Medusa Head: A Case Report of an Extracranial Arteriovenous Malformation with Multiple Aneurysms

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Background **---** Unlike those seen in the intracranial circulation, craniofacial high-flow malformations are rare, and no consensus has been reached with regards to its treatment. Embolization has become an integral part of the treatment of these malformations, either alone or in combination with surgical resection.

Case **---** We present a case of a 37 year old female with a huge facial mass and epistaxis. She was diagnosed with hemangioma and advised excision of the mass. She had an episode of epistaxis and bleeding from her left eye prior to admission. A four vessel angiography showed markedly tortuous bilateral external carotid arteries, arteriovenous malformation with multiple aneurysm formation in the left orbital and periorbital areas. A superselective embolization of the periorbital arteriovenous malformation was done. A week after the embolization, she then underwent wide excision of the Arteriovenous malformation with reconstruction.

Conclusion **---** For craniofacial arteriovenous malformation, superselective embolization followed by surgical resection may be considered in the management of the lesion. ***Phil Heart Center J 2012; 16(2):84-89***

Key Words: Arteriovenous Malformation ■ Superselective embolization

In the mid 1800s, Luschka and Virchow originally described arteriovenous malformations (AVMs) as abnormal connections between arteries and veins. They are seen as tangled masses of blood vessels with no intervening capillaries. The inherent lack of capillaries permits the flow of blood from a high pressure arterial supply to the low pressure venous system. Because of this, the veins enlarge as they try to accommodate this flow, causing some of them to stretch out, dilate aneurysmally and become susceptible to rupture and bleeding.[1] The surrounding normal tissues may be damaged as the arteriovenous malformations "steal" blood from those areas.

Majority of arteriovenous malformations, like most vascular malformations, are present at birth; however, some manifest later in childhood. These lesions tend to grow as the child grows; although, other factors, such as trauma, infection, hormonal changes or embolic or surgical intervention can cause these lesions to expand.[2]

Vascular malformations are further subdivided into high-flow lesions (arteriovenous malformations, arteriovenous fistulas), low-flow lesions (capillary malformation, venous malformations, LM), or combined vascular malformations. These lesions are 20-fold more common to occur in the intracranial as compared to extracranial vasculature.[3]

The estimated incidence rate of intracranial AVMs in the United States is pegged at 0.04-0.52%, which is similar to the average worldwide incidence.[4] In a review of 800 cases of AVMs, Krauenbuhl and Yasargil determined that extracranial AVMs accounts for only 8.1% of all cases

Finalist, Poster Presentation 18th PHC Annual Research Paper Competition held on February 23, 2010 at Philippine Heart Center,
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cases they have reviewed.[5] However, there has been no published report regarding the incidence of facial arteriovenous malformations locally due to the relative rarity of this anomaly.

Case

A 37 year old female presented with a huge facial mass and epistaxis. This facial mass noted at the left facial area started 23 years prior to admission. This was initially diagnosed as a hemangioma, and she was advised excision of the said mass.

This mass was initially a 1 cm x1 cm soft pulsating non tender mass on her left infraorbital area, which further enlarged to a size of 4 cm x 4 cm and extended to the frontal area. She became hypertensive three years prior to admission, and concurrent with her hypertension, the patient noted significant enlargement of the mass with associated protrusion of her left eye. Several days prior to admission, she experienced profuse nosebleeding from her left nostril (about half a glass) and bleeding from her left eye after she forgot to take her antihypertensive medication. The bleeding then resolved spontaneously.

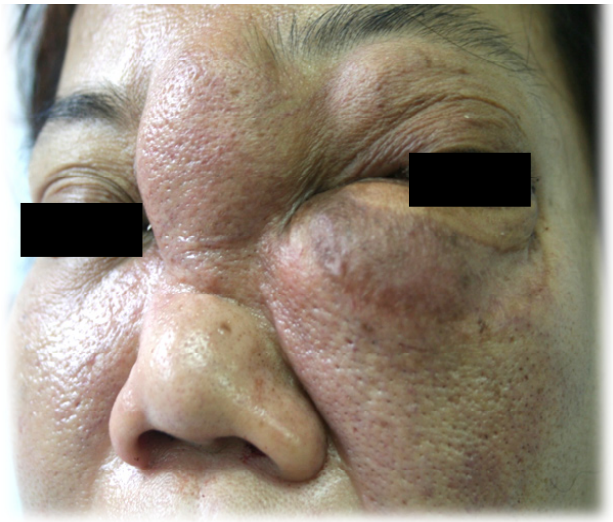

Figure 1. Preoperative picture of 37-year old female who presented with huge facial mass and epistaxis.

On the day of admission, she had another episode of epistaxis prompting admission for further work-ups.

Physical examination on admission showed intact extraocular movement and cone of light in both eyes. Her nasal septum was midline with blood clots noted along the septum and lateral nasal wall. The uvula and tongue were midline. There was a 4 cm x 4 cm soft nontender mass seen overlying the nasal bridge up to the medial canthus with associated prominent vascularization, pulsation and bruit. There is another soft nontender mass measuring 7 cm x 5 cm noted in the infraorbital area extending to the maxilla with associated bruit. (*Figure 1*)

With the admitting impression of hemangioma, she was referred for a four-vessel (4V) angiography. A Seldinger technique was done with a right femoral artery approach using a 5- french Berenstein catheter. Angiographic findings showed markedly tortuous bilateral external carotid arteries, arteriovenous malformation with multiple aneurysm formation in the left orbital and periorbital areas. The following feeding vessels were seen coming from bilateral ophthalmic, superficial temporal arteries, distal branches of both internal maxillary arteries, right facial and left superior alveolar arteries draining to the superficial temporal and external jugular veins. *(Figures 2a and 2c)*

Three weeks after angiography, she was referred back to our institution for super-selective embolization of the peri-orbital arteriovenous malformation. Branches of left external carotid artery were occluded, namely: 1. Internal maxillary artery, 2. Facial artery, 3. Superficial temporal artery. The particulate embolizing agents used were polyvinyl alcohol (PVA) and Gelfoam. (*Figures 2b and 2d).*

A week after the embolization, she then underwent wide excision of the arteriovenous

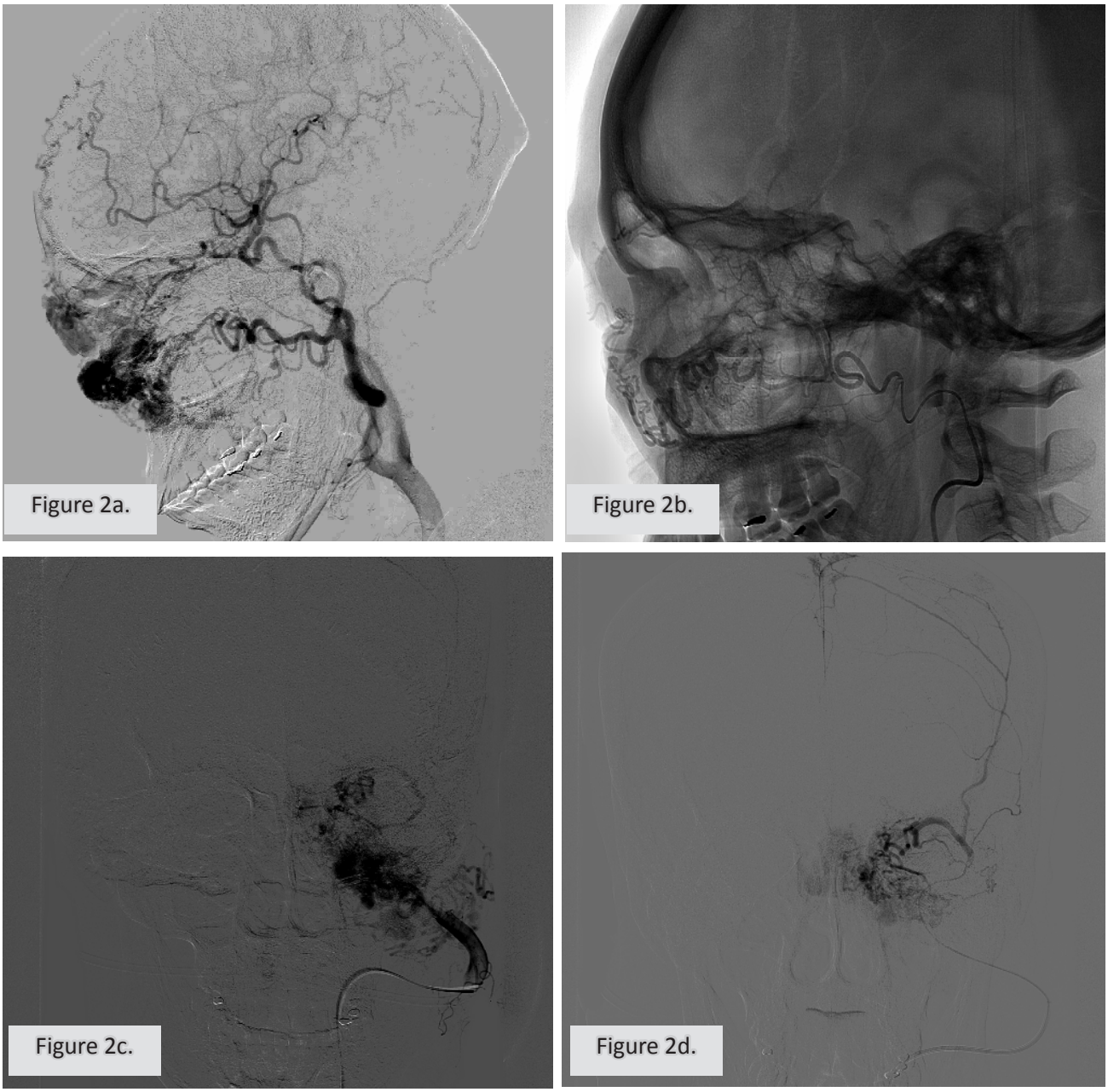

Figure 2. Four-vessel (4-V) angiographic images of a 37-year old female who presented with huge facial mass and epistaxis. Figure 2a is the lateral view of the facial AV malformation pre-operatively. Figure 2c is the anteroposterior view of the facial AV malformation pre-operatively. Figure 2b and 2d are taken post-operatively. Figure 2b is the lateral view, while Figure 2d is the anteroposterior view.

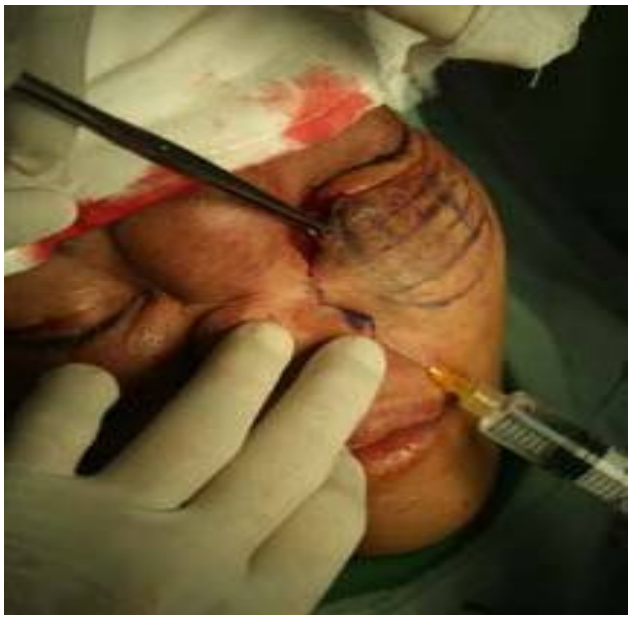

Figure 3. Intraoperative picture of a 37-year old female who presented with huge facial mass and epistaxis. She underwent wide excision of the AVM and facial reconstruction.

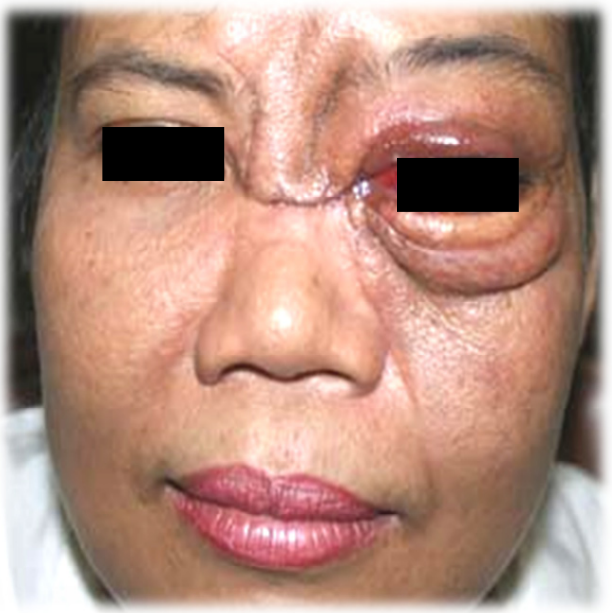

Figure 4. Postoperative picture of a 37-year old female who presented with huge facial mass and epistaxis.

malformation with reconstruction. (*Figure 3*) The said procedure was uneventful and the patient was discharged stable. (*Figure 4*)

Discussion

The approach to the management of vascular anomalies has evolved in the recent years. It now employs a multidisciplinary approach, involving clinicians, surgeons and radiologists, since no single specialty has sufficient skills and expertise to manage vascular anomalies that may involve any of the organ systems.[6]

Traditionally, high-flow anomalies such as arteriovenous lesions are managed by trans-arterial embolization.[7]

Lesions from the head and neck have a different presentation from that of the extremities since 'decompensation failure' is rare from AVMs from these sites. These extracranial AVMs usually occur in the more superficial layers and rarely communicate with the deep venous system.[7]

The usual symptoms of head and neck AVMs, such as cosmetic disfigurements and deformities, bleeding and ulceration, are related to the expansion of these lesions.[7]

Schrodinger proposed the staging of AVMs. The first stage shows pinkish-bluish stain and warmth of the skin. In the second stage, there is a pulsatile lesion with bruit and thrill. In the third stage, there are dystrophic skin changes, ulceration, bleeding and pain. In the fourth and final stage, there is already high-output cardiac failure.[7]

In asymptomatic patients, no treatment is necessary. However, if complications developed, such as bleeding, ulceration, pain or heart failure, treatment is warranted. Small and superficial AVMs can be resected by surgery. However, embolization is one of the treatment options for most vascular malformations. In this minimally invasive procedure, the arterial feeders are blocks that results to decrease in the size and blood flow and stabilizes the lesion. In some, serial embolizations can be employed to cause complete occlusion of the lesions. In selected patients, pre-operative embolization decreases intra- and post-operative risk.[7]

The patient belongs to the 3rd stage since she has a large facial mass with

discoloration of the overlying skin, pulsation, bruit, and experiencing profuse epistaxis already. The recommended management for her is embolization with excision of the mass.

The agents employed for AVM embolization include liquid adhesives, particles and alcohols. Polyvinyl alcohol (PVA) is usually used in the preoperative setting since particle agents are not considered as permanent embolic agent.[4] PVA, available as particles with a large range of sizes, is prepared through the reticulation of PVA with formaldehyde. A microcatheter is employed as a delivery catheter for particle size as large as 710 mm. Successful embolization is dependent on thrombus formation of the embolized vessel.[8]

Advantages of using PVA particle emboliza tion is its relative ease of use and favorable short-term histotoxicity. In large AVMs, a staged increase in particle size of PVA is necessary in particle embolization. "When flow in the AVM nidus has slowed significantly, final blockage can be achieved by using small microcoils."[4]

Gelfoam, a water-insoluble, off-white, nonelastic porous and pliable material, is one agent used to temporarily occlude abnormal vessels. This agent causes occlusion by forming nidus achieved thru holding many times its weight in blood and other fluids.[9]

Superselective embolization of the periorbital arteriovenous malformation was performed on the patient. The internal maxillary artery, facial artery, and superficial temporal artery, all branches of the external carotid artery, were occluded. The ophthalmic artery was spared to prevent blindness. The particulate embolizing agents used were polyvinyl alcohol (PVA) and Gelfoam. The former was used for the peripheral or distal vessels for its permanent effect and the latter was for the larger and more proximal vessels giving only a temporary effect. [9]

In some extracranial AVMs, "transarterial embolization with liquid adhesive material can achieve effective devascularization; but for preoperative devascularization to be effective, superselective catheterization of multiple feeding arteries and multiple injections are necessary.[10]

The goal of presurgical embolization are to decrease the nidus size of the AVM and to attempt to occlude deep, surgically inaccessible or deep arterial feeding vessels to facilitate surgical excision. Other goals of this procedure are to cause occlusion of intranidal aneurysms and high-flow fistulas to promote thrombus formation of the nidus of the AVM.[11]

Advantages of superselective preoperative embolization include reduction of blood loss and surgical time, "the applicability of strategically targeted embolization and the ability to occlude vessels deemed difficult to control by the operating surgeon, as well as the theoretical benefits of staging flow reduction in the nidus."[11] Failure to occlude the AVM nidus and proximal occlusion of the arterial feeding vessels will result to development of collaterals, which may have a detrimental effect on the surgical outcome.[11]

Conclusion

The usual approach in the management of arteriovenous malformation of the scalp and face is primarily thru a surgery, either by surgical excision or ligation of the feeding arteries. Concomitant with surgical excision is the risk of massive intraoperative hemorrhage and the need for reconstruction of the skin. On the other hand, ligation of the feeding arteries is problematic because of the recruitment of collateral vessel supply and the loss of access to the fistula for further embo-lization. With the introduction of new endovascular treatment techniques and new embolic agents, embolization in adjunct to surgery has become the treatment of choice for these lesions.

References

1. http://www. mayfieldclinic.com/ PE-AVM/ html.
2. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. Plast Reconstr Surg.1982 Mar;69(3):412-22.
3. Olivecrona H, Ladenheim J: Congenital arteriovenous aneurysms of the carotid and vertebral arterial systems, Berlin: 1957, Springer- Verlag. P-14 http://www. ijri.org./article
4. Koenigsberg RA. Brain, Arteriovenous Malformation. eMedicine Radiology. 2008. http://www. interestingradiology.blogspot.com/2008/12/emedicine-brain-arteriovenous/html
5. Huber P: Krayenbuhl / Yasargil Cerebral Angiography; 2nd completely rev. ed. Thieme Medical Publishers, Inc. New York. 1982; p373.
6. Murthy J. Vascular anomalies. Ind J Plast Surg. 2005;38(1):56-62
7. Chad S. Congenital vascular malformation. UTMB . 2006. http://www.utmb.edu/otoref/gmds/Congen-Vasc-Malfor-061220.doc
8. Konez O. Cho KJ (ed). Vascular Lesion Embolization Imaging. http://www.emedicine.medscape.com/article/419614/html.
9. Konez O. Hemangiomas and vascular anomalies. http://www.birthmarks.us/embolization.htm
10. Moon Hee H, Su OS, Hong Dae K, Kee-Hyun C, Kyung MY, Man Chung H. Craniofacial Arteriovenous Malformation: Preoperative Embolization with Direct Puncture and Injection of n-Butyl Cyanoacrylate. Radiology. 1999, 211, 661-6
11. Ogilvy CS, Stieg PE, Awad I, Brown RD, Kondziolka D, Rowenwasser R, et al. Recommendations for the Management of Intracranial Arteriovenous Malformations. Stroke. 2001;32:1458-71.