We present a rare case of an AVF of the basal vein of Rosenthal draining into a dilated vein of Galen that was managed by transarterial endovascular embolization. A male infant born at full term following a normal pregnancy and delivery with congestive heart failure, on investigation with MR imaging and MR angiography was found to have a basal vein of Rosenthal fistula with a dilated vein of Galen. His congestive heart failure was treated medically, and the AVF was managed electively at 10 months of age with successful transarterial endovascular embolization. The authors discuss the embryological aspects related to the pathological entity and the various clinical presentations, investigations, and management options. Management is primarily endovascular embolization; microsurgery is performed for a few selected cases, and radiosurgery has a limited role in older patients. Endovascular embolization is a safe and effective way to manage this malformation, with an excellent outcome if the AVF is eliminated by proper embolization at the fistulous point.

Key Words • basal vein fistula • vein of Galen • basal vein of Rosenthal fistula • vascular disorders

Abbreviations used in this paper: AVF = arteriovenous fistula; NBCA = N-butyl cyanoacrylate.

Case Report

History and Examination. This 10-month-old infant was born at full term following a normal pregnancy and delivery. In the neonatal period, he was found to have pulmonary hypertension that necessitated intubation for the first few days of life. Detailed workup with echocardiogram revealed fluid overload, with ventricular dilation. Clinically he had a pulmonary flow murmur with mild congestive cardiac failure. His condition was managed medically. His neuroimaging studies, consisting of MR imaging and MR angiography, demonstrated a dilated vein of Galen with normal brain parenchyma and no hydrocephalus. The head circumference was in the 95th percentile. The anterior fontanel was full but not tense. The child went on to achieve age-appropriate weight gain and developmental milestones.

Operation. By 10 months of age, it was decided to treat the child electively. Newly acquired MR images revealed an AVF between the lateral choroidal arteries of the posterior cerebral artery and the basal vein of Rosenthal at the level of the ambient cistern (Fig. 1). The enlarged basal vein traversed posteriorly and drained into the dilated vein of Galen. The straight sinus was hypoplastic, with a preserved median prosencephalic vein draining into the superior sagittal sinus (Figs. 1 and 2). A subsequent angiographic study demonstrated the AVF between the lateral choroidal arteries and the basal vein of Rosenthal that traversed posteriorly to drain into the dilated vein of Galen. The vein of Galen drained through the straight sinus, then through the falcine sinus, and then into the superior sagittal sinus. The child underwent transarterial endovascular embolization of the AVF at 10 months of age, in which NBCA mixed with ethiodol and
Tantalum was used (Fig. 3). Three fistulas were embolized; the first and second fistulas were embolized with 66% NBCA, and the third with 57% NBCA. The AVF was totally obliterated. The child did well, and the facial veins became less prominent after the procedure.

**Postoperative Course.** The follow-up angiograms (Fig. 4) obtained at 6 months and at 5 years postprocedure showed total obliteration of the AVF and remodeling of the vein of Galen and falcine sinus. The flow in the superior sagittal sinus was reversed. Follow-up MR imaging revealed total obliteration of the malformation, with remodeling of the feeding arteries and draining veins, with normal brain parenchyma.

**Discussion**

The development of the vasculature of the brain is a complex subject. The extraordinary work of Padget has advanced our understanding. The venous system of the brain is the earliest vascular system to develop, with large venous channels developing initially, followed by smaller, primitive venous channels developing in the 11- to 14-mm stage. Definitive arteries branching from the circle of Willis become recognizable later.

During the 20-mm stage, the pial arteries and veins are mere endothelial tubes crossing each other at various angles. The more acute the angle, the more maximal the surface area of crossing is, and the chance of developing an arteriovenous shunt is high. Embryologically, a fistula is a likely explanation of abnormally dilated capillary nets. Such fistulas are common transient stages during development, and they regress with the development of the proper capillary network and with vessel wall maturity. In our case, we hypothesize that the abnormal connection that developed at this stage of vascular development persisted.

The basal vein of Rosenthal is a complex venous channel, which develops secondarily from longitudinal anastomotic channels and that links several primary embryonic pial veins. The precursor veins are the telencephalic (deep middle cerebral vein), diencephalic (dorsal and ventral), and mesencephalic veins. These vessels are anatomically distant at development; they migrate and reorganize with the development of the large cerebral cortex to become anatomically closer together. Between the 20- and 80-mm stages, there is a constant change in the spatial orientation, connections, and drainage of the venous channels, with dynamic obliteration and development of new connections between them. At the 60- to 80-mm embryonic stage, the basal veins of Rosenthal become recognizable, draining into the vein of Galen. Earlier, each of its component veins had its own drainage. Commonly, the basal vein forms incompletely and drains through pial veins or alternate channels like the sphenoparietal sinus, cavernous sinus, and petrous sinus through the lateral mesencephalic or peduncular veins, and the transverse sigmoid sinus through the tentorial veins. At this stage, the internal cerebral vein is primitive, draining into the sinus of the embryonic tentorial sinus starts to regress.

Our hypothesis is that the fistula in this case arose from the persistence of the embryonic dilated capillary nets into one of the component veins that was later destined to form the basal vein. The early embryonic origin is supported by the persistence of the falcine sinus and hypoplasia of the straight sinus. This malformation is not a true vein of Galen malformation, which is an AVF between diencephalic vessels that are embryologically choroidal and the great cerebral vein. Usually in such a situation the internal cerebral veins are nonexistent, and the deep brain drains through an alternate route.

Neonates with an AVF malformation can present an-
Basal vein fistula with dilated vein of Galen

At present, MR imaging is the best initial modality for diagnosing these entities, with excellent sensitivity and specificity for accurate anatomical diagnosis. In this case it is important to differentiate the true vein of Galen from a vein of Galen aneurysmal dilation that also drains normal deep venous structures, and to avoid a venous approach that could result in catastrophic hemorrhage or venous infarction. Angiography remains the gold standard for investigation and is reserved for the time of treatment.

Management is primarily endovascular embolization; microsurgery is performed for a few selected cases, and radiosurgery has a limited role in older patients. The timing of the procedure is based on the clinical presentation. Clinically unstable or deteriorating cases need to be dealt with urgently, whereas a stable case like ours can wait until the child is mature enough for a major procedure.

The outcome is excellent if the AVF is eliminated by proper embolization at the fistulous point. Proximal occlusion of the feeding vessels will yield a poor outcome, because the active AVF will enhance angiogenesis from surrounding vessels, which are very hard to treat. We strongly discourage the use of any type of coil at the arterial site and believe that a fast tissue adhesive remains the best option.

Conclusions

This is a rare case of an AVF involving the basal vein of Rosenthal draining into a dilated vein of Galen and presenting as a vein of Galen malformation. Endovascular embolization is a safe and effective way to manage such a malformation.

Disclosure

The authors report no conflict of interest concerning the mate-
rials or methods used in this study or the findings specified in this paper.

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