it is being used on label in the endovascular treatment of arteriovenous malformations [4] and in Europe for the management of arteriovenous malformations and cerebral aneurysms [1].

Case report

We present a case of a baby boy who was born via Cesarean section at 38 weeks gestational age with an uncomplicated delivery. Apgar scores were 8 and 9 at 1 and 5 min, respectively. A cardiac murmur and tachypnea were noted at birth, prompting an echocardiogram revealing pulmonary

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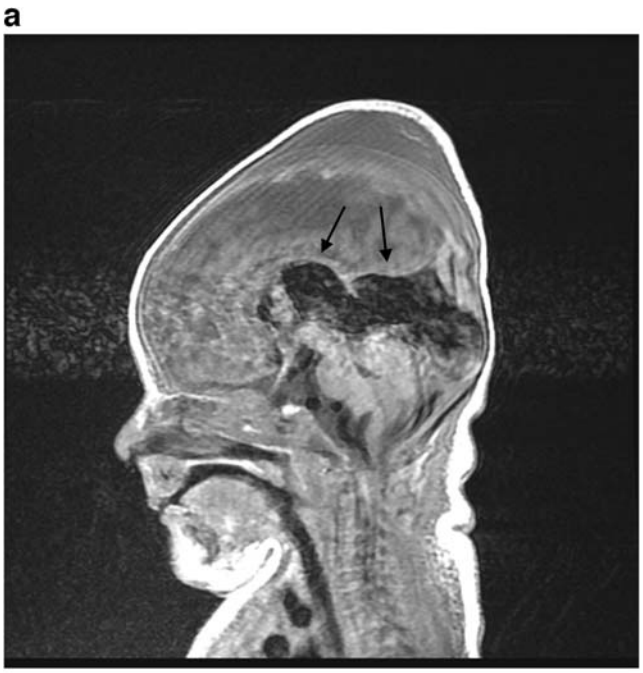
hypertension, poor cardiac contractility, and right ventricular enlargement. Chest X-ray revealed severe cardiomegaly consistent with CHF. The infant was intubated and underwent cranial ultrasound and eventual magnetic resonance imaging (MRI)/magnetic resonance venography (MRV) all of which revealed a VGM (Fig. 1a and b). Cardiopulmonary stabilization was achieved with multiple intravenous drips, and the patient was transferred to our facility. Electroencephalogram



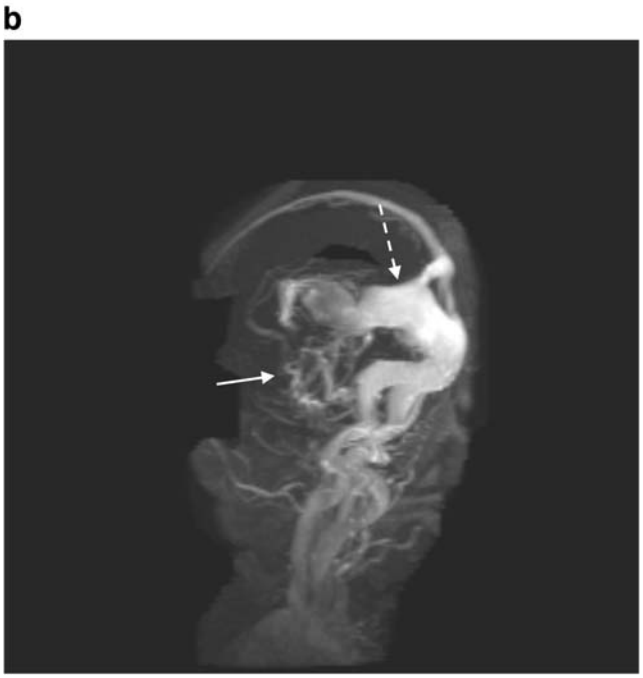


Fig. 1 **a** Sagittal MRI revealing vein of Galen malformation (*arrows*). **b** Sagittal MRV revealing arterial feeders (*white arrow*) and dilated venous structures (*dashed white arrow*) of VGM

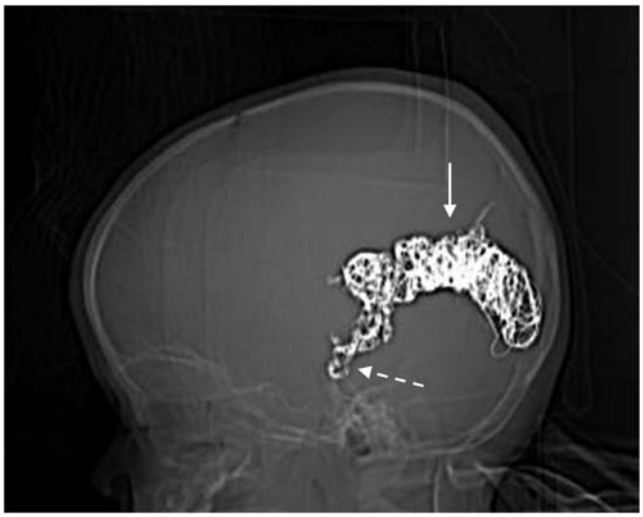


Fig. 2 Scout CT showing coils within the vein of Galen (*white arrow*) and within posterior cerebral arteries (*dashed white arrow*)

was within normal limits on multiple occasions, and clinical examination was otherwise normal. Despite aggressive medical therapy, the baby's heart failure continued to progress over the course of several weeks. Multiple multidisciplinary sessions were held, and a collective decision for endovascular intervention was made.

The infant's endovascular treatment began at the age of 20 days and weight of 3.5 kg. Given the patient's small size and circulating blood volume, contrast load was limited to 10 cm^3 per treatment. The endovascular treatment was intentionally staged to reduce the risk of renal toxicity and intracerebral hemorrhage from significant flow diversion in

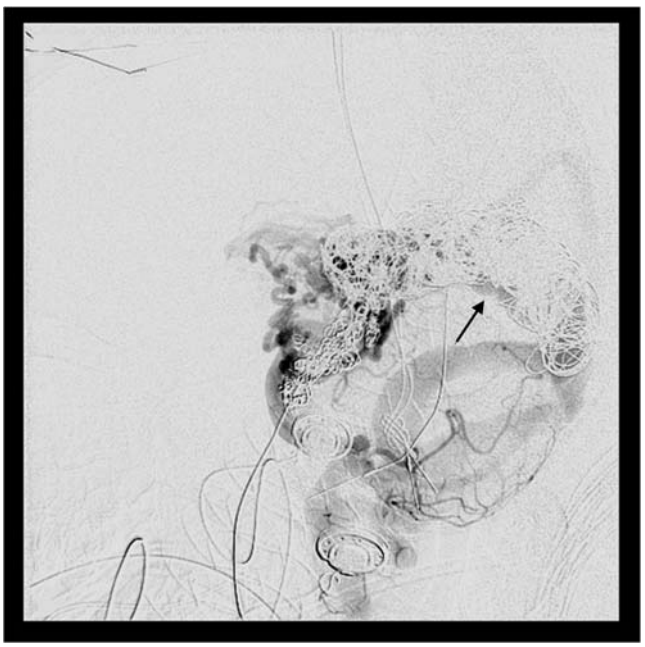


Fig. 3 Lateral view of left vertebral injection postcoiling alone. Note residual flow in draining veins as indicated by contrast within interstices of coil mass (*arrow*)

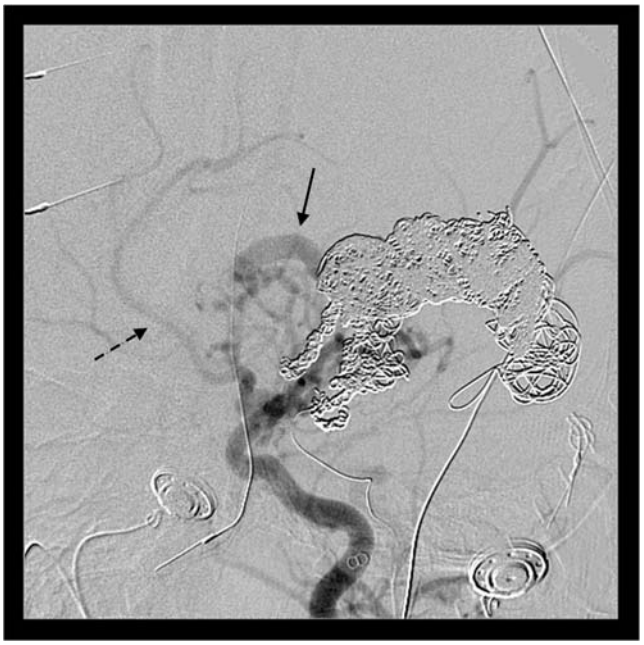


Fig. 4 Lateral view of left vertebral injection post-Onyx. Note stagnant contrast within fistulous vein (*black arrow*), Onyx within coil mass, lack of flow in the vein of Galen, and filling of anterior cerebral artery now that high flow to the posterior circulation is eliminated (*dashed black arrow*)

the acute setting. After a four-vessel diagnostic cerebral angiogram to completely visualize all feeding vessels, the patient underwent five staged coil embolization procedures performed with the aid of neurophysiological monitoring namely, electroencelphalogram and somatosensory-evoked potentials, and amytal testing, over the course of 5 consecutive days. Both transarterial and transvenous routes were utilized, and the main fistulous connections arising from the right and left posterior cerebral arteries and right and left anterior choroidal arteries were embolized using approximately 100 detachable platinum coils (Fig. 2). No intraoperative complications occurred. At the conclusion of these five procedures, the infant's baseline blood pressure increased by 15 torr, and all vasopressor agents were discontinued. Repeat echocardiograms revealed improved cardiac function. The patient was eventually extubated and discharged over the course of a few weeks with a stable neurological examination.

The patient returned 1 year later with signs of tachycardia. His weight had increased to 10 kg, he had met his usual milestones, and his neurological exam remained within normal limits. Although a repeat echocardiogram demonstrated normal cardiac function, the new onset tachycardia was believed to be a precursor of impending heart failure. A cerebral angiogram demonstrated residual fistula fed by the right and left posterior cerebral arteries and choroidal vessels (Fig. 3). Neurophysiological testing was performed, and the patient was treated again with simultaneous transarterial and transvenous catheterizations. After unremarkable injections with amytal, transarterial embolization of the right posterior cerebral artery was performed with 0.6 cm^3 of Onyx-18 over 2 min. Transvenous catheterization of the dilated venous structures was difficult because of the mass of coils present in the venous system from the prior embolizations. Once a microcatheter was positioned in the coil mass, catheter tip position was confirmed by releasing one detachable platinum coil in the existing coil mass using live subtraction techniques. Onyx-18 of 2.5 cm^3 was then injected over 5 min into the vein while periodic vertebral artery contrast injections were performed. Embolization was stopped once flow through the fistulous connections was eliminated (Fig. 4). The patient's oxygen saturation continued to remain 100%, thus ruling out the likelihood of Onyx pulmonary emboli. Follow-up MRI/MRV did not show any residual fistula (Fig. 5). The remainder of the hospitalization was unremarkable, and the infant's tachycardia improved. He was discharged in stable condition.

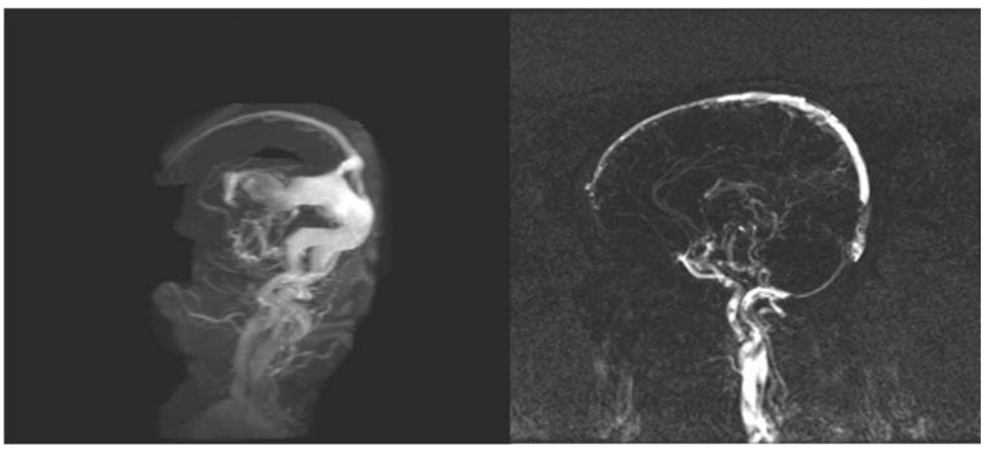


Fig. 5 Pretreatment and post-treatment sagittal MRV

Discussion

Given the cohesive but nonadhesive properties of Onyx and the ability to perform concurrent angiography, its controlled delivery allows for the precise administration of the liquid embolic agent. Although complications have been reported [9], there were no technically adverse events during this case. Studies are still underway to investigate the long-term efficacy of Onyx. However, in the acute setting for embolization in a symptomatic VGM, its use was well tolerated and effective for removing a high flow state, especially when used in conjunction with already existing coil scaffolding.

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