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CASE REPORT

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Ruptured Brainstem Arteriovenous Malformation Successfully Treated with Endovascular Surgery Used PHIL

Dante Valer[1]*, John Vargas[2], Osmar Ordinola[1], Giancarlo Saal[1] and Rodolfo Rodriguez[1]

[1]Médico Asistente del Servicio de Neurocirugía Endovascular, Departamento de Neurocirugía, Hospital Nacional Guillermo Almenara Irigoyen, Lima

[2]Médico Residente de Neurocirugía, Departamento de Neurocirugía, Hospital Nacional Guillermo Almenara Irigoyen, Lima

ABSTRACT

Brainstem arteriovenous malformations (AVMs) are rare and complex, presenting mainly with hemorrhage, which can be catastrophic. The objective of its treatment is the angiographic obliteration of the lesion without worsening or with improvement of the neurological symptoms. Embolization is an efficient treatment for these AVMs but there are very few reported cases, although total obliteration is difficult, but it can also serve to reduce the nest and close the most dangerous factors of the AVM. Brainstem AVM is a rare pathology with high associated morbidity and mortality, with a high rate of rebleeding without treatment, which in selected cases embolization can be curative with a good neurological prognosis.

Keywords: Brainstem arteriovenous malformation, Phil, Embolization, Angiographic obliteration.

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Introduction

Brainstem AVMs are rare and complex, constituting between 2-6% of all cerebral AVM and about ¼ of the posterior cranial fossa AVMs [1,2,3]. The initial presentation with hemorrhage has been reported in 76-100% of patients. It is postulated that it has a higher association with intranidal aneurysms which gives it its unfavorable behavior. The consequence of this bleeding can be catastrophic, with a mortality of 33% in treated patients and 66% in untreated patients [1, 4]. The annual risk rate for bleeding is 15 to 17.5% [1, 4].

The goal of treatment is angiographic obliteration of the lesion with improvement of the neurological symptoms [1]. The treatment of this type of AVM is not yet determined. In the last decade, embolization with ethylene vinyl alcohol copolymers, such as Onyx ®, is an efficient treatment of AVMs in other parts of the brain, but there are very few cases in the brainstem. Radiosurgery has been used, but it can cause neurological impairment and has a low obliteration rate relative to other locations in the brain [2].

Precipitating hydrophobic injectable liquid (PHIL; Micro-Vention, Tustin, California) is a new embolizing substance composed of a nonadhesive copolymer (polylactide-co-glycolide and polyhydroxy ethylmeth-acrylate) dissolved in DMSO with an iodine component (triiodo- phenol) covalently bound to the copolymer, causing radiopacity. PHIL is ready to use in prefilled syringes of 1 mL and does not require shaking before injection.

Total obliteration with embolization is difficult because they can have multiple afferents and extend to eloquent structures. Therefore, the therapeutic objective of embolization in some cases is to reduce the nest and try to close the most dangerous factors of AVM, such as aneurysms [2].

Yen et al in their series of cases on stereotaxic radiosurgery mention that the new embolic materials promise to reduce the size of the effective nest and obliterate the AVM, although the risks associated with surgery are high [5]. For this reason, we present the case of a patient with a total embolized ruptured pontine AVM in a single session, with a slow favorable neurological evolution, with mRS(Rankin) at 3 months of 1(Figure 4), with last study of MRI and brain angiography at 9 months of follow-up that shows absence of MAV (Figure 5).

Clinical Case

13-year-old female patient, with no significant medical history, 3 days before admission she suddenly presented oppressive global headache associated with explosive vomiting on multiple

Contact Dante Valer Gonzales neuro_valer@hotmail.com HEndovascular Neurosurgeon, Hospital Guillermo Almenara Irigoyen – EsSalud. Av. Grau 800 – La Victoria, Telf. 3242983 Anexos: 44157. Lima, Perú. .

occasions, for this reason, she is taken to her local hospital where a tomography (CT) of the brain showing a thick perimesencephalic subarachnoid hemorrhage (SAH) associated with a left pontine intracerebral hematoma (ICH) (Figure 01), she is referred to our hospital. She was admitted with a 15 point Glasgow scale (GA), without motor deficit, without sensory deficit, isochoric and photoreactive pupils, with preserved cranial nerves, with + / +++ neck stiffness. AngioTEM showing a ruptured left pontine arteriovenous malformation (AVM) (Figure 1B), cerebral panangiography showing a ruptutred left pontine AVM , moderate flow, moderate resistance, measurement 14.27 mm x 13.24 mm in anteroposterior (AP) and 7.64 mm x 15.67 mm laterally, with a single afference from a branch greater than 1mm originating from the anterior pontine segment of the left anteroinferior cerebellar artery (AICA), with the presence of an intranidal aneurysm, with efference towards the bilateral superior petrosal sinus and anterior mesencephalic vein (Figure 2). It was decided to embolize, for which an Envoy ® 6F guide catheter assisted with hydrophilic guidance was navigated to the end of the right V2 segment. Then it is navigated with the Sonic ® 1.5F microcatheter assisted with Hybrid ® 007 microguide, entering the left AICA and then reaching the nest of the AVM itself, where it is embolized with 0.3cc of Phil 25% ®. Angiographic control is performed where the total absence of the AVM is evidenced (Figure 3), with patency of neighboring vascular structures. Tomographic control is performed where there is no evidence of hydrocephalus or rebleeding, with embolization material in the left protrusion, with perilesional edema (Figure 4).

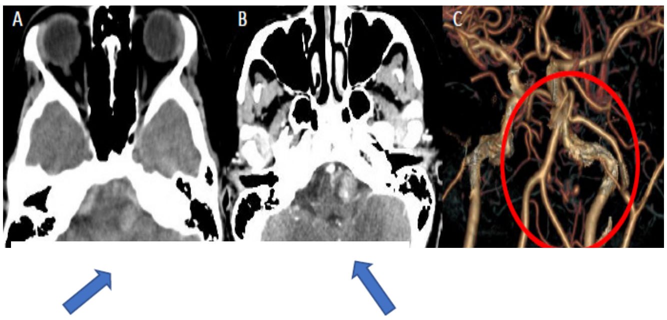

Figure 1. (A) Non-contrast cerebral CT in axial section showing hyperdense image in the lower left pons, compatible with a hematoma (arrow). (B) Brain CT with contrast in axial section showing a hyperdense contrast capturing lesion in the lower left pontine region (arrow). (C) AngioTEM showing vascular curl

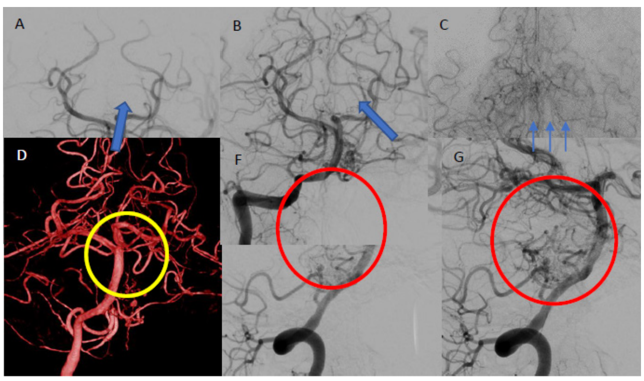

Figure 2. Angiography of the right vertebral artery. (A,D) AP incidence, in the arterial phase, where a left inferior pontine AVM is evident (thick arrow) arising from a small afferent greater than 1mm from the anterior pontine segment of the left anteroinferior cerebellar artery (thin arrow). B. AP incidence, in early venous phase, where venous drainage towards the left superior petrous sinus is evident (arrow) and anterior mesencephalic vein. (E,F) Lateral incidence, in arterial phase, where lower left pontine AVM is evident (circle).

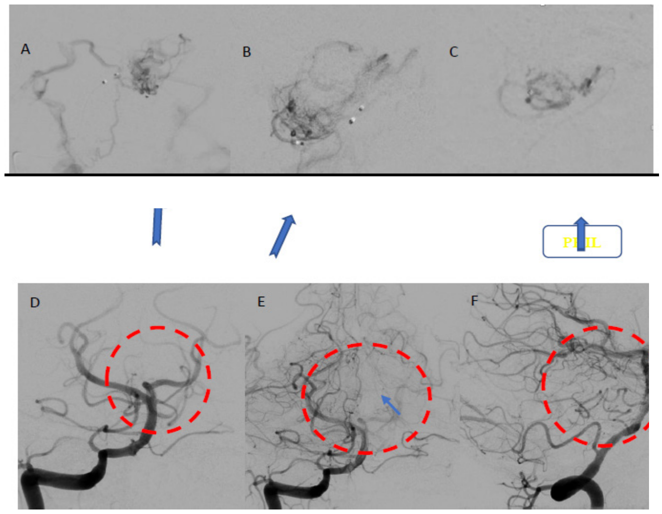

Figure 3. (A,B). Ultraselective angiography shows AVM in the pons with the presence of an intranidal aneurysm. (C) shows the embolizing substance Phil penetrating the AVM. (D,F) Angiography post embolization of the right vertebral artery in AP incidence where the absence of the AVM is evidenced (dashed circle) with patency of the left AICA (thin arrow). (G) Angiography of the right AV in lateral incidence showing absence of AVM (dashed circle).

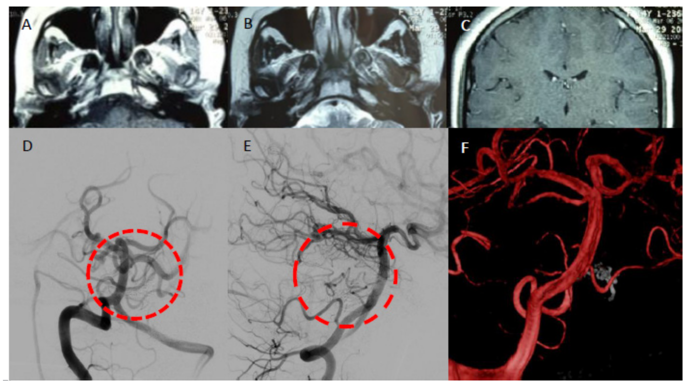

Figure 4. (A,B,C) Brain tomography shows Phill embolizing substance in the brainstem in the area of the arteriovenous malformation, not infarcts or bleeding. (D, E) The new embolizing substance Phil with the DMSO that differs from onyx which is dark, this is yellow color. (F) Shows the clinical evolution of the patient who at 02 months still presents still presents left sixth nerve palsy and right hemiparesis, managing to stand up. (G) At 03 months the patient is able to walk without any deficit in the limbs.

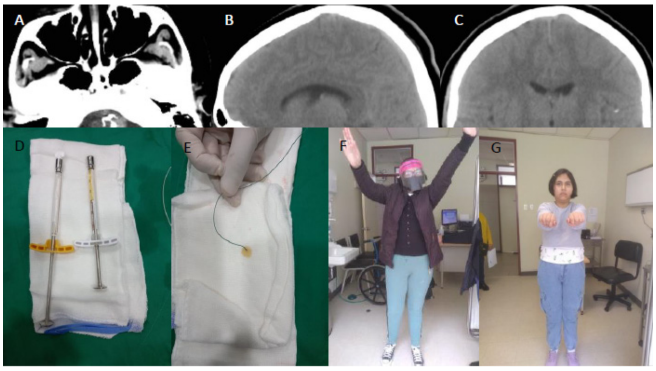

Figure 5. MRI and brain angiography at 9 months of follow-up that shows absence of MAV.

Postoperatively, the patient presented severe right motor deficit Daniels 1/5, associated with moderate left sixth and seventh cranial nerve (PC) paresis, in addition to persisting with mechanical ventilation. She tried three times to remove the mechanical ventilator, but it was unsuccessful. Intensive physical therapy is started from the second postoperative day. On day 15, she underwent a tracheostomy without intercurrences, after which the patient was able to get off mechanical ventilation. On postoperative day 30, 3/5 Daniels muscle strength was evidenced in the right hemibody, associated with mild left VI and VII CP paresis. On postoperative day 35, he was decannulated without complications and the patient had 4- / 5 Daniels muscular strength, tolerating the sitting position. On postoperative day 60, the patient began to walk with support and presented adequate dialogue. In the control at three months, the patient wanders without support, Daniels muscular strength 4 + / 5, adequate dialogue, left VI and VII CP paresis almost imperceptible.

Discussion

The largest series of brainstem AVMs are those of Han et al (29 patients), Kelly et al (29 patients), Nozaki et al (20 patients) report 56% total resection with 25% morbidity, Drake et al (15 patients), Yasargil (14 patients), Solomon and Stein (12 patients) who reported 75% total resection and 22% morbidity, Spetzler and Martin (10 patients), Lawton (8 patients) reported total resection in 100% and 0% morbidity [1,2,4], but in 2017 Madhugiri presented a series of 39 patients with brainstem AVM, of which 92.3% presented with hemorrhage, and it was determined that those who presented with Intraventricular hemorrhage (IVH) had better functional status than those who presented with SAH or ICH, but with a higher risk of needing a ventricular drain [1].

Patients with ICH damage the nuclei and tracts of the brainstem, which explains the poor neurological status [1, 4, 6]. SAH can generate spasm of the perforating arteries of the brainstem, generating focal ischemia and therefore develop deficits in its presentation [1]. Our case presented with ICH and SAH, but did not present preoperative neurological deficit.

In the ARUBA study it was identified that the average age of the patients with bulbar AVM was 60.9 years +/- 27.3 years, for the pontine AVM it was 41.85 years +/- 16.9 years, while for the midbrain AVM it was 33.3 years +/- 15.3 years, with statistical difference. But patients with pontine or bulbar ICH, being older, are considered hypertensive etiology and no further studies are done. Therefore, MRI angiography should be performed 6 to 8 weeks after the stroke. There, flow gaps may be evidenced, and if it is suggestive of AVM, you should go to digital subtraction angiography (1). For this reason, in our case, given the suspicion of cerebral angioTEM, it was decided to go for angiography, finding what had already been described.

92% of AVMs of the brainstem are unilateral and are divided into purely piales (71.8%) or with a parenchymal component. Pial lesions are considered to be accessible to microsurgical treatment, while parenchymal lesions are not [1].

Lawton classifies AVMs of the brainstem into 6 types, based on their location in the midbrain, pons, or medulla oblongata, as well as their anterior, posterior, and lateral orientation, which is helpful in selecting the patient for surgery and planning the surgical strategy and standardizing the expected results. The resection rate is 100% in lateral pontine or bulbar lesions, with a recovery rate or no change of 100% for the pontine and 75% for the bulbar. The rate of worsening or death is 50% in anterior pontine or posterior midbrain AVMs [2].

The anterior pontine AVM is located on a rectangular surface between the basilar groove medially and the root of the V PC laterally, and between the pontomesencephalic groove superiorly and the pontobulbar groove inferiorly. Despite being anterior, it is usually unilateral. It is located between the S1 segment of the ASCA superiorly and the A1 segment of the AICA inferiorly, and can be nourished by both arteries. The long perforating branches of the basilar trunk can also nourish this AVM. Venous drainage is collected medially in the median anterior mesencephalic vein or more frequently in the superior petrous vein and the superior petrosal sinus laterally [4]. This type being the pontine AVM found in our patient.

The lateral pontine AVM is situated on a triangular surface between the root of the V PC medially and the cerebellum fissure laterally. Its pial invasion varies, and may be only on the pial surface or penetrate deep to the middle cerebellar peduncle. These AVMs are nurtured by the a2 and a3 segments of the AICA, and more rarely by branches of the ASCA. Venous drainage goes to the superior petrous vein and the superior petrosal sinus [4].

A technique in which the afferent is coagulated at the surgical site without touching the parenchymal component has been described, but its effectiveness rate has not been determined [1]. For example, Han et al applied the occlusion in situ technique, where all the afferents were coagulated first, devascularizing the AVM, then the draining vein was closed, but the AVM was not removed. He showed that the rate of good results (no change or neurological improvement) was 83% in those resected and 55% in those occluded in situ, with a complete obliteration rate in 100% in those resected and 73% in those occluded in situ. situ [2].

The type of treatment of these AVMs is not determined, Liu et al indicate that embolization is a curative technique for brainstem

AVMs. Onyx ® being an embolizing substance that has better control during injection, injection time, with higher occlusion rates and lower complication rates. Jin et al in their series of 13 embolized patients, the most frequent types are anterior midbrain, with good results during embolization [2].

Due to its critical location with respect to the lower cranial nerves, which is why reflux is not tolerated, which is why pontine and bulbar AVMs have severe postoperative complications. Jin et al in their series found that, of the 13 patients, 6 had severe postoperative complications and of them, 5 had Onyx reflux. For this reason, slow and controlled injection with a high probability of penetrating through the arterioles without reflux is suggested. The cytotoxic effect of Onyx ® associated with injection of dimethyl sulfoxide (DMSO) must also be taken into consideration, and brainstem AVM has been shown to resemble perimedular AVM in the spinal cord, making them more sensitive to DMSO damage. Likewise, the development of edema and inflammation around the nest as a result of the injection of Onyx ® [2].

In our case, no reflux was observed, with patency of all the adjacent vascular structures, but in the postoperative period, he presented severe neurological deficit, which gradually recovered over the weeks, for which we can hypothesize that it is due to the effect cytotoxic of Phil ® and DMSO.

Embolization helps prevent repetitive bleeding, particularly for those in the acute state, but is still curative and has a potential risk of ischemic complications, because it can embolize only specific parts, such as intranidal aneurysms, or decrease the flow of the AVM [6].

Ischemia of the dorsal and lateral territory of the pons may occur because there is territory irrigated by the afferents but it is not evident in the angiography, therefore it can present with severe neurological deficit, but it is associated with better results than other locations [4].

Endo et al report 5 cases of AVM of the cerebellopontine angle (CPA) and also review the cases described in the literature, with a total of 31 cases, which were embolized, being predominantly male (71%), with a mean age of 57 years, of which 58% presented with trigeminal neuralgia and 42% with hemorrhage. 60% had a prenidal or nesting aneurysm as the cause of the hemorrhage. The main afference was through the superolateral pontine artery, in 89.66%, which originated from the basilar artery. N-Butylcyanoacrylate or Onyx 18 was used as embolic material, with extreme care at reflux [6].

Another treatment alternative is stereotaxic radiosurgery (ROSC) and its objective is the complete obliteration of the nest avoiding complications associated with radiation, but the bibliography mentions that this obliteration rate is between 44-73% after 3 years [3].

Kano et al found in their series of 67 patients that ROSC had a complete occlusion rate of 70% at 5 years and a mortality of 4.5%. He concludes that the best candidates for this therapy are those with small volume AVMs who receive at least 20 Gy in their margins [3]. Stereotaxic radiosurgery should be considered if the nest is subpial or epipial, and not when it is deep in the parenchyma [5].

Conclusion

Brainstem AVMs are a rare pathology with high associated morbidity and mortality, with a high rate of rebleeding if not treated, which increases morbidity and mortality even more, with an unclear therapeutic algorithm, but in selected cases embolization can be curative with good neurological prognosis [7-20].

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