

# An Infiltrative Angioarchitectural Variant of Arteriovenous Malformation of Temporalis

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## ABSTRACT

Vascular anomalies of the head and neck region pose a certain diagnostic and therapeutic paradox. Management of arteriovenous malformations (AVM) is a challenge owing to the presence of abnormal vascular communications and high recurrence. We report a case of a 19-year-old male patient, who presented with diffuse swelling in the right temporal region. Magnetic Resonance Angiography (MRA) suggested it to be an AVM in the temporalis muscle, having afferents in the ascending pharyngeal artery, with cavernous angioma. Surgical excision of the lesion was carried out under carotid control. Histopathology of the excised specimen utilizing special stains confirmed the presence of AVM. An absence of distinct nidus concomitant along with the exuberant proliferation of capillaries between the muscle fibres suggested it be an infiltrative angioarchitectural variant. The present case highlights significance of diagnosing AVM in temporalis muscle which is a rare occurrence in head and neck region. Also, the importance of ruling out other closely resembling vascular diathesis, both non neoplastic and malignant is discussed.

**Keywords:** Arteriovenous malformations, Elastic stains, Magnetic resonance angiography

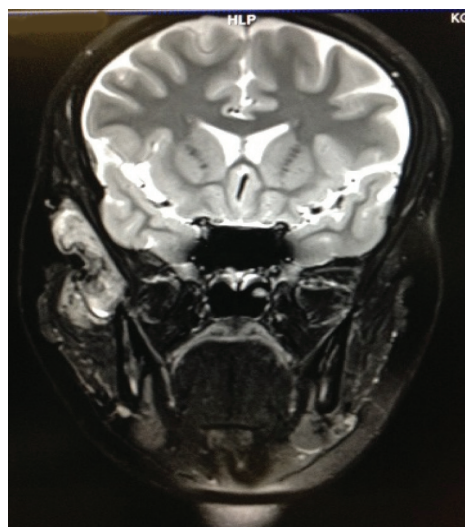
## CASE REPORT

A 19-year-old male reported with a complaint of swelling on the right upper part of the face. The patient noticed it about six months back, which gradually increased in size causing change in his facial profile. There was no associated pain or discomfort, weakness of the facial muscles, abnormal temporomandibular joint movements, and history of headache or trauma. Also, the patient did not provide any previous medical or any drug history for the same. Examination of the swelling revealed a diffuse enlargement over the right temporal region measuring about 5x4 cm, with no noticeable change in the colour of the skin. On palpation, the swelling was soft, non-compressible, non-tender, distinctly warm and pulsatile. Bruit could be appreciated by auscultation. A provisional diagnosis of vascular malformation was considered.

The magnetic resonance image (MRI) revealed a T1W hypointense and T2W mixed intensity lesion involving right temporalis and

masseter muscles extending from temporal fossa to the infra temporal fossa with flow voids measuring 6.37x4.40x2.81 cms [Table/Fig-1a]. The neck angiogram showed an evidence of intralesional flow voids supplied by the right ascending pharyngeal branch of external carotid artery and drained by external jugular vein, with cystic spaces [Table/Fig-1b]. A diagnosis of combined AVM with cavernous angioma of the right temporalis and masseter was made.

Complete surgical excision under carotid control via preauricular approach was carried out under general anaesthesia. The entire lesion was found to be located inside the temporalis muscle [Table/Fig-2]. Elective access osteotomy of the zygomatic arch was done in order to excise the lesion. The surgically excised specimen on gross examination measured about 4x2x1.5cm in dimension, creamish in colour, with an irregular surface [Table/Fig-3], and firm in consistency with the cut surface showing a few pigmented areas.



**[Table/Fig-1A]:** T2 weighted magnetic resonance image (coronal view) showing the extent of the lesion involving right temporalis and masseter muscle



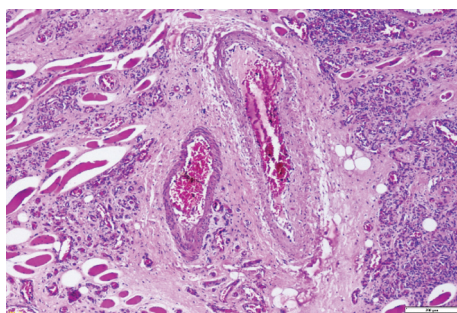
**[Table/Fig-1B]:** Magnetic resonance angiograph showing flow voids supplied by right ascending pharyngeal branch of external carotid artery and drained by external jugular vein, with cystic spaces



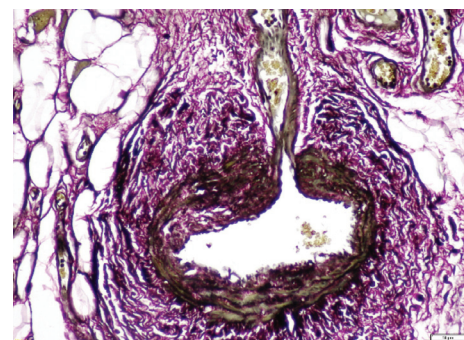
**[Table/Fig-2]:** Surgical photograph depicting lesion located in the temporalis muscle



**[Table/Fig-3]:** Gross examination of the excised specimen, measuring 4x2x1.5 cm in dimension



**[Table/Fig-4A]:** Photomicrograph showing hypertrophy of an artery and a vein with proliferating endothelial cells infiltrating between muscle tissue (Haematoxylin and Eosin, 4x)



**[Table/Fig-4B]:** Photomicrograph showing direct communication between artery and vein (Verhoff-van Gieson stain, 10x)

The haematoxylin and eosin (H&E) stained sections showed closely aggregated thin and thick walled vessels, arteries juxtaposed to thick-walled veins, proliferating capillaries amidst the fascicles of skeletal muscles and deposits of adipose tissue confirming the diagnosis of an AVM [Table/Fig-4a]. Staining of tissue with Verhoff – Van Gieson haematoxylin highlighted the internal elastic membrane of the arteriovenous shunts in the tissue section as thickened blue black line [Table/Fig-4b].

Patient is currently on regular follow up with no complications or any signs of recurrence which is confirmed by conducting MRA every six months.

## DISCUSSION

Vascular anomalies represent a heterogeneous group of vessel disorders which are diagnostically challenging and therapeutically enigmatic. Arteriovenous malformations (AVMs) are the congenital vascular malformations consisting of the arterial system shunting blood directly into a tortuous, dilated, usually multiple outflow veins through a vascular nidus without the usual resistance of an intervening capillary bed. A phase of early quiescence, followed by the gradual infiltration of soft tissue and bone, a late expansion, with the eventual destruction is the usual course of AVM [1]. Head and neck AVMs are stated to occur in 0.1% of the population. Extracranial AVMs account for only 8.1% of head and neck AVMs.

The arterio-venous shunt in AVM is thought to be due to failure of primitive arteriovenous channels to regress in utero. Trauma is considered to be one of the chief causes of AVM, including iatrogenic, as the one reported by Brown et al., [2] wherein, AVM was caused due to radical neck dissection. However, in the present case, no such history was noted.

AVMs grow in proportion with the growth of an individual and can exacerbate with trauma or hormonal change [1], particularly during puberty and pregnancy. Research has indicated that the dysregulation in apoptotic pathway, due to defect in transforming growth factor (TGF)-  $\beta$  signalling, leads to abnormal vascular proliferation in AVM. Also, increased expression of progesterone receptors in AVM clarifies their expansion during puberty. AVMs are usually sporadic, but mutation in gene RAS p21 protein activator 1 (RAS A1), expressing p120- Ras GTPase activating protein (Ras GAP), on chromosome 5q has been identified in families with AVM [3].

More than 50% of AVMs are located in head and neck region, common sites being midface and oral cavity. Occurrence of AVM in oral cavity, especially the base of tongue is taxing as it causes difficulty in reconstruction following surgical intervention. Maxillary and ethmoidal sinuses [4], mandible, nasopharynx are among the other sites where AVM has been reported to occur.

The histopathology shows considerable variation, suggesting a spectrum of disease and not just one disease process. Vascular recruitment and collateralization contribute to progressive expansion

of AVMs with the acquisition of feeding arteries and draining veins. AVMs when present intramuscularly show fatty overgrowth as a result of skeletal muscle atrophy and fatty replacement, as was observed in the present case. One of the chief features supporting the diagnosis of an AVM is the presence of an artery juxtaposed to a thick-walled vein with a larger lumen. The venous structures found in AVMs seem to strike a resemblance to an artery as they show medial hypertrophy and fibrointimal hyperplasia. Elastic stains are used as ancillary tools for making a definitive diagnosis of AVM [5]. Verhoff – Van Gieson haematoxylin staining in our case illustrated the thick elastic lamina of the arteries as well as veins with intercommunications, and clusters of exuberantly proliferating capillaries were evident adjacent to the skeletal muscle tissue with no significant feeding arteries.

Differentiating AVM from other closely related vascular anomalies is imperative as they differ in mode of treatment and prognosis. Intramuscular cavernous haemangioma (ICH) mimics AVM as both are slow growing masses with little or no superficial skin discoloration. However, ICH was differentiated from AVM by presence of large dilated blood filled vessels lined by flattened endothelium having a roughly lobular arrangement and absence of arteriovenous shunts.

Venous haemangioma was ruled out as it is slow flow lesion and histologically comprises of large veins with irregularly attenuated or disorganised walls, resembling the spokes of a wheel, with no feeding arteries, along with presence of intraluminal thrombi and phleboliths [6].

Presence of proliferating capillaries, as seen in present case, does resemble capillary haemangioma. However, capillary haemangiomas become clinically evident in an early stage in life and do not grow proportionally with growth of the patient. As they involute with age, capillary haemangiomas may show dilated residual vessels giving an impression of AVM. However, they lack the arteriovenous intercommunications seen in AVMs.

Presence of capillaries proliferating in between the muscle tissue may mimic angiosarcoma. However, absence of nuclear atypia and freely anastomosing sinusoidal pattern of capillary proliferation encountered in most angiosarcoma sets it apart from AVM [6].

Mixed vascular malformations including definite features of more than one pathologically discrete type of malformation within the same lesion have been reported [7], giving rise to several hypotheses about common pathogenesis among different lesions. The manifestation of proliferating capillaries in our case suggested the possibility of it being a hybrid lesion having features of more than one vascular malformation.

Physicians and surgeons must be aware of complications of AVM as they can be life-threatening, depending on the site of occurrence and the vessel involved. Uncontrollable epistaxis causing death of the patient due to AVM of geniomasstetric muscle has been reported by Cotulbea et al., [8]. AVM in tongue can not only cause haemorrhage and airway obstruction, but also difficulty in chewing and swallowing,



speech problems as well as orthodontic abnormalities [9]. Visual defects can also be one of the complications of AVM in head and neck region. Skeletal changes due to AVM include destructive and intraosseous changes, sometimes hypoplasia and demineralization. One of the most bothering complications of AVM to be reported is squamous cell carcinoma [10], thus requiring timely treatment.

Their hemodynamic characteristics, modality of growth and tendency to recur impose a therapeutic challenge for the practitioners. Angiography is preferred not only for diagnostic purposes but also as an initial therapeutic step in the form of embolization, as it helps reduction in vascularity and shrinkage of the lesion, making surgical ablation less extensive. Complete excision of the AVM (confirmed by pre- or postoperative angiography) is the most effective method of treatment, which was done in present case. Close postoperative observation with management of expected local recurrence is obligatory. Cyanoacrylate has been successfully used as the only mode of treatment for AVM of mandible in a 4-year-old child. A multidisciplinary team approach is required in the assessment and treatment of these lesions. Recent advances in microsurgery and in therapeutic radiology have greatly improved the prognosis for patients with these malformations.

## CONCLUSION

The characteristic clinical and histological presentation supported by relevant imaging findings in the present case confirmed AVM as

our diagnosis. AVM demands careful planned investigations and mode of treatment. Complications and clinical manifestations of AVM must be kept in mind by the physicians and surgeons, as it can prove to be fatal at times.

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