CASE REPORT

Management of a large diffuse maxillofacial arteriovenous malformation previously treated with ligation of ipsilateral arterial supply

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A 32 year-old male presented with a pulsatile facial mass with palpable thrill and audible bruit. Imaging revealed a very large diffuse left-sided facial arteriovenous malformation with extensive bilateral supply, as well as a previously ligated left external carotid artery. Endovascular treatment was required to control associated hemorrhagic events as well as for palliation and was delivered *via* the contralateral and ipsilateral collateral supply because of ligation of the direct route to the nidus. In addition, the patient received intravenous bevacizumab and intraarterial bleomycin therapy. Under such circumstances, endovascular embolization remains often the only option when emergent therapy for massive haemorrhage is required. Collaboration and treatment planning with head and neck surgery is imperative and should be performed from the onset, avoiding disastrous ligation of arterial feeders.

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Introduction

Arteriovenous malformations (AVMs) are congenital, non-involuting, vascular anomalies that may enlarge throughout an individual's life and have a variable pathogenesis. The head and neck region is the most common location for these vascular lesions outside of the brain. They represent an abnormal connection between one or more arteries and draining veins without an intervening capillary bed and are organized into tangled systems of vessels known as niduses, predisposing to pain, soft-tissue swelling, bleeding and infection. They can sometimes be part of a craniofacial arteriovenous metamerism syndrome involving specific intracranial and facial regions.¹ The facial lesions come to clinical attention due to associated haemorrhage,

cosmetic/psychosocial morbidity, or a combination of both. Depending on the location, magnitude and degree of extension, initial presentation may include haemorrhage, skin ulceration, loosening teeth, cardiac failure, pulsatile tinnitus, visual disturbances and epistaxis.²⁻⁴ Unfortunately, erroneous surgical interventions associated with proximal ligation of feeding arteries or similar interventional embolization with coil occlusion of feeding vessels will provoke development of indirect collateral supply and render subsequent management challenging.

AVMs can be identified clinically by the presence of erythema, warmth, palpable thrill or audible bruit, and diagnosis can be readily made on clinical examination. Attempt to biopsy or excise these lesions can cause massive haemorrhage and is strictly contraindicated. Before endovascular techniques were well established, surgical ligation was the mainstay of AVM management.

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However, microcatheter access and modern embolic agents have now established interventional treatment as the therapeutic technique with greatest efficacy and safety.⁴⁻⁸ The management team of a facial AVM should, from the onset, involve both head and neck surgery and interventional neuroradiology to facilitate the best possible outcome.

We report on a particularly challenging case of a large diffuse facial AVM with numerous indirect feeding arteries and emphasize the potential use of palliative and prophylactic embolization to mitigate further angiogenesis and manage symptomatic haemorrhagic events.

Case report

A 32-year-old individual with a steadily enlarging facial lesion since childhood, presented for possible treatment to our Interventional Neuroradiology service. Physical examination demonstrated a left maxillofacial pulsatile mass associated with a palpable thrill and audible bruit. Computed tomographic angiography and three-dimensional reconstruction demonstrated an extensive high-flow left-facial AVM with extensive perinidal angiogenesis and collateral vasculature. Imaging revealed secondarily recruited arterial supply from the stump of the ligated left external carotid artery (ECA), the internal carotid arteries bilaterally *via* trans skull base anastomosis and the ophthalmic systems, the ipsilateral vertebral artery and the right ECA (Figure 1). Review of his history revealed that the left ECA had been surgically ligated proximally in another country 3 years previously, restricting ipsilateral endovascular access to the nidus. In yet another country, 7 years prior, the lesion was misdiagnosed as a venous angioma and treated with pure ethanol sclerotherapy. This inadvertently caused a persistent hemiparesis for which rehabilitation was required. Subsequently, he had also undergone coil embolization for cheek, ear and gingival bleeding. Blood transfusions were frequently required.

Surgical removal even performed in combination with pre-operative embolization was at this point deemed to be of too high risk and not feasible due to the extensive involvement of the head and neck arterial and venous vasculature.

Before elective embolization could be arranged, the patient presented emergently with massive intraoral haemorrhage, and urgent transarterial glue embolization was successfully performed to arrest bleeding. Innumerable neovascular collateral channels arose over following repeat embolizations, which by their inherently fragile nature served as the potential source for future haemorrhage (Figure 2). Unfortunately, regional soft-tissue ischaemia resulted in further angiogenesis as well as cheek ulceration, and infection eventually necessitated drainage of a soft-tissue abscess. Eventually, he presented again with haemorrhage from a loose molar tooth and in collaboration with oral surgery, trans-arterial embolization and molar extraction were carried out successfully.

In view of the significant disease progression, experimental treatment with endovascular bevacizumab (400 mg of bevacizumab intravenously in 100 ml of normal saline, every 2 weeks) was carried out for 2 months. Although the overall mass effect of the lesion appeared to decrease in size following this treatment, ischaemic changes involving the skin became more prevalent, and this treatment was therefore discontinued.

He then underwent three sessions of intra-arterial infusion with bleomycin (intra-arterial infusion of 20 mg of bleomycin for 1 h) combined with gel foam embolization. Unfortunately, although this resulted in less frequent haemorrhage and decreased the overall mass effect, the skin over the lesion became more friable and he continued to have lip and cheek ulceration. He developed bacterial superinfection of these ulcers which was treated with cephalexin and mupirocin. The skin was covered by a soft conformable foam dressing. Subsequent to all of this, he also underwent two episodes of Onyx embolization, one of which was through a percutaneous approach.

Figure 1 (a) Diffuse facial arteriovenous malformation presenting as a steadily-growing fluctuant mass. (b) CT angiography, and (c) sagittal and (d) coronal three-dimensional reconstruction demonstrates supply to the arteriovenous malformation through left external carotid residual branches (post-ligation), with recruited supply from bilateral internal carotid arteries, vertebral arteries and the right external carotid artery.

The patient eventually succumbed to the disease from a combination of soft-tissue necrosis and septicaemia 5 years later (Figure 3).

Discussion

The differential diagnosis of head and neck vascular lesions includes facial AVM, venous malformation,

lymphatic malformation, capillary malformation and various vascular tumours. Their imaging appearances can mimic solid masses, and occasionally, angiography is necessary to identify vascular lesions.⁹ Once the diagnosis of a high-flow lesion has been made, facial AVMs are the main consideration. A subset of facial AVMs of modest size with compact nidus and relatively few arterial feeders can be cured by endovascular embolization if the nidus itself is obliterated with

Figure 2 (a) Arterial and (b) venous phase of anteroposterior projection right common carotid injection (arrow in a) shows diffuse arteriovenous shunting with innumerable collateral channels distal to the ligated external carotid artery feeding the lesion nidus (dashed arrow in a). (c) Arterial and (d) venous anteroposterior projection of the right facial artery demonstrates channels of high flow (arrow in c) which often provoked haemorrhage for our patient, with coils and liquid embolic material *in situ* (dashed arrow in c and e). (e) Arterial and (f) venous phase of left vertebral artery (arrow in e) injection demonstrate profound contralateral neoangiogenesis.

Figure 3 Follow-up with the patient 5 years after initial presentation shows drastic angiogenesis and skin breakdown following emergent treatments with *n*-butyl cyanoacrylate and polyvinyl alcohol particles, and elective treatments with endovascular bevacizumab and bleomycin.

a permanent embolic agent.¹⁰ Generally, cure of large-size facial AVMs represents a treatment challenge, even with modern endovascular and microsurgical advancements.¹¹ Stereotactic radiosurgery has no role in the management of this type of head and neck lesion. Unfortunately, there is still a lack of knowledge and understanding with respect to the fact that non-curative surgery or embolization should not be directed to proximally occlude arterial feeders. Such erroneous approach will prevent subsequent access to the nidus and worsen the long-term disease process by induction of angiogenesis and eventually lead to a disastrous outcome.¹⁰

Owing to the naturally rich arterial supply to the face, facial AVMs may possess a complex angioarchitecture, requiring a treatment plan of equal intricacy. In addition, a significant proportion of these AVMs are supplied bilaterally, in particular, when they extend towards the midline and/or are associated with very high flow. Embolization or surgery, as a unique treatment or in combination, has been shown to be potentially curative.^{10,12} However, surgery is not always feasible due to the diffuse and deep extension of some malformations and their extensive collateral recruitment, such as in our patient. Unfortunately, malformations of this complexity are mostly incurable with either embolization alone or post-embolization excision.^{6,11} Hence, the role of palliation in the setting of acute haemorrhage cannot be overstated, as it is often the only effective modality. In emergent situations,

palliations cannot always be delivered to the nidus. Unfortunately, embolizations which result in proximal occlusion of feeding arteries will likely provoke further angiogenesis, skin necrosis and other unwanted long-term complications such as heart failure.¹⁰

Our patient had, previously, the major supply to the AVM ligated, which prompted florid angiogenesis and collateral vessel development requiring future palliative embolizations to be performed *via* the contralateral external carotid and ipsilateral vertebral arteries. Angiogenesis occurs within weeks to months after improper embolization and proximal ligation.⁷ The angiogenetic arteries are predisposed to bleeding and are technically more difficult to penetrate in order to reach the nidus *via* endovascular methods.^{5,7} There are a number of important lessons to be learned from this case which can be generalized to large diffuse head and neck AVMs as a group.

The care team should be familiar with the various indications of consecutive embolization procedures. Unfortunately, the immediate clinical goal can be at odds with the delayed angiographic results of endovascular treatment. For instance, it is clear that the clinical goal of arresting haemorrhage is achieved by emergent embolization, but the long-term goal of lesion obliteration is not achieved and potentially rendered less feasible. Similarly, endovascular bleomycin reduced lesion bulk for our patient, but with drastic and undesirable cutaneous side effects.

It is important to note that our treatments were chosen in an environment where the original treatments with ligation and coiling of feeders precluded proper subsequent management.¹² Under such circumstances, subsequent mechanical obliteration is not likely and the role of antiangiogenic drugs will need to be further explored. In addition to bevacizumab and bleomycin, there is a renewed interest in the use of thalidomide as a therapeutic agent for these lesions.¹³ Pingyangmycin, another vascular endothelial growth factor inhibitor in the bleomycin family has been used with concomitant electrochemical therapy and mattress cerclage to some effect in selected patients.^{14,15}

Large diffuse AVMs of the head and neck region remain a major therapeutic challenge. A multidisciplinary treatment plan should be formulated at the beginning of the management process. Surgical or endovascular treatment with proximal ligation of feeding vessels is contraindicated, as it results in the subsequent development of new vessels precluding proper endovascular access to the nidus in later treatment sessions. The use of antiangiogenic drugs such as intravenous bevacizumab and intra-arterial bleomycin is still experimental for treatment of arteriovenous malformations and while unsuccessful in our patient, they could be further explored in other patients where proximal ligation of arterial feeders has mistakenly been performed or for palliative management of diffuse large head and neck arteriovenous malformations.

References

1. Bhattacharya JJ, Luo CB, Suh DC, Alvarez H, Rodesch G, Lasjaunias P. Wyburn-Mason or Bonnet-Dechaume-Blanc as cerebrofacial arteriovenous metamerism syndromes (CAMS)—a new concept and a new classification. *Interv Neuroradiol* 2001; **7**: 5–17.
2. Nagasaka S, Fukushima T, Goto K, Ohjimi H, Iwabuchi S, Maehara F. Treatment of scalp arteriovenous malformation. *Neurosurgery* 1996; **38**: 671–7. doi: <https://doi.org/10.1227/00006123-199604000-00007>
3. Goel S, Gupta S, Singh A, Prakash A, Ghosh S, Narang P. The current approach to the diagnosis of vascular anomalies of the head and neck: a pictorial essay. *Imaging Sci Dent* 2015; **45**: 123–31. doi: <https://doi.org/10.5624/isd.2015.45.2.123>
4. Buckmiller LM, Richter GT, Suen JY. Diagnosis and management of hemangiomas and vascular malformations of the head and neck. *Oral Dis* 2010; **16**: 405–18. doi: <https://doi.org/10.1111/j.1601-0825.2010.01661.x>
5. Marotta TR, Berenstein A, Zide B. The combined role of embolization and tissue expanders in the management of arteriovenous-malformations of the scalp. *Am J Neuroradiol* 1994; **15**: 1240–6.
6. Berenstein A, Scott J, Choi IS, Persky M. Percutaneous embolization of arteriovenous-fistulas of the external carotid-artery. *Am J Neuroradiol* 1986; **7**: 937–42.
7. Zheng JW, Zhou Q, Yang XJ, Wang YA, Fan XD, Zhou GY, et al. Treatment guideline for hemangiomas and vascular malformations of the head and neck. *Head Neck* 2010; **32**: 1088–98. doi: <https://doi.org/10.1002/hed.21274>
8. Cohen JE, Gomori JM, Grigoriadis S, Sibly Z, Rajz G. Complete and persistent occlusion of arteriovenous malformations of the mandible after endovascular embolization. *Neurol Res* 2009; **31**: 467–71. doi: <https://doi.org/10.1179/174313208X355468>
9. Dmytriw AA, Song JS, Gullane P, terBrugge KG, Yu E. Venous vascular malformation of the floor of mouth masquerading as a dermoid tumor. *Am J Otolaryngol* 2016; **37**: 383–6. doi: <https://doi.org/10.1016/j.amjoto.2015.12.008>
10. Dmytriw AA, Ter Brugge KG, Krings T, Agid R. Endovascular treatment of head and neck arteriovenous malformations. *Neuroradiology* 2014; **56**: 227–36. doi: <https://doi.org/10.1007/s00234-014-1328-0>
11. Churojana A, Khumtong R, Songsaeng D, Chongkolwatana C, Suthipongchai S. Life-threatening arteriovenous malformation of the maxillomandibular region and treatment outcomes. *Interv Neuroradiol* 2012; **18**: 49–59.
12. Morgan P, Keller R, Patel K. Evidence-based management of vascular malformations. *Facial Plast Surg* 2016; **32**: 162–76. doi: <https://doi.org/10.1055/s-0036-1581141>
13. Colletti G, Dalmonte P, Moneghini L, Ferrari D, Allevi F. Adjuvant role of anti-angiogenic drugs in the management of head and neck arteriovenous malformations. *Med Hypotheses* 2015; **85**: 298–302. doi: <https://doi.org/10.1016/j.mehy.2015.05.016>
14. Lasjaunias P, terBrugge KG, Berenstein A. Cerebrofacial arteriovenous metamerism syndrome. *Surgical neuroangiography: clinical and interventional aspects in children*. Berlin, Germany: Springer; 2006. pp. 359–88.
15. Cheng C, Xi SL, Xu DP, Zhai QK, Gong H, Wang XK. Mattress cerclage combined with electrochemical therapy and pingyangmycin injection after embolization for management of arteriovenous malformation on oral and maxillofacial region. *J Craniofac Surg* 2015; **26**: e110–13. doi: <https://doi.org/10.1097/SCS.0000000000001333>