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A Rare Case of Low Flow Vascular Malformation of Head and Neck Region Presenting with Multiple Phleboliths

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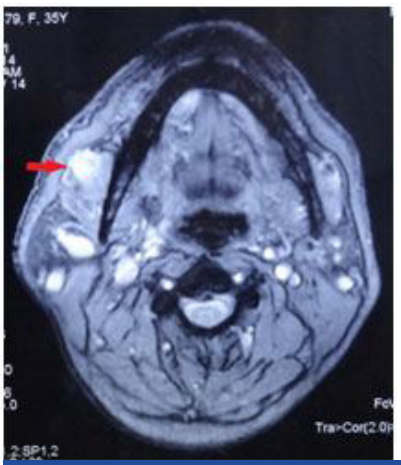
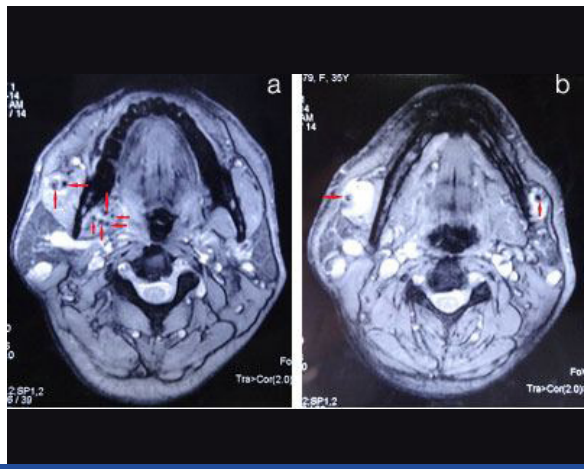
A 37-year-old male patient reported to outpatient department with chief complaints of swelling on the tongue for past five years. Patient also noticed a new swelling on the left side of the neck which appeared two months back which used to increase in size on lying down. There was no previous history of trauma and any other systemic illness. Family history was non-contributory. On extraoral examination, a swelling was noted on left side of neck approximately 2×3 cm in size, bluish purple discoloration, non-pulsatile, soft, compressible & non-tender [Table/Fig-1]. Intraoral examination revealed two well defined, soft, compressible, non-pulsatile, non-tender swelling with slight bluish hue measuring approximately 1×1 cm and 0.5×0.5 cm on right and left dorsum of tongue respectively [Table/Fig-2]. Diffuse reddish blue macular lesion was present on left ventro-lateral surface of tongue posteriorly [Table/Fig-3]. No difficulty in swallowing & speech was present. Provisional diagnosis of vascular malformation was made. Differential diagnosis of haemangioma, arterio-venous malformation was given Magnetic resonance imaging (MRI) neck was done. Right masseter muscle showed heterogenous postcontrast enhancement and enhancing focal lesions of altered signal intensity on T2-weighted images [Table/Fig-4]. Similar lesions were noted in right medial pterygoid muscle, left masseter muscle and along right side of tongue on T2-weighted images [Table/Fig-5]. Areas of signal voids were noted in lesions on FLASH MRI (Fast Low Angle Shot Magnetic Resonance Imaging) which were suggestive of phleboliths in bilateral masseter muscle and right lateral pterygoid muscle on T2-weighted images [Table/Fig-6a&b]. Heterogenous postcontrast enhancement of the right parotid gland was also noted on T2-weighted images [Table/Fig-7]. MR Angiography neck revealed bilateral common carotid arteries, external carotid arteries, internal carotid arteries with normal caliber and enhancement. No evidence of arterial feeders or early draining vein was seen [Table/Fig-8]. Above mentioned MRI findings were suggestive of slow flow vascular malformations likely venous malformation. Excisional biopsy was done of the lesion on dorsum

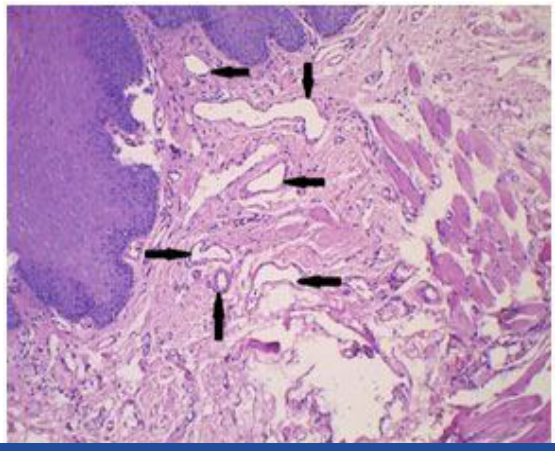
[Table/Fig-1]: Lesion on left side of the neck shown by an arrow **[Table/Fig-2]:** Lesion on the dorsum of tongue shown by the arrows **[Table/Fig-3]:** Lesion on left ventro-lateral surface of tongue shown by an arrow





[Table/Fig-4]: T2 weighted MRI image showing enhancing lesions in right masseter depicted by an arrow **[Table/Fig-5]:** T2 weighted MRI image showing enhancing lesions in right medial pterygoid, left masseter, and right side of tongue depicted by the arrows **[Table/Fig-6a,b]:** FLASH MRI showing signal voids suggestive of phleboliths depicted by the arrows



[Table/Fig-7]: MRI showing enhancement of right parotid gland depicted by an arrow **[Table/Fig-8]:** MRI angiography showing normal enhancement and caliber of the vessels **[Table/Fig-9]:** Microphotograph at 10X power showing numerous variably sized and shaped endothelial lined blood spaces in stromal tissue depicted by the arrows

	Gray scale US	Doppler US	MRI
Slow flow vascular malformations			
• Venous	Solid echogenic lesion with phleboliths	Monophasic venous or no flow pattern	-Heterogenous, intermediate signal on T1W -Enhancement on T1W post
• lymphatic	Variable multicystic lesions with or without fluid	Lesion with no flow except in septa	-Low-intermediate signal on T1W -High signal on T2W -No enhancement (except in septa) on T1W post gadolinium
Fast flow vascular malformations			
• Arteriovenous malformations and fistulas	Well defined, cluster of vessels	Arterial and venous signals with arterializations of venous structures	Serpiginous signal voids on T1W and T2W

[Table/Fig-10]: MRI and US features of vascular malformations

of tongue & histopathological sections revealed stromal tissue showing numerous variably sized and shaped endothelium lined blood spaces supported by dense fibrous connective tissue [Table/Fig-9]. These features were suggestive of vascular malformation. Hence, final diagnosis of low flow venous malformations was made. Since there was no difficulty in speech and swallowing was associated & also patient was also not willing for management so no treatment was provided & patient is under regular follow up for the past one year.

Vascular malformation is proposed to be the consequence of congenital defect of vascular morphogenesis. Different varieties of vascular malformation pose some of the most difficult challenges from diagnosis as well as therapeutic point of outlook. Imaging modalities including Plain Film Radiography, Ultrasound, Color Doppler Imaging, Computed Tomography (CT), Magnetic Resonance Imaging (MRI), and MR angiography have prompt role in diagnosis and providing pre-treatment information for the workup of such cases. Ultrasound (US) is often used and it may define the extent of superficial lesions. Color Doppler imaging (CDI) has proven to be an excellent diagnostic tool in the initial evaluation and later follow up of the cases since it is non-invasive, cost-effective, and provides real time images. CDI generally shows arterial or venous nature of the flow, as well as its velocity [1].

Currently MRI has been established as a mainstay for the initial diagnosis, to access endovascular treatment as well as follow up. High flow vascular malformations such as arterio-venous malformation demonstrates usually a signal void on long TR/TE sequences which can be predominantly due to turbulence related de-phasing contributing to signal loss. Whereas low flow vascular malformations such as venous malformations (VM) and lymphatic malformations in general have high signal intensity on long TR/TE sequences which is attributed to stagnant flow in these abnormal vascular spaces [2]. Presence of septations and rounded signal voids on MRI corresponds to existence of phleboliths and are additional distinguishing feature of VM [3]. The characteristic MRI and US features of vascular malformations are summarized in [Table/Fig-10]. High spatial resolution of MR angiography allows for highly accurate measurement and mapping of feeding and draining vessels and association with adjacent structures to assist in interventional radiologic or surgical planning. Treatment options include conservative measures such as head of bed elevation and compression, laser therapy, sclerotherapy, and surgery. Comprehensive multidisciplinary approach should be adopted to achieve most effective results. Complications in head and neck area include significant cosmetic defects, recurrent bleeding, obstruction of airway and interference with normal speech and dentition.

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