Successful Treatment of a Child with Ruptured Arteriovenous Malformation Using Onyx Embolization: A Case Report

Seidu A. Richard¹, Saurav S. Shrestha⁴, Changwei Zhang¹,⁴, Wei Fu¹, Ting Wang¹,⁴, Wu Cong¹,⁴, Xiaodong Xie¹,⁴*

¹Department of Neurosurgery, West China Hospital, Sichuan University, Chengdu, China
²Department of Immunology, Jiangsu University, Zhenjiang, China
³Department of Surgery, Volta Regional Hospital, Ghana, West Africa
⁴Department of Neuroradiology Intervention, West China Hospital, Sichuan University, Chengdu, China

Email: *gbepoo@gmail.com

Abstract

Introduction: Arteriovenous Malformations (AVMs) arising deep in the basal ganglia and thalamus encompass a very small fraction of total cerebral AVMs and are rarely encountered in neurosurgery. The rate of rupture of these AVMs is very uncommon making our case more thought-provoking.

Case presentation: We present a case of 8-year-old boy with 3 days' history of an intermittent headache with nausea and vomiting. Computer tomographic angiography (CTA) confirmed right basal ganglia and dorsal thalamic hematoma with numerous tortuous blood vessels around the hematoma.

Conclusion: We successfully treated this child with ruptured deep seated cerebral AVMs using Onyx embolization with no further neurological deficits.

Keywords

AVM, Onyx Embolization, Basal Ganglia, Thalamus, Hematoma

1. Introduction

Arteriovenous Malformations (AVMs) refers to anomalous proliferation or collection of dilated arteries and veins within the brain parenchyma with an anomalous superseding capillary bed, instigating shunting [1] [2] [3]. The occurrences of cerebral AVMs are uncommon in pediatric age group, although they are believed to be congenital [3] [4]. In children, AVMs can manifest as spontaneous hemorrhage resulting in hydrocephalus, non-specific headaches, epilepsy.
or focal neurological deficit [5]. Management of cerebral AVMs usually encompasses multiplicities of treatment modalities such as embolization, surgical removal radio surgery and conservative. Efficient treatment usually focuses on lessening the possibilities of bleeding/rebleeding [6] [7]. Therefore, complete or proximately complete obliteration of the feeding vessels as well as vascular nidus is required. Coil embolization of the AVM followed by surgical resection or radio surgery was the ultimate treatment modalities although complete obliteration of the nidus via embolization was not possible in most cases [3] [6] [7] [8] [9] [10]. Other embolic agents like N-butyl cyanoacrylate (n-BCA) has been used before but with a success rate of 10% [6] [8] [11].

Currently, a liquid embolic agent called Onyx (eV3, Irvine, California) is the most preferred agent. It is made up of ethylene-vinyl alcohol dissolved in dimethyl sulfoxide (DMSO). It is able to penetrate deeper into the nidus because of its nonadhesive nature as well as slow solidification rate with longer polymerization time. These features allow for a more lengthy and meticulous injection [10] [12] [13] [14] [15] [16]. We present a case of rupture AVM in a child which we successfully managed with Onyx embolization.

2. Case Report

An 8-year-old boy presented with 3 days’ history of an intermittent headache with nausea and vomiting. He was apparently well until the above complaints started prior to presentation at a local hospital. His parents deny any history of loss of consciousness, seizures, incontinence, chest pain or any respiratory distress. His headaches were relieved after vomiting and aggravates mostly in the mornings. He was put on analgesia with no improvement of his condition. His immunization is completed according to his age. Past medical history was unremarkable. Computed Tomography (CT-Scan) done at the local hospital suggest hypothalamic and dorsal thalamic hemorrhage with cerebral AVMs (Figure 1(a) and Figure 1(b)). The patient was referred to our center for further evaluation and treatment. On examination, we saw a young boy who was not acutely ill. Systemic examination revealed no abnormalities. Neurological examination revealed normal cranial nerves. The muscle bulk on the limbs as well power was normal. Routine laboratory and auxiliary investigation (CXR, ECG etc.) were all normal.

Computer tomographic angiography (CTA) confirmed right basal ganglia and dorsal thalamic hematoma with numerous tortuous blood vessels around the hematoma (Figures 2(a)-(d)). The blood vessels supplying the right side of the brain especially internal veins and large cerebral veins are thick, considered as AVMs while the right side of anterior cerebral artery (A1) segment and left posterior cerebral artery (P1) segment are slightly thinner. Digital subtracting angiography (DSA) also indicated right basal ganglia and dorsal thalamus cerebral hemorrhage with thick and numerous tortuous blood vessels measuring about $2.7 \times 1.4 \times 1.5$ cm in diameter with hematoma in right ventricle (Figure 3(a) and Figure 3(b)). We made a diagnosis of right basal ganglia and dorsal
Figure 1. (a) & (b) are preoperative CT-Scan images indicating hematoma while (c) & (d) are postoperative CT-Scan images showing the resolution of the hematoma after Onyx embolization.

Figure 2. (a)-(d) are computer tomographic angiography (CTA) confirmed right basal ganglia and dorsal thalamus hematoma with numerous tortuous blood vessels around the hematoma.
thalamic AVM. His parents were taken through a series of education and counseling and endovascular embolization treatment scheduled for the next day.

Patient was put into a supine position. Under general anesthesia, 6th F sheath was placed into the right femoral artery. Cerebral angiography done intraoperatively revealed that the right posterior choroidal artery and posterior arterial branch of the right anterior cerebral artery were draining straight into the sinus. With a continuous pressurized drip of normal saline, the catheter and micro-guide wire was guided into the right internal carotid artery and advanced into the right choroidal artery to reach the AVM and fistula. The micro-guide wire was removed and Onyx embolization agent gradually injected into the AVM. Intraoperative angiography done immediately after the procedure revealed successful obliteration of the AVM (Figure 3(c) and Figure 3(d)). Postoperative CT-Scan done two [2] days after the procedure revealed near total reduction in the hematoma size (Figure 1(c) and Figure 1(d)). The patient recovered very quickly and was discharged home 2-days after the treatment. Outpa-

Figure 3. (a) & (b) are preoperative DSA images show the AVM while (c) & (d) are postoperative DSA images showing total occlusion of the AVM with onyx embolization agent.
tient department visits were scheduled every 3-month. Two years’ follow-up revealed no recurrence of his disease.

3. Discussion

AVMs arising deep in the basal ganglia and thalamus encompass a very small fraction of total cerebral AVMs. They are rarely seen in typical clinical setting. Studies have established that AVMs in the basal ganglia and thalamus forms about 4.3% - 11% of all AVM cases [7] [17] [18]. Fleetwood et al observed an incident rate of about 54.2% [7]. On the other hand, pediatric AVMs constitutes about 3% of all AVMs which mean that in the pediatric age group, AVM are not as frequent compare to their adult counterparts [3] [19] [20] [21]. Therefore, our case is very unusual. The etiology of AVMs is still a matter of debate although their angioarchitectural physiognomies and occurrence in all age groups suggest that AVMs possibly have embryonic origin. Notwithstanding this, a sizable number of AVMs don’t appear to have hereditary source, though sporadic incidents of familial manifestation have been seen in literature [2] [22]. Occasionally, spontaneously disappearances of AVMs have been reported in literature as well as recurrence after total angiographic obliteration [2] [23] [24].

Studies have indicated that AVMs trigger about 2% of all strokes and 4% of hemorrhagic types [2] [25]. Our patient presents with hemorrhagic stroke as result of the reputed AVM. The younger age group is more prone to stroke as compare to their adult counterpart’s. AVMs causes one-third of hemorrhagic strokes in children or young adults [2]. Our case is also falls within the pediatric age group. Hemorrhage arising from AVM is mostly less life threatening as compare to the rupture of an intracranial aneurysm or spontaneous hypertensive intracranial hemorrhage, although AVM rupture with accompanying intraparenchymatous hemorrhage frequently leads to substantial neurologic disability. The mortality rate and risk of lifelong morbidity as a result of ruptured AVM differ extensively in various studies, ranges from 5% - 25% and 10% - 40%, correspondingly [2] [26]. Numerous anatomic factors have been implicated in hemorrhagic manifestation of AVMs. Neutral risk factors for hemorrhagic include small size, deep venous drainage, indefinite or nonborder zone (watershed) as well as infratentorial locations, accompanying aneurysms, hypertension, small number of draining veins, venous ectasias, and high feeding artery pressure [2] [27] [28]. Symptomatic epilepsy, which is seen in about 18% - 35% of cases is the second most common manifestation of AVMs [2] [26] [28]. Our case did not presented with epilepsy. Anatomic AVM features linked to epileptic manifestations includes large size, cortical location of the nidus or the feeders, and site of the AVM with reference to the middle cerebral artery [2] [29] [30].

The radiological characteristics of a nidus variety of cerebral AVM are consistent with its definition [31]. The diagnostic principles of AVM comprise 1) the existence of a nidus embedded within the brain parenchyma, recognized by both cross-sectional imaging like CT and MRI or conservative angiography; and 2)
early venous drainage, which is preeminently observed on dynamic studies with typical pointers being conservative catheter angiography. The precise elucidation of early venous drainage can be determined only if the veins are observed in the “arterial” phase, which is usually recognized on typical magnetic resonance angiography (MRA) or CTA if the shunt volume and draining veins are sizable enough [31]. Furthermore, radiological techniques like dynamic MRA and CTA are gradually being used in the recognition of early drainage for tinier AVMs and thus in determining the diagnosis. CTA was very useful in ruling out other hemorrhage sources as the case of bleeding in our patient. Nevertheless, the above-mentioned principles are imperative in distinguishing brain AVMs from other vascular diseases of the brain [31] [32] [33] [34].

Moreover, two subtypes of anomalous webs of vessels can be seen if a nidus exists. The characteristic form is the glomerular or compact kind nidus, which comprises of anomalous vessels without any interposed normal brain tissue. The second kind which is infrequently observed is the supposed diffuse or proliferative kind nidus, in which normal brain parenchyma is interposed throughout the web of vessels [31]. If the above exist, proliferative angiopathy or cerebrofacial arteriovenous metameric syndrome (CAMS) should be key differential diagnosis. These differentials can be differentiated from true brain AVMs on the basis of the lack of early venous drainage which usually occurs in proliferative angiopathy as well as the typical site and interrelation with facial AVMs observed in CAMS [31]. The arterial feeder vessels and venous drainage of a brain AVM will rely on the site of the nidus. It is now very clear that deep and ventricular AVMs usually have feeders arising from the perforator such as lenticulostriate, thalamoperforator branches and choroidal branches such as anterior, medial, and lateral posterior arteries correspondingly, while venous drainage will classically be via the deep venous plexus [31]. On the other hand, AVMs located superficial or cortically have their main arterial supplies from the pial arteries [31].

Besides conservative treatment, there are three other treatment modalities for brain AVMs. These other modalities include; endovascular embolization, surgical removal or radiosurgery. All this option has their own advantages and disadvantages thus, a multimodality treatment is most generally advisable [4] [31] [35]. Furthermore, there are certain radiological and clinical qualities that are essential in deciding the securest and most efficient management option. Although surgical removal offers a fast remedy in appropriate circumstances; the complication rate relies mainly on the Spetzler-Martin grade: The higher or lower the grade, the higher or lower the complication rate respectively. Thus, surgical resection may be advantageous for cases with smaller and cortical-based brain AVMs [31] [36] [37].

Endovascular embolization is a radiological treatment modality that can be used to rapidly eradicate angiographic risk factors. Although there are no tangible contraindications for endovascular treatment, the outcome of treatment rate with embolization alone is about 10% - 20% which is comparatively very low.
except in high flow pialarteriovenous fistulas or small lesions [31] [38]. Furthermore, AVM embolization is not just limited to incomplete nidus embolization, although therapeutic embolization as an alternative to radio surgery or surgical resection is achievable in particular cases. The fundamental facts on therapeutic AVM embolization have distinctly been reported in literature. These facts are: 1) the AVM must be small- or medium-sized measuring about 1 - 3 cm, not situated in brain stem or deep structures, with feeders from a single vascular zone, reachable by microcatheters and permitting reflux of onyx for 2 - 3 cm, 2) the nidus must be well demarcated, and 3) the location of the proximal segments of the draining vein must be clear to identify venous filling with onyx. Emphatically, all these anatomical characteristics also make n-BCA appropriate as an embolization agent too [12] [39] [40].

On the other hand, catheter entrapment and angiotoxicity are the particular drawbacks to the use of Onyx embolization in brain AVMs [6]. Furthermore, the reflux of Onyx during the procedure acts as a double-edged sword. Nevertheless, accurate reflux aids in the incessant penetration of Onyx into the nidus to attain acceptable embolization compared with n-BCA. Conventionally, inappropriate reflux usually leads to hitches in removing the microcatheter, which may lead to hemorrhagic complications or catheter entrapment [6]. Xu et al. are of the view that the tortuosity of the feeding artery, the length of the Onyx reflux and the injection time are the key triggers of catheter entrapment. They explain further that Onyx is not a good embolic agent for apparently small tortuous feeding arteries [6]. If Onyx has to be used in cases of small tortuous feeding arteries, the use of microcatheter with detachable tips may aid in decreasing peculiar risks accompanying nondetachable microcatheters [6] [41]. Onyx embolization has also been linked to angiotoxicity with vasospasm or angionecrosis because of its action potential-reducing properties in DMSO [6] [13]. Another most efficient treatment option for deep seated brain AVMs is radio surgery although it may not be appropriate for the treatment of AVMs with angiographic risk factors because of imminent hemorrhage [31]. Furthermore, AVMs with a volume greater than 12 mL are linked to lower treatment rates as well as higher complication rates. Nevertheless, due to the radiation consequences, it is advisable to use radio surgery treatment options with caution in children [4] [31] [42]. Conservative treatment is normally used when the risk posed by the three [3] treatment modalities above are too high, like in large and deep seated AVMs or in asymptomatic cases in whom imminent hemorrhage is very unlikely [31]. The prognosis of pediatric AVM is usually good if well managed however, re-rupture frequency is projected to be 2 - 4% with a death rate up to 25% per every incidence of re-rupture; this risk is greater within 5 years after identification of the AVM [3] [19].

4. Conclusion

We successfully treated this child with ruptured deep seated cerebral AVMs us-
ing Onyx embolization with no further neurological deficits. The first line treatment for our pediatric patient was certainly embolization therapy because of the location of the AVM. Furthermore, surgical removal will be very difficult and radiotherapy did not seem appropriate because of the risks factors associated with radiation in this age group. Two years’ follow-up also revealed no recurrence of his disease meaning our treatment has been very effective so far.

Consent for Publication
The patient and his relatives were dually informed about our intention to publish his case and he/they fully concerted to the use of his documents. The hospital also concerted to the use of this information for publication.

Authors’ Contributions
SAR conceived the project and designed the study. FW, SSS, WT collected patient’s data. CWZ, CW and XX provided technical assistance in the study. SAR analyzed the data, prepared the illustrations and wrote the paper. SSS, FW and WT contributed equally to the manuscript. All authors approved the paper for the submission.

Competing Interests
All the authors have no competing interest to disclose.

References


