VOLUME 9 | ISSUE 2

CLINICAL CASE - TEST YOURSELF
Head and neck imaging

A 46-year-old female with swelling in right cheek

Karthikeya Patil, Meera Theenathayalan, Eswari Solayappan
Department of Oral Medicine and Radiology, JSS Dental College and Hospital, JSS Academy of Higher Education and Research, Mysuru

SUBMISSION: 23/11/2023 | ACCEPTANCE: 27/01/2023

PART A

A 46-year-old female patient presented with swelling in her right cheek region [Fig 1a,1b] for two years, accompanied by intermittent pain and enlargement of a mass when lying down [Fig 1c]. The patient gives no history of trauma. There is no history of sudden increase or decrease in the size of the swelling. The swelling was visible when the patient clenched her teeth and while sleeping [Fig 1c]. Blood investigations were within normal limits. Digital Panoramic radiograph [Fig 2] was taken followed by Cone Beam Computed Tomography (CBCT) [Fig 3a,3b,3c], Ultrasonography (USG) [Fig 4a,4b,4c] was performed.

CORRESPONDING AUTHOR, GUARANTOR

DR Karthikeya Patil, MDS, Professor and Head,
Department of Oral Medicine and Radiology,
JSS Dental College and Hospital,
JSS Academy of Higher Education And Research,
Mysore - 570 015
Email: dr.karthikeyapatil@jssuni.edu.in
Phone +91 94498 22498.
ORCID ID: 0000-0002-7941-2467

VOLUME 9 | ISSUE 2

Figure 1a *- Frontal Profile View of the patient,* ***Figure 1b*** *- Right Lateral View of the patient,* ***Figure 1c*** *- Supine Position of the patient*

Figure 2 *– Orthopantomogram*

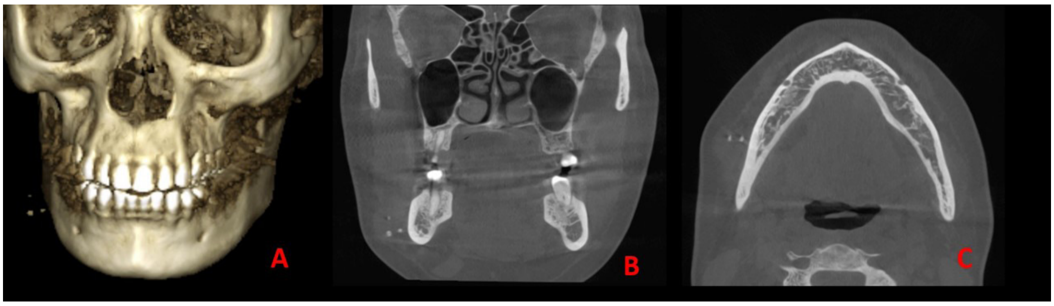

Figure 3a *– Cone Beam Computed Tomography section of 3D reconstruction,* ***Figure 3b****- Cone Beam Computed Tomography section of Coronal Section,* ***Figure 3c****- Cone Beam Computed Tomography section of Axial section*

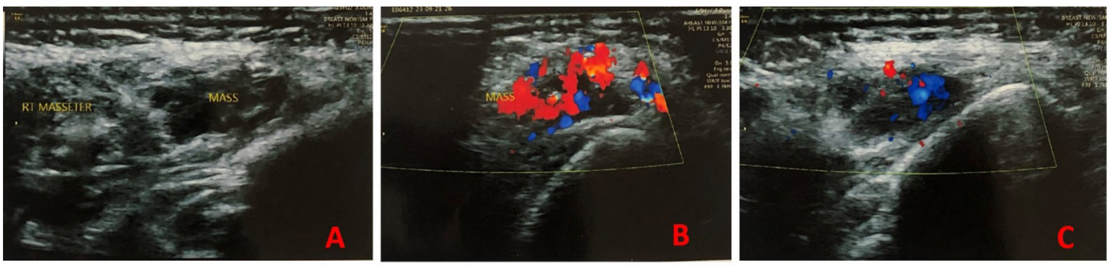

Figure 4a, 4b, 4c *– Ultrasonography of the right masseter region*

PART B

DIAGNOSIS: Intramuscular Arteriovenous Malformation in Right Masseter Muscle with Turkey Wattle Sign

Diagnostic imaging was necessary to determine the cause and extent of the abnormalities. The orthopantomogram findings of the patient were insignificant. Further examination with Cone Bean Computed Tomography [CBCT] revealed multiple bony nodules involving the masseter region. Three small nodules were noted, each measuring roughly about 2x2mm in size. For further evaluation, Ultrasonography [USG] was taken, which revealed diffuse swelling in the inferior border of the right masseter muscle, measuring roughly about 17X9mm, and had clusters of venous channels noted with areas of calcification suggestive of Phleboliths. The impression was an arterial malformation of the right masseter muscle.

Arteriovenous malformations (AVM) is a medical condition that occurs when an artery and vein connect directly without capillaries. These malformations are typically detected at birth or in early childhood and are often located inside the skull. However, there is another type of AVM called extracranial AVM, which is a more common local aggressive disorder primarily observed during puberty or adolescence [1]. This type of malformation can cause cosmetic and functional disturbances, such as facial asymmetry. AVM is caused by defects in TGF-beta signaling and a genetic two-hit hypothesis. The development of the mesenchyme primordia is stopped during the undifferentiated capillary network stage, leading to vascular malformation. As differentiation occurs, primitive vessels penetrate deeper into the subcutaneous layer, muscle, or bone tissue. Gradual replacement of the immature plexiform network by mature vascular channels results in vascular malformation. This type of vascular tumor is difficult to diagnose due to its cellular nature and firmness. The tumors show more mitotic figures, perineural infiltration, and lymphoid follicles than cavernous types, but less than capillary types. They are only found in 5% of the head and neck region [2].

AVM, or Arteriovenous Malformation, can be triggered by various factors such as trauma, hormonal changes, ischemic events, and puberty. Skeletal muscle vascular tumors are rare, but can occur in the head and neck region, with the masseter muscle being the most commonly affected (accounting for 4.9% of all intramuscular malformations), followed by the sternocleidomastoid and trapezius muscles [3]. While some people may be born with AVM, it may not become apparent until childhood. It is most frequently found in the head and neck, followed by the limbs, trunk, and viscera. It can also occur on the anterior two-thirds of the tongue, palate, gingival, and buccal mucosa in the oral cavity. AVMs can be classified using the Schobinger clinical staging system. Initially, the lesions are latent but develop into a warm pink-bluish skin lesion with a pulsatile thrill in adolescence [4]. Over time, the lesion will expand, leading to dystrophic skin changes, bleeding, ulceration, and tissue necrosis. While the causes of these tumors are still unknown, some evidence suggests that the trophoblast, a placental cell, may be the origin of vascular malformations [5]. Intramuscular vascular malformations are difficult to detect through physical examination as they are usually deep within the muscle. The condition is characterized by swelling, pain, and mobile, tense, and pulsatile lesions along the muscle's vertical axis [6].

Unlike vascular lesions on the skin, these malformations rarely cause skin discoloration and do not grow over time. When they occur in the masseter muscle, they can cause swelling, pain, or facial paralysis. The expansion of the mass can also cause pressure on the nerve, and the lesion becomes more prominent when the Valsalva maneuver or muscle contraction is performed [7]. Tumors can often be mistaken for intramuscular vascular malformations due to their proximity, and they must be distinguished from parotid gland neoplasms. Increased venous pressure can make the tumor more visible. Arteriography is necessary for the detection of vessel communication, while sialography has been replaced by computerized tomography with contrast enhancement, which makes it easier to differentiate between parotid and extra parotid masses [8,9]. Ultrasound is used to diagnose lesions, while computed tomography (CT) or MRI is better for determining the extent of the lesions.

Imaging is crucial in characterizing arteriovenous malformations (AVMs) and planning treatment. Investigations such as Doppler ultrasonography, CT,

VOLUME 9 | ISSUE 2

Figure 1a *- Frontal Profile View shows solitary swelling in the right body of the mandible,* ***Figure 1b*** *- Right Lateral View shows a solitary swelling measuring approximately 1X1.5cm in size was found 1cm anterior to the right angle of the mandible.,* ***Figure 1c*** *- Supine Position – Depicting swelling in the Right inferior border of the mandible Region which was visible and palpable when the patient was clenching or sleeping, which is referred to as the "turkey wattle sign"*

Figure 2 *– Orthopantomogram showing no pathological variations arising from the odontogenic origin.*

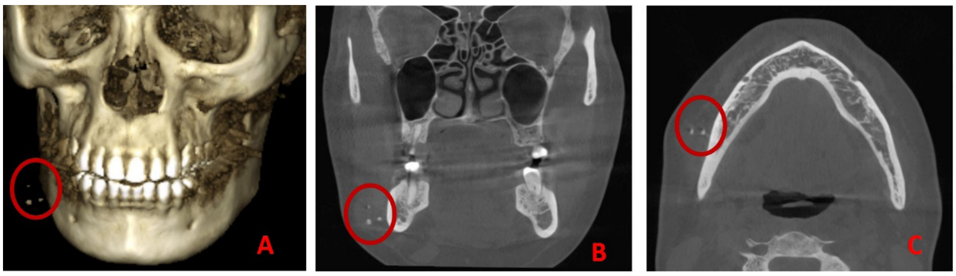

Figure 3a *– Cone Beam Computed Tomography section of 3D reconstruction revealed multiple bony nodules involving the right masseter region,* ***Figure 3b****- Cone Beam Computed Tomography section of the coronal Section shows 3 specks of calcification in the right body of the mandible region.,* ***Figure 3c****- Cone Beam Computed Tomography section of axial section representing 2 specks of calcification in the right body of the mandible region.*

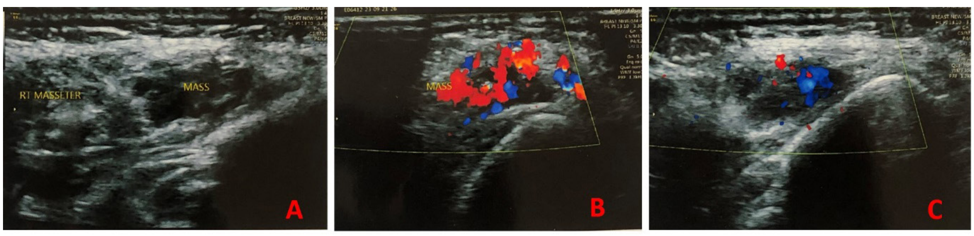

Figure 4a, 4b, 4c *– Ultrasonography of the right masseter region reveals clusters of venous channels noted within the mass measuring 17X9mm with specks of calcification.*

and MRI can diagnose arteriovenous shunts. Ultrasound and Doppler imaging can reveal high systolic and diastolic flow, arteriovenous shunting, and arterial waveforms. MRI shows AVMs as a mesh of dilated arteries and veins connected by shunts. Magnetic resonance angiography (MRA) shows arterial feeders, shunting volume, and nidus location. Contrast-enhanced CT can provide an accurate structural assessment of arteries, veins, and nidus and flow analysis for preprocedural planning [8,10].

When dealing with vascular malformations, doctors must examine their flow characteristics to determine the appropriate treatment. Low-flow malformations can be treated with sclerotherapy, laser therapy, or cryosurgery, while high-flow malformations require presurgical embolization followed by aggressive ablative therapy. The ideal time between embolization and surgical treatment varies based on the goals of embolotherapy. After 2-3 weeks, the benefits of embolization may decrease due to collateral supply and recanalization of vessels. There are several treatment options available, including medical therapy, embolotherapy, sclerotherapy, lasers, cryosurgery, and surgical therapy. The timing of treatment depends on the goals of embolotherapy [4,7].

In conclusion, diagnosis of intramuscular AVMs is challenging and requires a multidisciplinary approach including plastic surgery, vascular surgery, and interventional radiology. Endovascular embolization is an effective therapeutic choice in this instance, offering a less intrusive way to obstruct abnormal arteries and lessen symptoms. To keep an eye out for recurrence and evaluate the effectiveness of the intervention, long-term follow-up is essential. Furthermore, to improve early diagnosis and patient outcomes, healthcare providers must be able to acknowledge the clinical presentations of intramuscular AVMs, particularly the "turkey wattle" sign. This rare case report adds to the knowledge of intramuscular AVMs by highlighting the significance of a thorough diagnosis process and management, when dealing with these unusual vascular diseases. **R**

Conflict of interest:
The authors declared no conflicts of interest.
Funding
This project did not receive any specific funding.

KEY WORDS | Arteriovenous malformation, Ultrasonography, Masseter

REFERENCES

1. Eerola I, Boon LM, Mulliken JB, et al. Capillary malformation-arteriovenous malformation, a new clinical and genetic disorder caused by RASA1 mutations. Am J Hum Genet 2003; 73(6): 1240-1249.
2. Karankot, G., Arutla, R., Rajan, R., & Reddy, S. Multifocal arteriovenous malformation of face: A rare entity. *J Indian Acad Oral Med Radiol 2020; 32*: 77 - 80.
3. Lakkasetty YT, Malik S, Shetty A, Nakhaei K. Multiple vascular malformations in head and neck - Rare case report. J Oral Maxillofac Pathol 2014;18(1):137-142
4. Richter GT, Friedman AB. Hemangiomas and vascular malformations: current theory and management. Int J Pediatr. 2012; 2012: 645678.
5. Mulligan PR, Prajapati HJ, Martin LG, Patel TH. Vascular anomalies: classification, imaging characteristics and implications for interventional radiology treatment approaches. Br J Radiol. 2014; 87(1035): 20130392.
6. Nair, Sanjiv C., and Srinivasa R. Chandra. Management of Head and Neck Vascular Lesions: A Guide for Surgeons. 2022. <https://doi.org/10.1007/978-981-15-2321-2>.
7. Kim JB, Lee JW, Choi KY, et al. Clinical Characteristics of Arteriovenous Malformations of the Head and Neck. Dermatol Surg. 2017;43(4):526-533.

8. Rai P, Setia S, Kalra N, Upreti L. Intramuscular vascular malformation of the masseter muscle presenting with turkey wattle sign. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2006; 102(5):618.
9. Lee JW, Chung HY. Vascular anomalies of the head and neck: current overview. Arch Craniofac Surg 2018; 19(4): 243-247.
10. Kumar A, Mittal M, Srivastava D, Jaetli V, Chaudhary S. Arteriovenous Malformation of Face. Contemp Clin Dent 2017;8(3):482-484.

READY - MADE CITATION

Karthikeya Patil, Meera Theenathayalan, Eswari Solayappan. A 46-year-old female with swelling in right cheek. *Hell J Radiol* 2024; 9(1): 50-55.