Radiology Case Reports 16 (2021) 2202–2206

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

Giant parapharyngeal high-flow arteriovenous malformation causing airway compromise: emergency embolo-sclerotherapy via an endovascular-only approach ☆

Vishnu Naidu, MBBS, BSc, Ayyaz Quddus, BMBS, BMedSci, MRCS, FRCR, Ruhaid Khurram, MBBS, BMedSci (Hons), Priyesh Karia, MBBS, BSc, MRCS, FRCR, Jocelyn Brookes, MBBS, FRCP, FRCR*

Royal Free Hospital NHS Trust

ARTICLE INFO

Article history:
Received 7 May 2021
Accepted 8 May 2021

Keywords:
Arteriovenous malformations
Head and neck
Interventional radiology
Embolosclerotherapy

ABSTRACT

Head and neck arteriovenous malformations are the commonest extracranial vascular malformations but demonstrate a unique challenge in the limited available surgical options secondary to their intimate association to vital structures. We present a case of middle-aged female patient who presented with threatened upper-airway obstruction and bleeding secondary to a slowly enlarging parapharyngeal arteriovenous malformations. She was treated with an endovascular-only approach with the proximal arteriole branches selectively undergoing embolo-sclerotherapy with an optimal radiological and clinical outcome. We also demonstrate the utility of elective tracheostomy prior to intervention.

Introduction

Arteriovenous malformation (AVMs) are amongst a broad category of vascular malformations that are commonly acquired in the early gestational period, resulting in abnormal arterial and venous anatomy [1]. They are a high-flow, low resistance connection between dysplastic arteries and draining veins, connected by highly vascular nidi. Unlike capillaries, a nidus is unable to regulate high blood flow, and thus the draining veins expand to cope with the altered flow dynamics. AVMs most commonly occur intracranially; extracranial AVMs have a propensity to involve the head and neck with the majority of these involving the mid face and oral cavity [2–4].

AVMs are initially clinically indolent, but may become progressively symptomatic until adulthood, resulting in a variety of symptoms ranging from swelling and ulceration to treatment-refractory bleeding and congestive cardiac failure [5]. Proposed exacerbating factors include infection, trauma or

☆ Competing Interests: The authors have declared that no competing interests exist.

* Corresponding author. V.Naidu
E-mail address: vishnu.naidu@nhs.net (V. Naidu).

https://doi.org/10.1016/j.radcr.2021.05.027

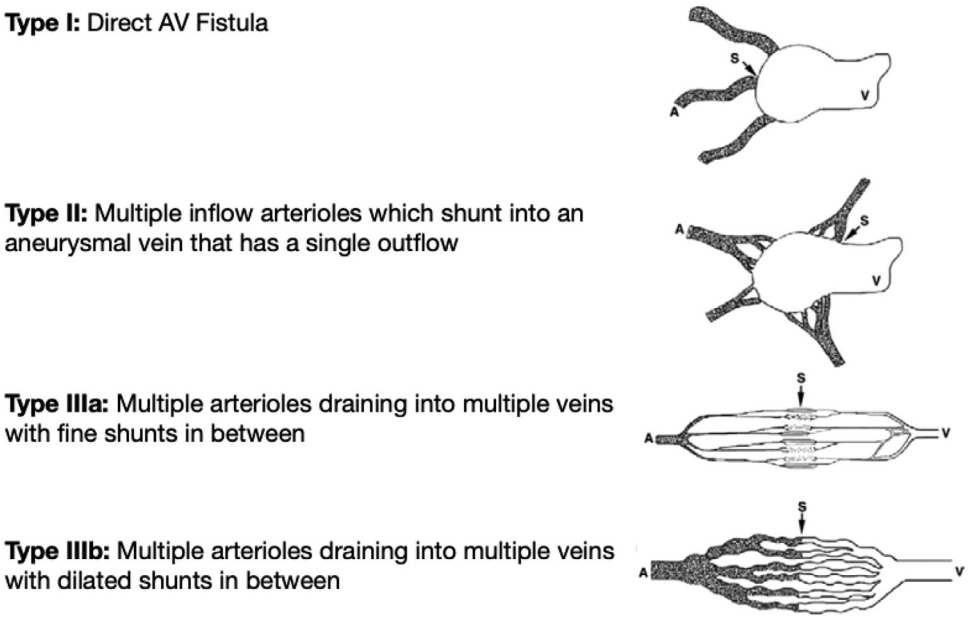

Fig. 1 – AVM classification by Cho et al.[8].

as a result of hormonal imbalances such as during pregnancy [6].

Clinically, the four-stage Schobinger system exists to classify AVMs, which may guide a decision as to whether treatment is warranted [7]. *Stage 1* describes the *quiescent* phase, whereby the lesion is asymptomatic but may be visible for example, a vascular skin stain. *Stage 2* refers to an *expansive* phase, which is a combination of *Stage 1* and a clinically pulsatile, tortuous AVM, which may invade deeper structures. *Stage 3* represents a *destructive* phase whereby an AVM, which in addition to *Stage 2*, causes dystrophic skin changes, skin ulceration and bleeding. *Stage 4* is the decompensative phase, whereby an AVM results in congestive cardiac failure alongside the features of *Stage 3* and is extremely rare [7].

Classification systems proposed by both Cho et al and Yakes et al. (see Figs. 1 and 2) alongside the introduction of new radiological techniques have enabled the angioarchitexture to be better interrogated and treatment planning and strategies to be optimized [8,9]. Treatment often include a combination of endovascular (transarterial, retrograde or percutaneous intranidal) and surgical management, but are generally bespoke based upon the angioarchetexture, location and presenting symptoms.

AVMs within the head and neck poses a unique challenge given the proximity to vital structures. We present a case of a patient presenting with treatment-refectory bleeding and threatened airway obstruction secondary to a giant neck AVM, which was successfully treated via an endovascular-only approach.

Case

A 42-year-old female presented to the A&E department, bleeding from the left ear which was controlled temporarily by packing. She had a 10-year history of surveillance of a complex, large high-flow, left-sided, parapharyngeal AVM causing symptoms of dysphagia, sleep apnoea and more recently, left-sided deafness.

On examination, significant swelling was seen in the left submandibular region and within the oral cavity, from the left buccal membrane extending to the right tonsillar arch. She had threatened airway obstruction, with a Mallampati class of III/IV and her oxygen saturations were 92% on air with a respiratory rate of 21. She was haemodynamically stable, with a heart rate of 80 bpm and a blood pressure of 114/86 mmHg. In addition, there was no focal neurology.

A contrast-enhanced CT of the neck demonstrated an 11 × 10 × 10 cm left-sided, left paraphyngeal high-flow AVM drawing from left carotid artery with early fistulation to left jugular vein, threatened airway obstruction, and dilated vessels adjacent to the left external auditory canal (see Fig. 3).

Following MDT discussion with the regional AVM centre, she underwent an urgent tracheostomy to secure the airway while arranging transfer (see Fig. 4).

Embolosclerotherapy under general anaesthetic was carried out taking a right femoral approach to access the left common carotid artery. After DSA assessment, selective catheterization of the left external carotid artery branches was then carried out in a systemic fashion, accessing different fistulating nidi of the AVM in turn. Sclerosis of the nidi was carried out transarterially using 3% STS foam, dehydrated ethanol and gelfoam sequentially under fluoroscopic guidance with no percutaneous intervention as the venous origins of the nidi were inaccessible behind the ramus of the mandible.

On recovery, there was no significant pain except around the new tracheostomy. No visual or neurological deficit was demonstrated. Overnight, the patient's symptoms of dysphagia and dyspnoea had significantly improved with almost complete devascularisation of the nidus radiologically (see Fig. 3). The tracheostomy was removed successfully at 10 days and the patient discharged home pending further follow up.

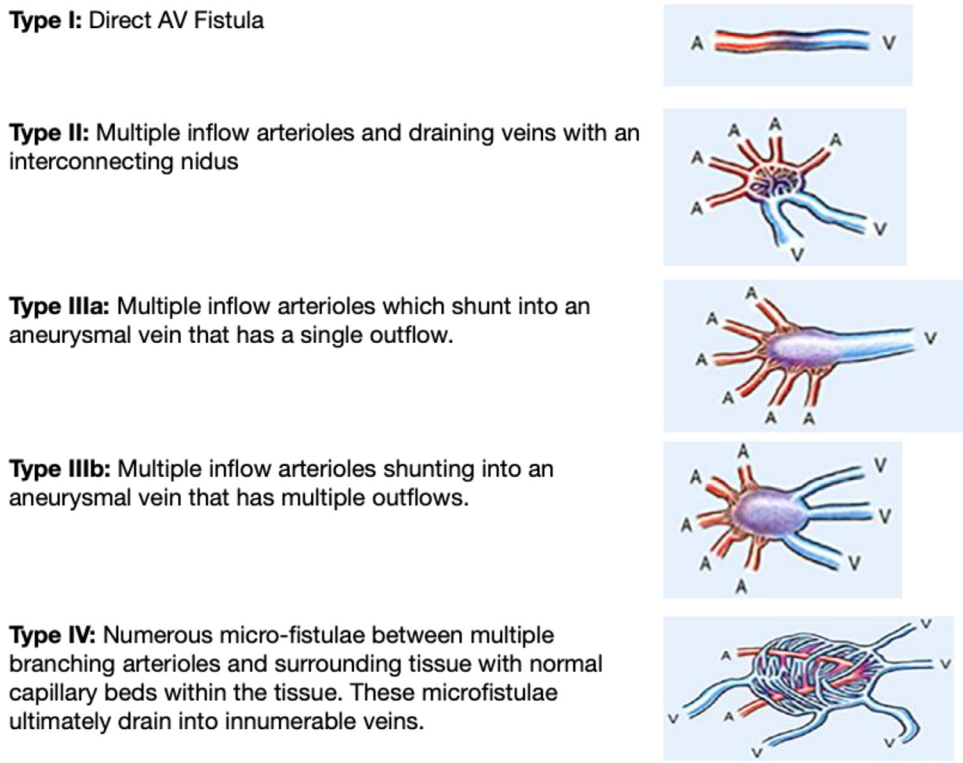

Fig. 2 – AVM classification by Yakes et al.[9].

Discussion

We demonstrate a case of a giant high-flow, airway-obstructive and bleeding parapharyngeal AVM which was successfully treated with a transarterial-only approach.

Cases of AVMs causing extrinsic airway obstruction have been described in the literature [10,11]. However, these patients had unfortunately presented very late, with treatment either not being possible or a surgical approach being taken. To our knowledge, this is the first case in the literature of a head and neck AVM causing extrinsic airway obstruction, successfully treated with a transarterial-only approach.

Investigation into AVMs starts with clinical examination and classification with the Schobinger system. The initial radiological mode of investigation is Doppler ultrasonography, which allows spectral analysis of the feeding vessels. Both arterial and venous limbs would be dilated, with increased systolic/diastolic velocities seen on the arterial side and an arterialized waveform on the venous side. The nidus appears as a hypervascular lesion, which is indistinguishable from a hypervascular tumour. The next mode of investigation includes either CT or MR angiography. This allows the angioarchitexture of the AVM to be interrogated to allow planning. MRI has the additional advantage of assessing for local soft tissue invasion. However, digital subtraction angiography remains gold-standard for pre-embolisation angiographic planning [2,12].

Treatment approaches to AVM, are premised on shutting down the fistulating nidus by means of embolization, and sclerosis of the causative endothelial cells in the wall of the nidus itself. Recognition and access to the nidus usually involves a combination of angiography to display the anatomy of the nidus and direct percutaneous puncture of the nidus because direct trans-arterial access is rarely possible. Cho and Yakes both devised anatomical classification of AVMs (see Fig. 1 and 2), which enables the best therapeutic approach to be chosen; transarterial, retrograde, percutaneous (intranidal) or a combination [8,9]. With our case, and with most head and neck AVMs, the classification was deemed as a Yakes type II/ Cho type IIIb, in which the lesion is characterised with multiple inflow arteries and draining veins with a hypervascular nidus connecting the two.

Cervical AVMs of the head and neck are particularly challenging due to their proximity to vital structures, especially neurovascular bundles and brain, whilst surgery carries a prohibitively high risk of intra-operative bleeding. On this occasion, it was possible to negotiate deeply into the AVM which had not previously been disturbed by surgical or angiographic procedures, and treat the several distinct AVM nidi systematically by direct trans-arterial approach (using a micro-catheter) without requiring percutaneous access.

Liquid embolic agents are generally used to treat AVMs via the arterial or percutaneous route; proximal arterial embolisation with coils are avoided due to their association with ischemia-related angiogenesis and high likelihood of recurrence [13,14]. The liquid embolics generally used are ethanol, ethylene vinyl alcohol copolymer (EVOH also known as Onyx) and glue. Venous occlusion is variable and may be accompolished with a combination of mechanical occlusion, coils, and liquid agents [2]. Ethanol is the only agent that is potentially

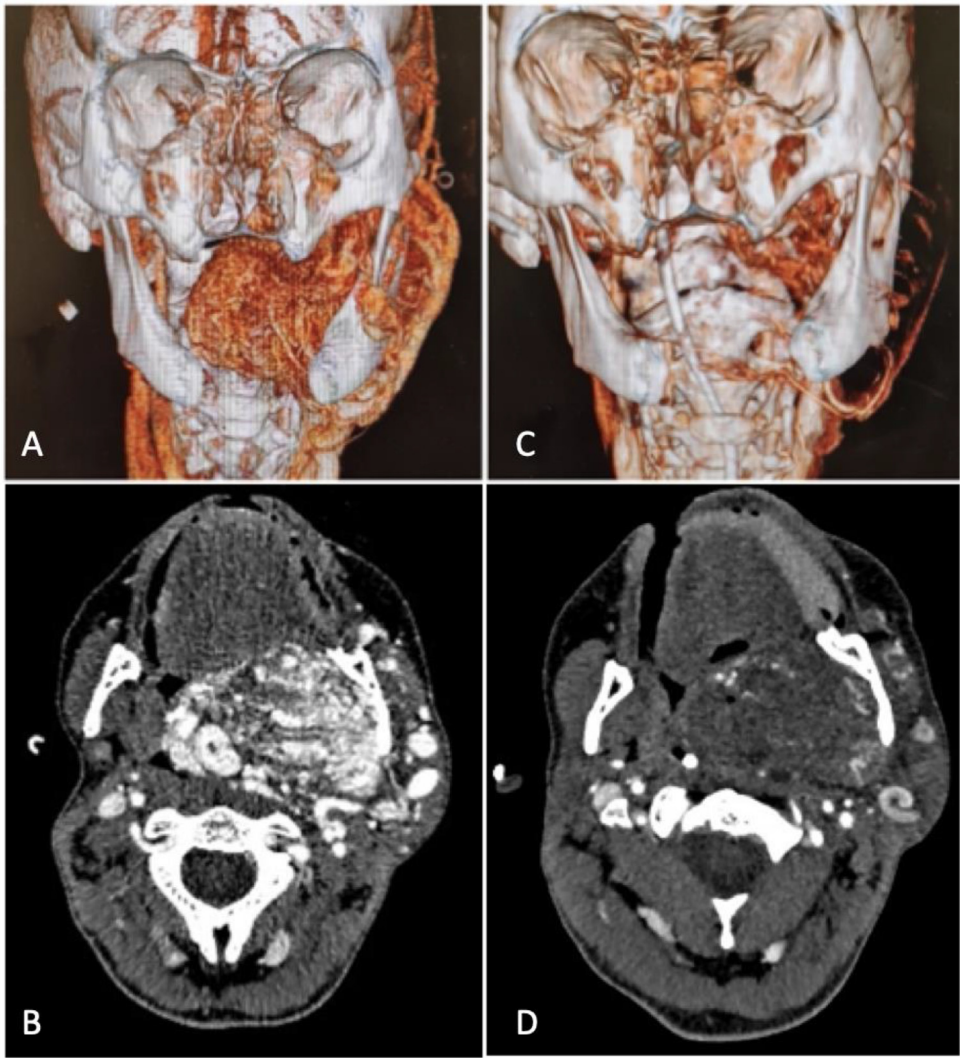

Fig. 3 – (A): Volume rendered CTA showing giant left-sided parapharyngeal AVM feeding from left carotid on presentation. (B) Same pre-treatment CTA but in axial section showing gross swelling of left-sided AVM across midline, threatening airway. (C) Same format 48 hours after embolo-sclerotherapy showing significant clearing of the AVM. NG tube in situ. (D) Post-op CTA axial section, showing substantial devascularisation of the AVM with preliminary reduction in volume.

curative; it's mechanism of action is centred on the destruction of the internal lamina of endothelial walls, which promotes thrombosis but also inhibits secretion of angiogenesis related factors [15,16]. The danger of ethanol includes potential pulmonary arterial spasm, leading to systemic collapse; this is why anaesthetic support and close pulmonary artery monitoring is paramount during embolosclerotherapy [17].

Conclusion

AVMs are rare, but can present with catastrophic consequences. An awareness of their clinical appearance and the investigations of choice is important for swift diagnosis and timely management. Head and neck AVMs are often associated with both cosmetic issues as well as serious consequences including life-threatening airway obstruction. Our case demonstrates two potentially life-threatening consequences; bleeding and airway obstruction, both of which were mitigated by a multidisciplinary decision for an endovascular-only approach with an optimal therapeutic and cosmetic outcome. It also demonstrates the utility of elective tracheostomy for treatment of head and neck AVM

Patient consent statement

The patient has provided written consent with regards to publication of their case.

They understand the following:

1. The Information will be published without my name attached and every attempt will be made to ensure

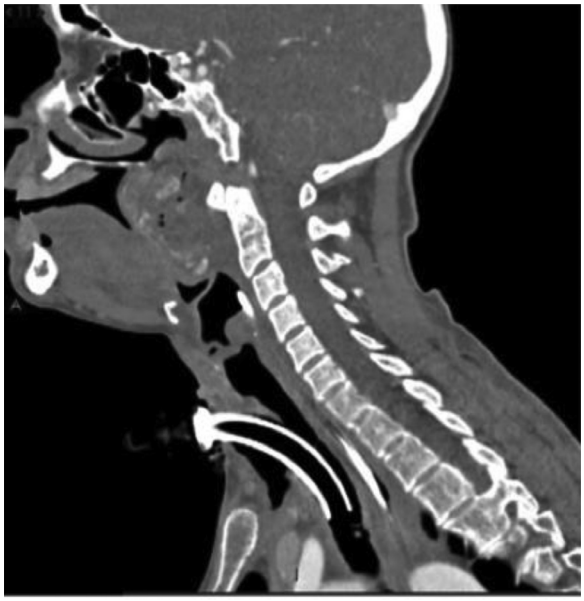

Fig. 4 – (E): Post-op CTA at 48 hours sagittal section showing substantially devascularised pharyngeal AVM (gas bubbles from treatment foam) and elective tracheostomy.

anonymity. I understand, however, that complete anonymity cannot be guaranteed. It is possible that somebody somewhere - perhaps, for example, somebody who looked after me if I was in hospital, or a relative - may identify me.

2. The Information may be published in a journal which is read worldwide or an online journal. Journals are aimed mainly at health care professionals but may be seen by many non-doctors, including journalists.
3. The Information may be placed on a website.
4. I can withdraw my consent at any time before online publication, but once the Information has been committed to publication it will not be possible to withdraw the consent.

REFERENCES

[1] Yakes WF, Rossi P, Odink H. How I do it. Arteriovenous malformation management. Cardiovasc Intervent Radiol 1996;19(02):65–71. doi:10.1007/BF02563895.

[2] Gallant SC, Chewning RH, Orbach DB, Trenor CC 3rd, Cunningham MJ. Contemporary management of vascular anomalies of the head and neck-Part 1: Vascular Malformations: A Review. JAMA Otolaryngol Head Neck Surg 2021;147(2):197–206. doi:10.1001/jamaoto.2020.4353.

[3] Fowell C, Jones R, Nishikawa H, Monaghan A. Arteriovenous malformations of the head and neck: current concepts in management. Br J Oral Maxillofac Surg 2016;54(5):482–7. doi:10.1016/j.bjoms.2016.01.034.

[4] Richter GT, Suen JY. Clinical course of arteriovenous malformations of the head and neck: a case series. Otolaryngol Head Neck Surg 2010;142(2):184–90. doi:10.1016/j.otohns.2009.10.023.

[5] Caruqeja IM, Sousa J, Mansilha A. Vascular malformations: classification, diagnosis and treatment. Int Angiol 2018;37(2):127–42. doi:10.23736/S0392-9590.18.03961-5.

[6] Elluru RG, Azizkhan RG. Cervicofacial vascular anomalies. II. Vascular malformations. Semin Pediatr Surg 2006;15(2):133–9.

[7] Finn M C, Glowacki J, Mulliken J B. Congenital vascular lesions: clinical application of a new classification. J Pediatr Surg 1983;18(06):894–900.

[8] Cho SK, Do YS, Shin SW, Kim D, Kim YW, Park KB, et al. Arteriovenous malformations of the body and extremities: analysis of therapeutic outcomes and approaches according to a modified angiographic classification. J Endovasc Ther 2006;13(4):527–38. doi:10.1583/05-1769.

[9] Yakes W, Baumgartner I. Interventional treatment of arteriovenous malformations. Gefasschirurgie 2014;19(04):325–30.

[10] McLoughlin JM, McBrien ME. Acute upper airway obstruction secondary to late presentation of a massive oropharyngeal arteriovenous malformation. J Clin Anesth 2009;21(4):278–81 Epub 2009 Jun 6PMID:19502034 . doi:10.1016/j.jclinane.2008.07.006.

[11] Nabawi A, Abraham N, Nabawi A. Disfiguring high-flow cervicofacial arteriovenous malformations. J Surg Case Rep 2020;2020(10):rjaa435 PMID:33101645PMCID: PMC7569479. doi:10.1093/jscr/rjaa435.

[12] Gilbert P, Dubois J, Giroux MF, Soulez G. New treatment approaches to arteriovenous malformations. Semin Intervent Radiol 2017;34(3):258–71. doi:10.1055/s-0037-1604299.

[13] Alomari A, Dubois J. Interventional management of vascular malformations. Tech Vasc Interv Radiol 2011;14(1):22–31. doi:10.1053/j.tvir. 2010.07.006.

[14] Jin Y, Zou Y, Hua C, Chen H, Yang X, Ma G, et al. Treatmentof early-stage extracranial arteriovenous malformations with intralesional interstitial bleomycin injection: a pilot study. Radiology 2018;287(1):194–204. doi:10.1148/radiol.2017162076.

[15] Prasetyono TO, Kreshanti P. Efficacy of intra-lesional alcohol injection as alternative and/or complementary treatment of vascular malformations: a systematic review. J Plast Reconstr Aesthet Surg 2010;63(07):1071–9. doi:10.1016/j.bjps.2009.04.020.

[16] Su L, Wang D, Han Y, Wang Z, Zheng L, Fan X. Absolute ethanol embolization of infiltrating-diffuse extracranial arteriovenous malformations in the head and neck. Eur J Vasc Endovasc Surg 2015;50(1):114–21. doi:10.1016/j.ejvs.2015.02.020.

[17] Shin BS, Do YS, Cho HS, Kim DI, Hahm TS, Kim CS, et al. Effects of repeat bolus ethanol injections on cardiopulmonary hemodynamic changes during embolotherapy of arteriovenous malformations of the extremities. J Vasc Interv Radiol 2010;21(1):81–9. doi:10.1016/j.jvir.2009.09.026.