

Transvenous Endovascular Embolisation of Nongalenic Arteriovenous Fistula

Islam Eldesouky*, Benjamin Gory, Roberto Riva, Nicolae Dorin and Francis Turjman

Department of Interventional Neuroradiology, Hopital Neurologique Pierre Wertheimer, France

***Corresponding Author:** Islam Fathallah Mohammed Kamel Eldesouky, Department of Interventional Neuroradiology, Hopital Neurologique Pierre Wertheimer, Hospices Civils de Lyon, Lyon, France.

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Abstract

Nongalenic arteriovenous fistula (NGAVF)- also known as intracranial pial arteriovenous fistula (AVF), cerebral AVF, or brain AVF is a rare vascular malformation. There are few retrospective case series describing their epidemiological, clinical and radiographic characteristics as well as technical elements of treatment. Combined transarterial and transvenous embolization of a 4 month old female with a multi-hole pial AVF is described. The patient underwent treatment by transarterial and transvenous embolisation in a staged fashion and without neurological complication.

Keywords: Arteriovenous fistula; Embolisation; Transvenous approach

Abbreviations: AVF: Arteriovenous Fistula; MRI: Magnetic Resonance Imaging, DSA: Digital Subtraction Angiography; CHF: Congestive Heart Failure; EVT: Endovascular Treatment; NGAVF: Nongalenic Arteriovenous Fistula; VOGM: Vein Of Galen Malformation; HHT: Hereditary Hemorrhagic Telangiectasia Syndrome; n-BCA: n-Butyl Cyanocrylate

Introduction

NGAVF- also known as intracranial pial AVF, cerebral AVF, or brain AVF is a rare vascular malformation. NGAVFs are distinct in that 1 or more pial arteries feeds directly into a cortical vein without the intervening nidus of a typical arteriovenous malformation (AVM) [1,2]. Venous drainage varies with the location of the fistula. An NGAVF is distinguished from a dural arteriovenous fistula (DAVF) by the location of the fistula site in the subpial meningeal space, and from a vein of Galen malformation (VOGM) by the lack of direct involvement of the embryonic median prosencephalic vein [1].

The incidence and prevalence of NGAVFs are unknown. In previously published series at University of California-San Francisco and the Mayo Clinic (Rochester, Minnesota), NGAVFs comprised 1.6% and 4.7% of AVMs [2]. NGAVFs usually become symptomatic in childhood. The most common presentation of NGAVF in neonates is congestive heart failure (CHF). Infants may present with increased head circumference or focal neurologic deficits. The goal of treatment for an NGAVF is occlusion of the AVF site, or of the feeding arteries and proximal draining vein as close to the fistula as possible. Transarterial embolisation is the primary method of treatment but open surgery and radiosurgery also have a role [3].

Case Report

History and clinical examination at presentation

A 20 weeks old female with no family history of genetic diseases, presented with increased head circumference. Head MRI raised suspicion of an AVM supplied by branches of anterior cerebral artery, with moderate tetraventricular hydrocephalus (Figure 1). Neurological

Neurological examination revealed no motor deficits. Echocardiography was done showing pulmonary arterial hypertension (PAH - 46/40 mm Hg). We decided after both endovascular and neurosurgical consultations to treat her by endovascular approach in a staged fashion.

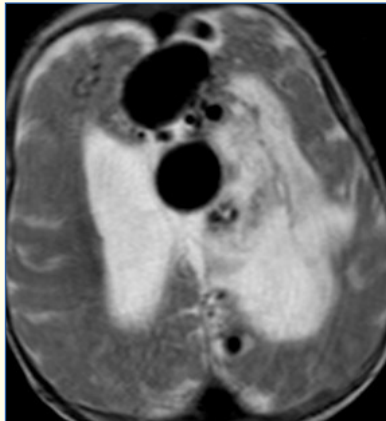


Figure 1: Axial T2 weighted brain MRI demonstrating moderate hydrocephalus, periventricular white matter changes, and multiple dilated veins with signal void.

Treatment and progress of the case

With the patient under general anesthesia and a sterile technique, left femoral artery was punctured using Microset. Ultraflow microcatheter and Mirage 008 microwire were used for catheterisation of right carotid and left vertebral arteries. (Figures 2 and 3). Navigation through right internal carotid artery was realized, then through anterior communicating artery to the left anterior cerebral artery where 3 feeders were embolized using n-BCA glue injection and two spiral platinum coils. This resulted in complete occlusion of those fistulous points with no procedural or post procedural complications.

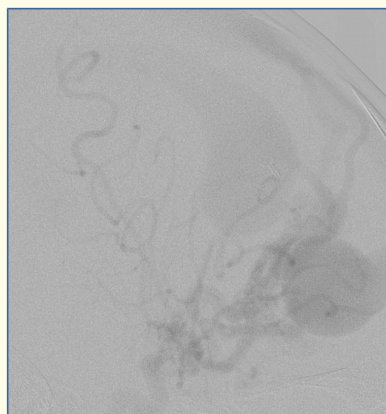


Figure 2: Left internal carotid AP arterial angiogram demonstrating multiple pial fistules supplied by branches of left anterior and middle cerebral arteries. The fistulous points drain into cortical veins that drain into dilated superior sagittal sinus. Notice the aneurysmal dilatation on the drainage veins.

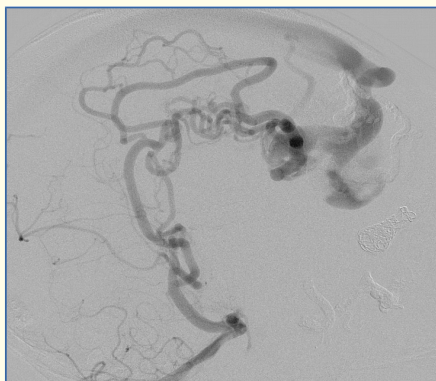


Figure 3: Left vertebral artery arterial angiogram demonstrating feeders from parietal branch of left posterior cerebral artery.

After six months a second session of EVT resulted in the occlusion of another 2 fistulous points with feeders from the anterior cerebral artery. A third session was realized 3 months later, using a 5F introducer and 5F vertebral to catheterize the left common carotid. Using an Ultraflow micro catheter and Mirage 008 micro wire, we catheterized a fistulous feeder from the frontal branch of left middle cerebral artery. 30% glue was injected in this feeder, with occlusion of the fistulous point. No procedural or post procedural complications were observed.

A fourth and last session was realized three months later. Distal catheterization of left posterior cerebral artery was attempted using Ultraflow microcatheter and Mirage microwire, but because of the tortuosity of the feeder the fistulous point could not be reached. Proximal injection of glue 50% resulted in pedicle occlusion, but persistence of the fistulous point (Figures 4 and 5). We decided to embolize this fistulous point by transvenous approach. A 5F introducer was put in the right femoral vein and Envoy 5F was used as a guide catheter. Ultraflow microcatheter and Hybrid microwire were used to catheterize left internal jugular vein, transverse sinus, superior sagittal sinus to the fistulous point and the drainage vein (Figure 6). 1 cc of onyx was used to occlude both the fistulous point and the proximal drainage vein, while preserving the sinus (Figure7). No procedural or post procedural complications were noticed.

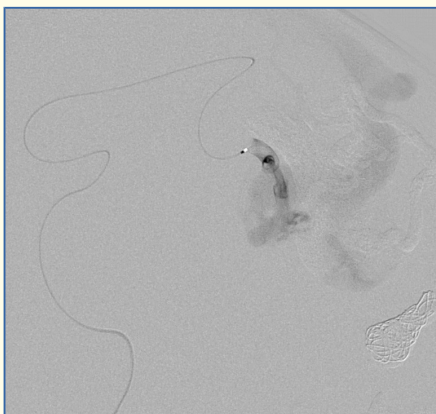


Figure 4: Supraselective angiogram demonstrating the Ultraflow microcatheter in the parietal branch of the posterior cerebral artery.



Figure 5: Left vertebral artery angiogram after injection of the glue demonstrating occlusion of the feeder with persistence of the fistula and its drainage vein.

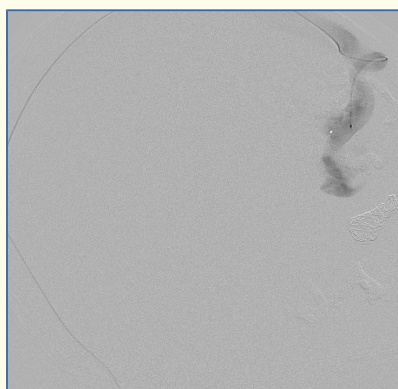


Figure 6: Venous phase of left vertebral artery angiogram demonstrating the Ultraflow microcatheter in the fistulous point after navigation through the superior sagittal sinus before injection of the onyx.

Discussion

Although NGAVFs are heterogeneous, the data suggests that these lesions can be separated into 2 distinct groups based on the patient's age at presentation and the complexity of AVF angioarchitecture, which are not independent of each other. Patients who present in the first 2 years of life are more likely to harbor large, complex multi-hole AVFs that have high flow arteriovenous shunting leading to CHF. Massive dural venous sinus dilation was also more common in these younger patients with marked arteriovenous shunting [4].

Children presenting after 2 years of age had more neurologic deficits, or intracranial hemorrhage. Other authors have noted that intracranial hemorrhage is an infrequent presentation for NGAVF [3].



Figure 7: Left vertebral artery angiogram demonstrating disappearance of the fistula and its draining vein with no onyx diffusion in the sinus.

Authors have reported NGAVF associated with hereditary hemorrhagic telangiectasia syndrome (HHT), Ehlers-Danlos syndrome, and neurofibromatosis type-I, Klippel-Trenaunay-Weber syndrome, encephalocraniocutaneous lipomatosis and aortic coarctation [5].

Neonatal NGAVFs are particularly difficult to treat. Due to small total blood volumes and coexistent CHF, surgery is unsafe in neonates. Stereotactic radiosurgery is reserved for patients with NGAVFs who also have an AVM nidus. Radiosurgery has limited ability to induce regression of larger caliber fistulas [6].

From an endovascular perspective, challenges include the presence of a high-flow fistula, tortuous intracranial feeding arteries, a small femoral artery access site, and limitations in the volume of contrast that can be used [7].

From a technical perspective, because of the accuracy of embolic deposition at the fistula site and the ability to deploy multiple coils without requiring recatheterization, the use of coils and glue in the dilated drainage vein is preferred as we did in the first session.

Transarterial embolisation is the first line of endovascular therapy, because it preserves patency of the physiological vein [8]. But in the setting where transarterial approach is limited due to tortuosity of the feeder, or fails to occlude the fistula as in our case, transvenous approach can be used. Transvenous approach has been described in the treatment of some cases of VOGMs and this method proved effective with associated morbidity not significantly different from that of the more common transarterial approach [8].

Conclusion

Transvenous approach can be used as complementary method in cases with difficult or failed transarterial approach for the treatment of pediatric NGAVF as in our case report with no neurological complications.

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