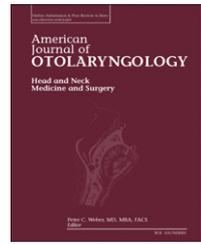


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# Hypopharyngeal venous malformation presenting with foreign body sensation and dysphagia

Andrew M. Vahabzadeh-Hagh, MD<sup>a,\*</sup>, Ali R. Sepahdari, MD<sup>b</sup>, Jayson Fitter<sup>a</sup>, Elliot Abemayor, MD, PhD<sup>a</sup>

<sup>a</sup> Department of Head and Neck Surgery, David Geffen School of Medicine at UCLA, Los Angeles, CA USA

<sup>b</sup> Department of Radiological Sciences, David Geffen School of Medicine at UCLA, Los Angeles, CA USA

## ARTICLE INFO

### Article history:

Received 26 August 2015

## ABSTRACT

**Objective:** Review the importance of imaging selection and clinicoanatomic correlation for a vascular malformations presenting with unique symptomatology.

**Methods:** Case study and literature review.

**Results:** A 64-year-old female presented with globus and dysphagia ongoing for 40 years. Esophagogastroduodenoscopy discovered a hypopharyngeal mass. A CT scan showed a soft tissue mass with shotty calcifications. Flexible laryngoscopy revealed a bluish compressible mass. MRI showed T2 hyperintensity with heterogeneous enhancement resulting in the diagnosis of a low-flow vascular malformation.

**Conclusions:** All globus is not equal. Attention to symptoms, anatomy, and imaging selection is crucial for the diagnosis and treatment of vascular malformations uniquely presenting with dysphagia.

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## 1. Introduction

Vascular anomalies, including vasoproliferative/vascular neoplasms and vascular malformations remain a diagnostic and therapeutic challenge. Unlike vascular neoplasms, vascular malformations are composed of abnormally formed vascular channels lined by endothelium that do not exhibit abnormal cellular turnover or mitosis. Vascular malformations are congenital lesions that never regress and grow proportionally with time, trauma, and/or hormonal changes. Vascular malformations may be of the low-flow variety, including capillary malformations (CM), venous malformations (VM), and lymphatic malformations (LM), or of the high-flow variety, including arteriovenous malformations (AVM). In the head and neck, venous malformations (VM) are the third most common vascular

mass and may be seen within the muscles of mastication, lips, tongue, or elsewhere within the upper aerodigestive tract. Imaging is critical in the diagnosis and management of vascular malformations. See [Table 1](#) for the importance of imaging selection for each vascular malformation subtype [1].

Here we report on a patient with chronic dysphagia found to harbor a large hypopharyngeal low-flow vascular malformation. We highlight the importance of imaging selection and the dictums of long-term management.

## 2. Case report

A 65-year-old retired teacher presented with a history of globus and dysphagia since she was twenty years old. She has always had problems with swallowing pills and intermittently

\* Corresponding author at: 10833 Le Conte Avenue, 62-132, Los Angeles, CA 90095, USA. Tel.: +1 310 206 6688.

E-mail addresses: [AVahabzadehHagh@mednet.ucla.edu](mailto:AVahabzadehHagh@mednet.ucla.edu) (A.M. Vahabzadeh-Hagh), [ASepahdari@mednet.ucla.edu](mailto:ASepahdari@mednet.ucla.edu) (A.R. Sepahdari), [JFitter@mednet.ucla.edu](mailto:JFitter@mednet.ucla.edu) (J. Fitter), [Abemayor@ucla.edu](mailto:Abemayor@ucla.edu) (E. Abemayor).

**Table 1 – Imaging of vascular malformations.**

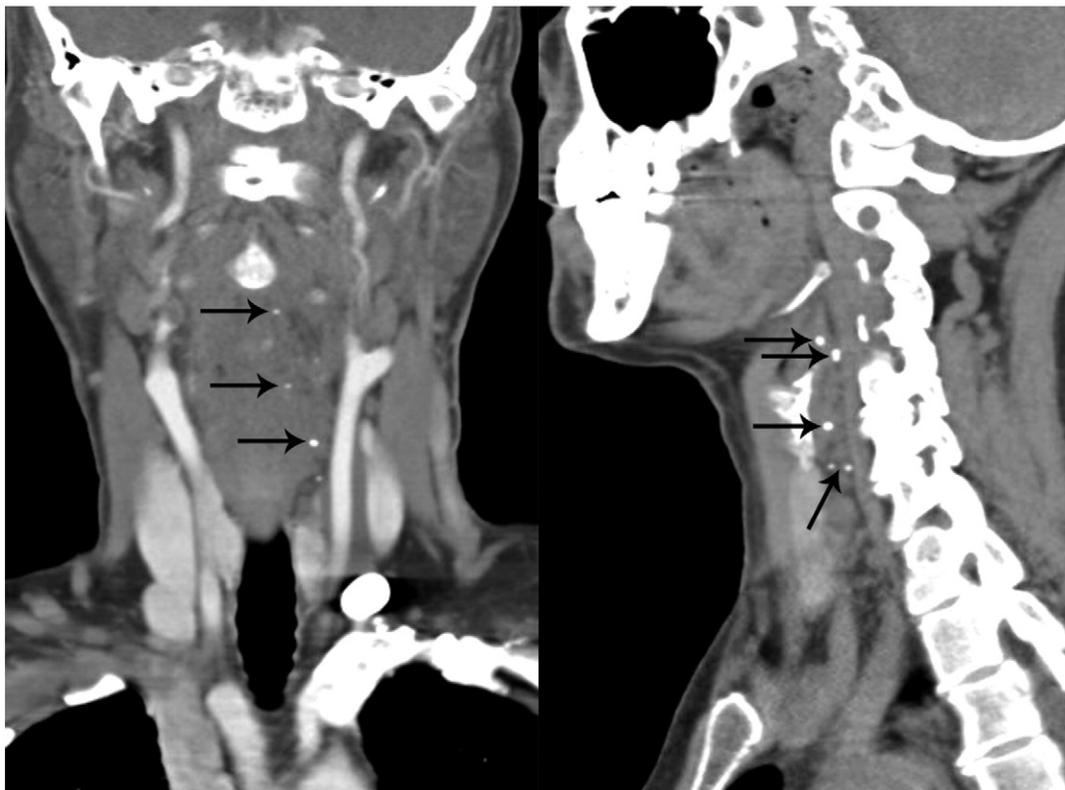
Malformation	Importance of imaging
Capillary	Imaging usually not needed unless V1 (ophthalmic division of trigeminal nerve) or midline distribution to rule out leptomeningeal involvement
Venous	MRI most useful, small lesions may need formal angiogram to fully characterize
Lymphatic	Ultrasound/color flow Doppler; helps to identify characteristic cystic appearance; MRI is useful to evaluate extent of disease
Arteriovenous	Ultrasound and MRI to identify high flow pattern as well as lesion extent. CT can be helpful especially for bony AVMs and angiography can assist in identifying feeding and draining vessels prior to therapeutic intervention

finds difficulty with solid foods. She otherwise has a history of GERD for which she takes a proton pump inhibitor and antacids. Her social history includes 1 glass of wine daily and an 8 pack-year smoking history, a habit she quit approximately 30 years ago. Extensive work-up at an outside facility including esophagogastroduodenoscopy led to the discovery of a hypopharyngeal mass. A CT scan revealed 3 × 1.5 cm soft tissue mass in posterior midline hypopharynx with shotty calcifications, and small bilateral cervical lymph nodes. The initial radiological interpretation raised concern for malignancy (Fig. 1).

Flexible fiberoptic laryngoscopy revealed a bluish hued lesion at posterior hypopharynx, at the level of the vocal cords. This lesion appeared partially compressible with swallow and hyolaryngeal elevation (Fig. 2). An MRI was obtained which demonstrated T2 signal hyperintensity as well as heterogeneous contrast enhancement suggestive of a low-flow or mixed vascular malformation (Figs. 3–6). This further demonstrated the trans-spatial nature of these lesions, which tend to traverse fascial planes as opposed to causing local distortion and mass effect as can be seen with malignancies. Further review of the CT scan at our institution recognized the calcifications as phleboliths, a pathognomonic feature of venous malformations. The combination of our examination and these imaging features solidified the diagnosis of a hypopharyngeal venous malformation.

### 3. Discussion

VMs are the most common type of vascular malformation with an incidence of 1–4%. VMs tend to grow proportionally with age and often increase in size with hormonal changes, such as in puberty. They tend to undergo thrombosis resulting in the pathognomonic radiologic signs of phleboliths, intralesional calcifications. Ultrasound is a good imaging modality if the lesion is accessible, but MRI tends to be the most useful modality to characterize the full extent of disease as they often are intimately associated with adjacent musculature and



**Fig. 1 – Coronal and sagittal contrast-enhanced CT images — multiple, small, round calcifications in the hypopharynx and left carotid sheath, consistent with phleboliths and strongly suggestive of a low-flow venous malformation.**



**Fig. 2 – View of hypopharyngeal mass via flexible fiberoptic laryngoscopy — bluish hued lesion at posterior hypopharynx at the level of the vocal cords. Otherwise normal appearing mobile bilateral vocal cords; images left to right show vocal cords adducting. AE — aryepiglottic fold, Ant — anterior, L — left, Post — posterior, Py — pyriform sinus, R — right, VM — venous malformation, # — false vocal fold, ★ — true vocal fold.**

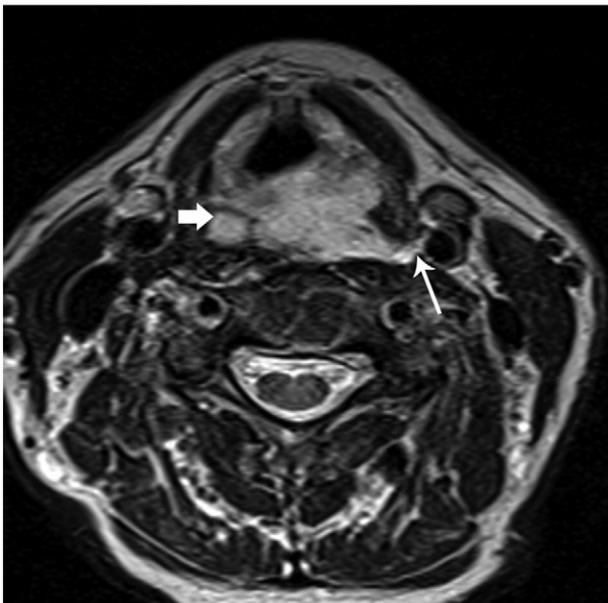
may be multifocal. This case report highlights three key imaging features of venous malformations. Namely, phleboliths seen on CT, hyperintense T2 signal on MRI, and the trans-spatial behavior of these lesions in the absence of local tissue distortion or mass effect.

Phleboliths have been described in literature reviews and case reports of venous malformations and hemangiomas. However there remains an ambiguity between what is labeled a vascular malformation versus a vascular neoplasm in these reports [2–5]. As Mulliken and Glowacki described in 1982, the differentiation of vascular neoplasms from vascular malformations relies on the histopathologic demonstration of increased endothelial cell turnover [6]. With an incomplete clinical picture and absence of histopathology it then becomes difficult to label these lesions definitively. As such, despite the presence of such reports, we continue to support phleboliths as a pathognomonic radiologic sign of venous malformations.

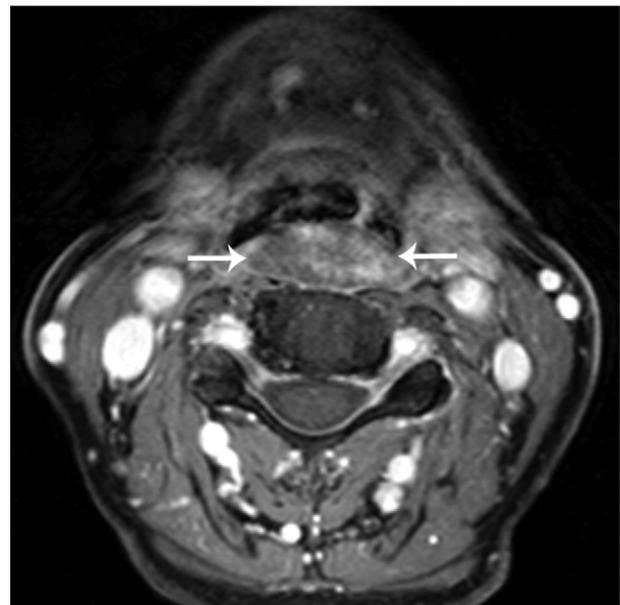
Although the imaging characteristics of this lesion were classic for venous or mixed low-flow vascular malformations, the clinical

presentation was not. This difference may be attributed to the unique anatomic location of this lesion. VMs typically are localized lesions on the face, limbs, or trunk [1]. Leung et al reviewed 40 children with low-flow vascular malformations and found the majority of patients presented with a visible mass or swelling, pain with bleeding, discoloration of skin, or obstructive airway symptoms, with only one case having difficulty feeding [7]. Here we find a submucosal venous malformation within the hypopharynx, a unique location explaining this rare symptomatic presentation and likely contributing to the prolonged pathway to a final diagnosis.

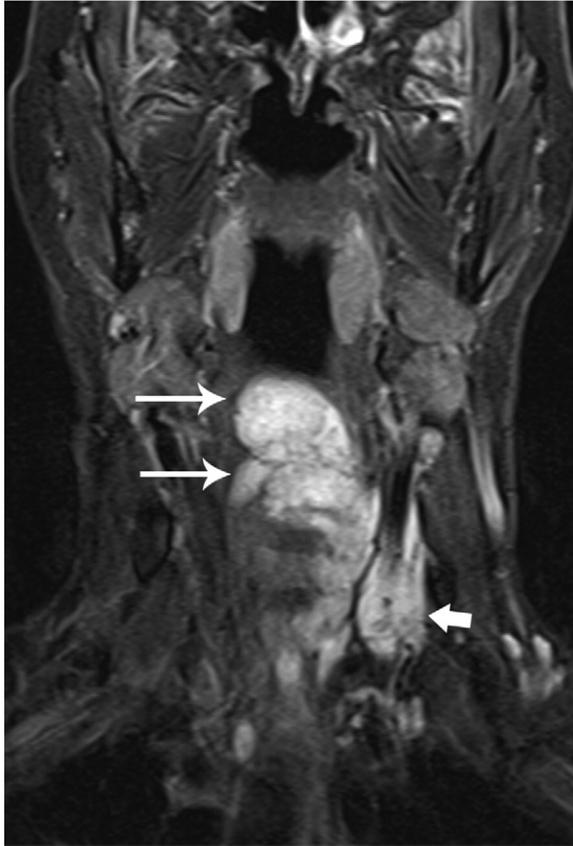
The natural history of venous malformations includes lesion expansion, development of thrombi, and eventual alteration in function and/or esthetics. Treatment is expectant in nature. Therapeutic options are based on three pillars; namely, surgery, sclerotherapy, and laser treatment. Surgery is usually a second-line treatment option or is implemented in conjunction with sclerotherapy. Laser, specifically Nd:YAG is well suited to treat mucosal lesions of the upper aerodigestive



**Fig. 3 – Axial T2-weighted image through the level of the hypopharynx — markedly T2-hyperintense lesion (short arrow) that smoothly effaces the airway, and also extends toward the left carotid sheath (long arrow). Markedly bright T2 signal indicates a non-squamous cell lesion.**



**Fig. 4 – Axial fat-suppressed post-contrast T1 through the level of the hypopharynx — minimal enhancement of this component of the lesion (arrows), suggests the presence of a lymphatic component in addition to the venous component.**



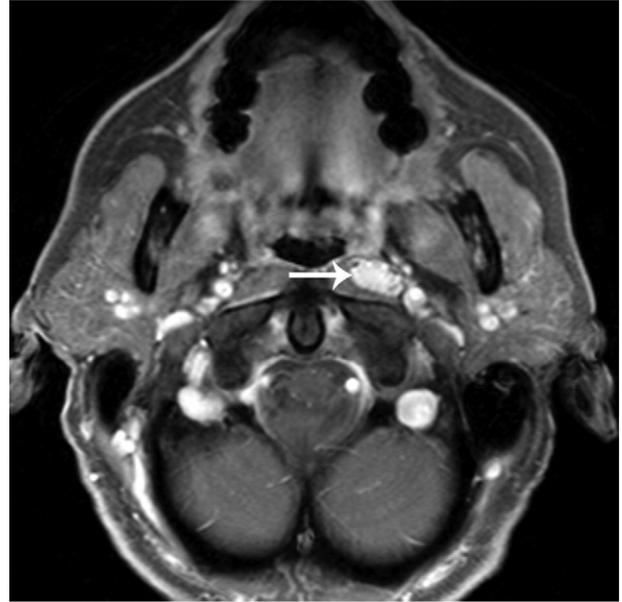
**Fig. 5 – Coronal inversion recovery image — hyperintense signal in the hypopharynx (long arrows), and also infiltrating about the left carotid sheath (short arrow).**

tract [8]. Goals of therapy tend to revolve around controlling the disease with periodic interventions without harming preexisting form or function. As such, long-term follow-up is key in these patients [7,9].

The patient in this case maintained relatively good function without any truly debilitating symptoms. We discussed the options of laser treatment, robotic resection, external beam radiotherapy, or sclerotherapy but recommended watchful waiting with interval imaging to assess its growth pattern given that therapeutic intervention could potentially cause a functional setback.

#### 4. Conclusion

Venous malformations are common vascular malformations found in the head and neck. Symptomatology is mainly a function of their size and location. In our case we see the rare presentation of dysphagia caused by a submucosal VM within the hypopharynx. Imaging is critical in avoiding a more invasive diagnostics pathway and, although multiple



**Fig. 6 – Axial fat-suppressed post-contrast T1 at the level of the skull base — enhancement within the prevertebral muscles on the left (arrow), reflecting extension of the vascular malformation high within the prevertebral muscles.**

treatment modalities exist, management is often observational and expectant in nature.

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