Head and neck arteriovenous malformation: A rare case and review of literature

Danu Yudistira, MD[1,2], Sagung Rai Indrasari, MD[1,2], Camelia Herdini, MD[1,2], Agustian Winarno Putra, MD[1,2]

[1]Department of Otorhinolaryngology-Head and Neck Surgery, Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada, Yogyakarta, Indonesia, [2]Dr. Sardjito General Hospital, Yogyakarta, Indonesia

SUMMARY

Arteriovenous malformation (AVM) of the head and neck region is an extremely rare existence. This condition can lead to various complications due to the surrounding complex anatomical structures of the head and neck. From the clinical manifestation, AVM mass can be easily mistaken for other soft tissue masses. The gold standard diagnostic procedure for AVM is angiography. The current approach in managing AVM involves a combination of conventional surgical technique and endovascular procedure. The ultimate goal is to achieve either complete removal of the nidus or total occlusion of its blood flow. The management of head and neck AVM demands a comprehensive, multidisciplinary approach involving various medical specialties.

INTRODUCTION

Arteriovenous malformation (AVM) of the head and neck region is an extremely rare existence. This condition can lead to various complications due to the surrounding complex anatomical structures of the head and neck. From the clinical manifestation, AVM mass can be easily mistaken for other soft tissue masses. The gold standard diagnostic procedure for AVM is angiography. The current approach in managing AVM involves a combination of conventional surgical technique and endovascular procedure. The ultimate goal is to achieve either complete removal of the nidus or total occlusion of its blood flow. The management of head and neck AVM demands a comprehensive, multidisciplinary approach involving various medical specialties.

CASE PRESENTATION

A 20-year-old male presented with a mass on the right neck. The patient said that the swelling initially was as big as a peanut and had been present since childhood. However, the swelling started growing larger until it reached the size of a tennis ball within a period of 4 months. The patient felt difficulty and pain during swallowing but denied any difficulty of breathing or hoarseness. Physical examination revealed significant swelling within the right neck region with clear margin and soft consistency upon palpation. From the cranial nerve examination, all was within normal limits. The patient was haemodynamically stable with normal vital signs.

Laryngoscopy showed a vascularised mass at the right vallecula, deviating the epiglottis to the left. The mass extended into the supraglottic region (Fig. 1). The vocal cord movement was symmetrical during adduction and abduction. A contrast-enhanced computed tomography (CT) of the neck demonstrated a well-margined amorphous mass with the dimension of 4.1 × 5.3 × 5.6 cm in right neck region with density of 41 Hounsfield units (HU) precontrast and lined up to 58 HU postcontrast. The mass drawing vascularisation from right external carotid artery and superior right thyroid artery was also seen from the CT result. Thus, a high vascularised mass was suspected.

A multidisciplinary meeting with the interventional radiologist and anaesthesiologist was held to decide the best approach to treat the patient. Because of the suspicion of highly vascularised mass in the neck which extend into the supraglottic region, it was then decided for the patients to have tracheostomy to secure the airway before angiography. the interventional radiologist would confirm the AVM during angiography and proceed with the embolisation.

Embolisation under general anaesthesia was carried out. The procedure involved a right femoral approach to access the right common carotid artery. After digital subtraction angiography (DSA) was assessed, selective catheterisation of the right external carotid artery branches was then carried out in a systemic fashion, followed by transarterial embolisation using polyvinyl alcohol (PVA) sequentially under fluoroscopic guidance (Fig. 2).

During recovery, there was no significant pain except around the recent tracheostomy site. No visual or neurological deficit was found. Within one night, the patient's symptoms of dysphagia had significantly improved with almost complete devascularisation of the mass radiologically, the patient was then discharged home one day after.

Follow up was done 1 month after the embolization to detect any early signs of recurrence and to plan possible decannulation procedure when the airway was completely safe. At follow-up, there was no clinical complaints nor any enlargement of the neck and no signs of complication from the embolisation procedure (Fig. 3). A follow up, contrast-enhanced CT angiography of the neck was done for evaluation and detected shrinkage of the mass by 32%. There was a hypovascularised mass with size of 3.6 × 4.6 × 5 cm without airway narrowing. Thus, patient was instructed to undergo routine long-term follow-up to assess the progress of his condition.

This article was accepted: 13 February 2024
Corresponding Author: Danu Yudistira
Email: dhanoetht@gmail.com

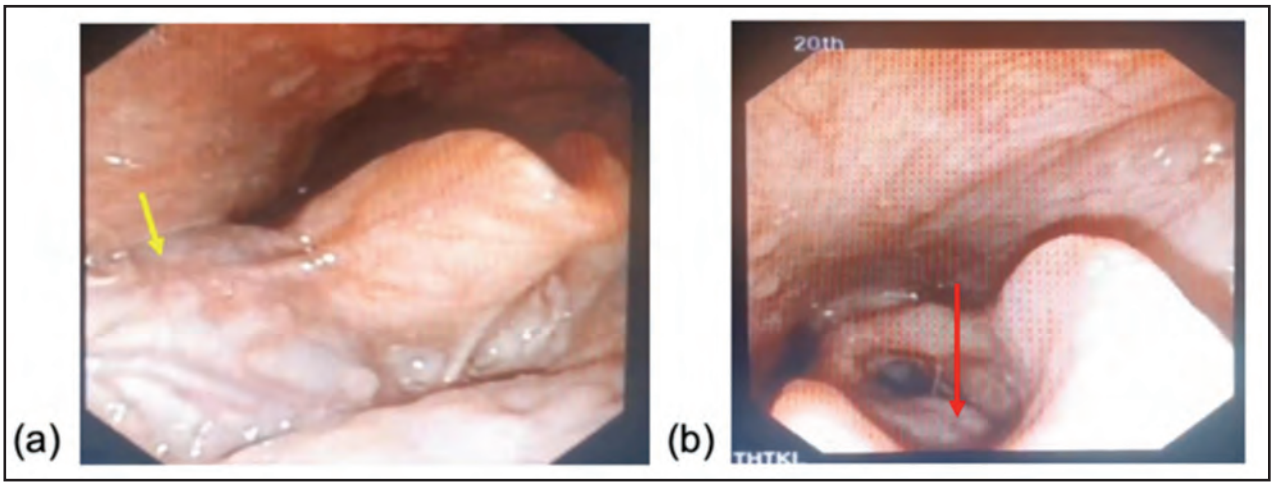

Fig. 1: Laryngoscopy a) a mass on the right vallecula deviating the epiglottic to the left (yellow arrow), b) a mass on the supraglottic area, the vocal cord showed symmetrical movement (red arrow)

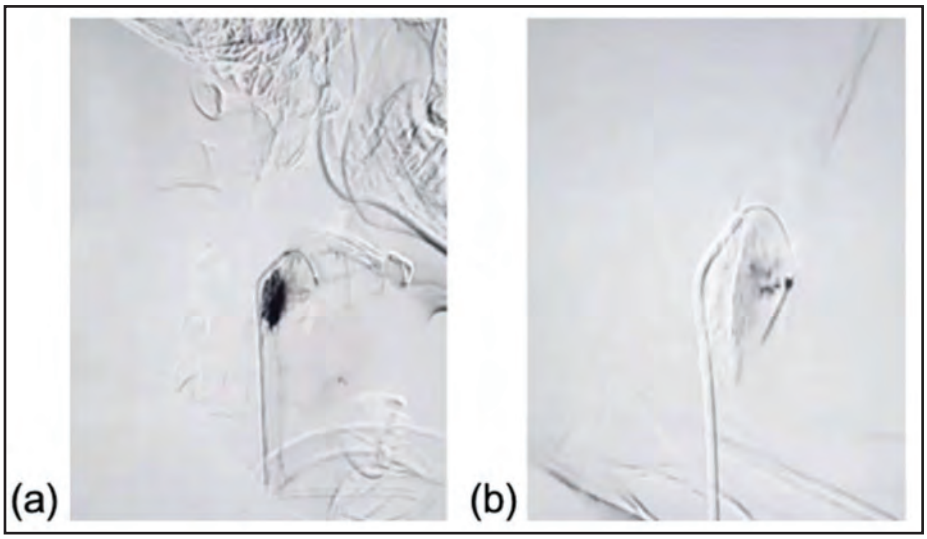

Fig. 2: Angiogram showing the AVM nidus drawing from right carotid artery and right superior thyroid artery. a) Pre-embolisation – showing tumour blush appearance. b) Post-embolisation – showing no visible blushing

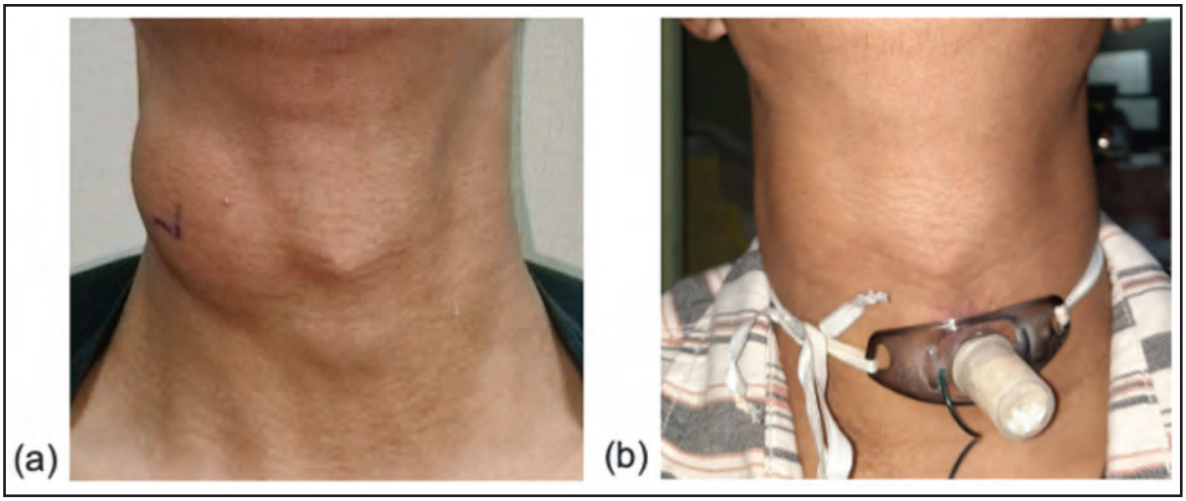

Fig. 3: Clinical picture of the patient before and after embolisation. a) Pre-embolisation. b) Post-embolisation

DISCUSSION

AVMs fall under the category of congenital high-flow vascular anomalies, constituting a mere 4.7% of all vascular irregularities. While AVMs most frequently occur within the central nervous system, they are seldom found extracranially.[3] AVMs usually exist as clinically dormant but may become progressively symptomatic until adulthood, leading to disfiguring and life-threatening complications. Contributing factors might include infection, injury, or hormonal imbalances during phases like pregnancy or puberty.[2] Their diagnosis relies on complete patient history, clinical assessment and specific imaging traits. There are various classification systems for AVMs (Table I).[4]

AVMs have diverse clinical presentations and pose challenges to manage due to high rates of recurrence irrespective of therapeutic approach. The classification and management of AVMs have considerably changed in the last few years. Endovascular treatment is currently almost always part of the treatment of AVMs often combined with surgical resection.[1] AVMs within the head and neck poses a unique challenge given the proximity to vital structures.[5]

In our case, for radiological imaging we use contrast-enhanced CT scan and angiography. CT scan was a useful diagnostic imaging tool for its high spatial resolution and quick scanning time, it also could detect soft tissue abnormalities and bone involvement. It provided detailed evaluation of the angiographic structure of the mass which in our case was originated from right external carotid and right superior thyroid artery. Head and neck AVM may be fed by multiple or bilateral arteries including external and internal carotid, vertebral, or subclavian artery depending on the distribution around the region. Arteriography remains the gold standard to diagnose AVM is mainly characterised by tortuous, dilated arteries with arteriovenous shunting and enlarged draining veins. Even though in this case we did not use ultrasonography with colour doppler, it is an accessible and inexpensive modality to assess disease progression and treatment outcomes.[1] Biopsy is not necessary to diagnose AVM and should be avoided as far as possible due to the risk of bleeding and the possibility of triggering growth of an AVM.[6]

The available literature highlights instances of high success rates in treating head and neck AVM using a singular treatment approach, particularly emphasising embolisation. Various embolic material can be used for the endovascular treatment of the AVM. It has been recommended that transarterial embolisation with slow injection with penetration to draining veins along with shunt points using low concentration n-butyl-2-cyanoacrylate (NBCA) can achieve better embolisation effect of AVMs.[7] Although there had been many results reported of a vast variety of embolic agents for transarterial embolisation, the use of NBCA had become the most commonly utilised due to its proven properties and effectiveness. This liquid embolic material was frequently used for its permanent embolic effect and wide blockage from the vascular feeder to drainers via fistula. NBCA is considered to depict high thrombogenicity compared to other liquid embolic materials.[7]

The application and selection of embolic agents in the management of head and neck AVMs depend on several factors like the vascular characteristics, the AVMs classification, the expertise of the treatment team, and the availability of the embolic agents, also the main goal of the procedure. We used transarterial PVA which is a permanent type of agent used worldwide. In our case, we use PVA because it was readily available, inexpensive, and has good capillary control.[7] Thus, the outcome of our case still obtains quite satisfactory results in the aspect of function and appearance. The main focus of our treatment was to preserve the surrounding anatomical structure of the neck region in order to avoid potential complication with swallowing and breathing. Ischemic complications in the nervous system may arise due to the migration of embolic materials into the cerebral arteries or the vessels that supply the cranial nerves, known as the vasa nervorum. Such occurrences can result in mortality or severe neurological deficits. To mitigate these risks, particularly in embolisation involving liquid embolic material, it is crucial to carefully observe and address angiographic findings demonstrating the neural arteries. Injection of liquid embolic material or small-sized particles is contraindicated in the head and neck arteries that have anastomotic channels with cerebral arteries and vasa nervorum.[7]

Surgical treatment alone might be considered the initial treatment choice for smaller, isolated AVM characterised by well-defined feeding vessels and without any involvement of bones. For larger or previously treated AVM, a combination of super selective embolisation and surgical intervention has been shown to yield better intraoperative management and the highest rates of success.[1]

The patient was instructed to undergo routine long-term follow-ups because of the possibility of recurrence, as reports suggest that it can occur in as many as 80% of cases following embolisation or resection. Incomplete removal or embolisation of the nidus can stimulate aggressive growth of the remaining lesion, leading to a risk of progression as high as 50% within the initial five years. Furthermore, recurrences have even been observed a decade after treatment, underlining the necessity of extended post-treatment follow-up for timely detection. It's important to consider that the interpretation of the term 'cure' varies within the literature and reported instances of 'cure' might be influenced by limited follow-up periods. Some reported cases of 'cure' refer to an asymptomatic state following embolisation, rather than a complete absence of the condition.[1]

CONCLUSION

Head and neck arteriovenous malformations (AVMs) are rare vascular abnormality that is not commonly found in clinical practice. Because they come with such dire consequences often associated with both cosmetic issues as well as serious consequences including life-threatening bleeding, swallowing problem and airway obstruction. Awareness of the clinical appearances and correct choice of modalities is important to uphold swift diagnosis and accurate management. Our case demonstrated head and neck AVM treatment based on multidisciplinary decision for a single

Table I: Overview of various classification systems for AVMs.[4]

Focal vs diffuse		Schobinger	Suen-Richter
Focal	Diffuse	Stage I: Quiescence, cutaneous blush, warmth	T: Size of AVM T_1: 1 Cervicofacial subunit T_2: 2 Cervicofacila subunit T_3: 3 Ccervicofacila subunit T_4: Bilateral/multifocal disease
Discrete border with central nidus	Multiple or no discrete nidus	Stage II: Expansion; active growth, pulsations, bruit	D: Depth of AVM invasion D_1: Skin and/or subcutaneous involvement D_2: Subcutaneous and muscle involvement D_3: Subcutaneous, muscle, and cartilage or bone involvement D_4: Skull base or intracranial extension
Firm to palpation	Compressible with rapid rebound	Stage III: Destruction; same as stage II but symptomatic (pain, bleeding, disfigurement)	S: Schobinger stage modified: Kohout and colleague S_0: Quiscence S_1: Expansion (Bruit, pulsation, rapid growth S_2: Destruction (Ulceration, bleeding, pain)
1-2 arterial feeders	Multiple arterial feeders	Stage IV: Decompensation; same as stage III but with high- output cardiac failure	Stages: Stage I: $T_{1\text{-}2}$ D_1S_0, $T_1D_1S_1$, $T_1D_2S_0$ Stage II: $T_1D_3S_0$, $T_2D_{1\text{-}2}$ $S_{1\text{-}2}$, $T_2D_2S_0$ Stage III: $T_1D_3S_{1\text{-}2}$, $T_3D_{1\text{-}2}S0$, $T_2D_3S_{0\text{-}2}$ Stage IV: $T_3D_3S_{0\text{-}2}$, any D_4, any T_4
Good treatment outcomes	Higher risk of recurrence	-	-

endovascular approach with an optimal functional and cosmetic outcome.

ACKNOWLEDGMENTS

The authors would like to thank all the hospital staff who were involved in the patient's care.

DECLARATION

The authors declare no conflict of interest.

REFERENCES

1. Fernández-Alvarez V, Suárez C, de Bree R, Nixon IJ, Mäkitie AA, Rinaldo A, et al. Management of extracranial arteriovenous malformations of the head and neck. Auris Nasus Larynx 2020; 47(2): 181-90.
2. Cuong TC, Thang LM, Giang NL, Trung NA, Qui LV, Tha TTT, et al. Recurrent head and neck arteriovenous malformations: a case report. Radiol Case Rep 2023; 18(3): 766-70.
3. Eivazi B, Werner JA. Extracranial vascular malformations (hemangiomas and vascular malformations) in children and adolescents – diagnosis, clinic, and therapy. GMS Curr Top Otorhinolaryngol Head Neck Surg 2014; 13: 1-19.
4. Rosenberg TL, Suen JY, Richter GT. Arteriovenous malformations of the head and neck. Otolaryngol Clin North Am 2018; 51(1): 185-95.
5. Naidu V, Quddus A, Khurram R, Karia P, Brookes J. Giant parapharyngeal high-flow arteriovenous malformation causing airway compromise: emergency embolo-sclerotherapy via an endovascular-only approach. Radiol Case Rep 2021; 16(8): 2202-6.
6. De Miguel R, López-Gutierrez JC, Boixeda P. Arteriovenous malformations: a diagnostic and therapeutic challenge. Actas Dermo-Sifiliográficas 2014; 105(4): 347-58.
7. Tanoue S, Tanaka N, Koganemaru M, Kuhara A, Kugiyama T, Sawano M, et al. Head and neck arteriovenous malformations: clinical manifestations and endovascular treatments. Interv Radiol 2023; 8(2): 23-35.