Intramuscular Hemangioma of the Masseter and Mentalis Muscles with the Turkey Wattle Sign and Phleboliths-A Case Report

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Abstract
Intramuscular hemangiomas are uncommon benign tumors in the head and neck region. The masseter muscle is the most commonly involved muscle in the orofacial region, but simultaneous involvement of both the mentalis and masseter muscles has not yet been reported. Preoperative diagnosis of such lesions is difficult because of the rarity of such tumors, their deep intramuscular location, and their unfamiliar clinical presentation. A case of intramuscular hemangioma of the masseter and mentalis muscles in a young Indian male patient is presented with a review of the role of various imaging modalities in such tumors.

Introduction
Vasoformative tumor is a broad term that includes hemangiomas and vascular malformations. The recent concept of hemangiomas is based on rapid endothelial cell proliferation in early infancy, followed by involution. Vascular malformations are characterized by a normal endothelial cell growth cycle that affects the vascular system, i.e., veins, capillaries, and lymphatics, and it does not show involution (1). Developmentally, hemangiomas are considered to be hematomas arising from abnormal embryonic rests. These pathologic tissues often go undetected for long periods and may become noticeable in the second decade of life due to proliferation of a pre-existing embryonic malformation, which can be stimulated by trauma to the site or hormonal changes (2).

Hemangiomas are uncommon benign vascular tumors affecting skeletal muscle in less than 1% of cases; of these, less than 15% occur in the head and neck region. The masseter is the most commonly involved muscle, followed by the trapezius and sternocleidomastoid muscles (3). An intramuscular hemangioma of the head and neck region involving the masseter muscle is often mistaken for a parotid swelling, and accurate preoperative diagnosis is achieved clinically in only less than 8% of cases (4).

A case of intramuscular hemangioma of the masseter and mentalis muscles in a young male is presented to illustrate the typical clinical presentation along with its radiological features. This case report emphasizes the importance of radiological investigation in such cases for accurate preoperative diagnosis.

Case Report
A 27-year-old Indian man complained of a 5-month history of swelling in the left cheek region. The patient noted that the swelling was more prominent on lying down on the bed and on clenching his teeth. The swelling had not changed in shape or size since it was first noticed. The patient gave no history of difficulty with mastication, pain, or discharge with respect to the swelling. Extra-oral examination revealed a solitary, ovoid in shape, well-defined swelling extending 6 cm anterior to the left tragus and 4 cm inferior to the left zygoma in the left masseter region, measuring 2 cm superioinferiorly and 1 cm mediolaterally in its greatest dimension on clenching the teeth (Turkey wattle sign). There were no visible pulsations. The overlying skin was normal. (Figs. 1, 2) On palpation, the swelling was rubbery
inconsistency and not tender. There was no local increase in temperature, and the mass was not fixed to underlying structures. Intra-oral examination revealed no significant findings. Informed consent was obtained from the patient, and the fine needle aspiration cytology (FNAC) procedure was explained to the patient. The FNAC procedure was done, and the aspirate of the swelling yielded fresh blood. Cytological analysis showed blood with reactive hematogenous cells, with no epithelial or lymphoid cells (Fig.3). A panoramic radiograph showed multiple (4-7 in number) radiopacities, well-defined, rounded to ovoid in shape, varying in size (0.5-1mm), along the left ramus of the mandible slightly above the mandibular notch and in the mandibular anterior body, and characterized by concentric lamellae of calcified material separated by radiolucent bands (Fig.4). A similar radiographic appearance was seen on a lateral oblique radiograph of the left ramus of the mandible (Fig.5). A mandibular occlusal radiograph showed 4 radiopacities, well-defined, rounded to ovoid in shape, varying in size (0.5-1mm), beyond the confines of adjacent bone on the lingual side in the area of teeth 34-37, and characterized by concentric lamellae of calcified material separated by radiolucent bands (Fig.6). Color Doppler ultrasound features were suggestive of hemangioma with phleboliths related to the left masseter and mentalis muscles (Figs.7-10).

MRI was performed with a 1.5-Tesla whole body MRI scanner. Axial T1 and T2, coronal T1 and STIR were obtained through the neck. A contrast-enhanced dynamic study of the neck was performed, and additional post-contrast axial, coronal, and sagittal T1-weighted images (T1WI) were obtained. A lobulated, soft tissue lesion with internal septations within the left masseter was seen along
with hemorrhagic foci and calcifications. The lesion was intermediate to slightly hyperintense on T1 (compared to surrounding muscles) and hyperintense on STIR images. There were oval foci of hypointensity on T2-weighted images (T2WI) scans within the lesion. A dynamic contrast study showed subtle patchy enhancement in the arteriovenous phase with progressive fill-in of the lesion on sequential venous phase images and complete opacification of the lesion except for oval hemorrhagic areas in the delayed phase. These findings favored the diagnosis of intramuscular hemangioma with calcification in the masseter and mentalis muscles (Fig.11A, B, C). The patient was informed of his condition, and treatment options were discussed, but he elected to forgo treatment at that time.
Discussion

Intramuscular hemangioma affecting skeletal muscles represents 0.8% of all benign vascular neoplasms, with 13.8% of these found in the head and neck region. The masseter is the most commonly involved muscle, followed by the trapezius and sternocleidomastoid muscles, and mentalis involvement is rarely reported (3-5).

A higher male prevalence was reported in studies by Wolf and Daniel (3, 4), whereas Sund and Ingelies cited a higher female prevalence (5, 9, 10). The present case report involved a lesion in a male patient. Clinically, the lesion presents as a slow growing soft tissue mass in the parotid region, occasionally associated with pain. Palpation of the mass may reveal a diffuse, soft swelling, compressible to firm in consistency, without any surface discoloration, bruits, or thrills. Preoperative diagnosis is inaccurate in more than 90% of cases for the following reasons: tumor rarity, deep intramuscular location, lack of specific signs suggesting vascular etiology (discoloration of overlying skin, presence of pulsations, bruits, thrills), and lack of sensitization of dental professionals towards the characteristic clinical/radiological presentations (7, 8).

The “turkey wattle sign” (8, 9) is an unusual pathognomonic manifestation in such lesions in which enlargement of a facial mass occurs on dependency of the head. This may be due to vascular engorgement within the lesion, which impedes venous return from the head to the superior vena cava. The turkey wattle is a red vascular structure in the neck of a male turkey that has the ability to increase in size when filled with blood (9). In the present case, the turkey wattle sign was the chief sign suggesting the possibility of a vascular lesion.

A rare characteristic manifestation within intramuscular hemangiomas is calcification either in the form of phleboliths or mature bone. In intramuscular hemangiomas, as a result of localized vascular stasis in tortuous vascular channels, platelet aggregation and formation of thrombus are initiated, resulting in calcification and formation of phleboliths. As reported previously, the incidence of phleboliths in such lesions ranges from 3-5%, and they are most commonly associated with cavernous hemangioma (10-13). In the present case, multiple phleboliths, 3-5 in number, were seen on an orthopantomogram and a mandibular occlusal radiograph.

The use of FNAC in hemangiomas is still a controversial procedure, because some reports stress the potential risk of hemorrhage following aspiration (14, 15), whereas other reports suggest that the hazards of aspiration in vascular lesions are overstated (15). According to a study done by Khurana and Mortelliti, FNAC is valuable not only in confirming the benign nature of a mass, but also in the
follow-up of such cases (16). In the present case, FNAC was done, and the aspirate showed blood.

**Imaging for intramuscular hemangiomas of the head and neck**

The role of imaging is vital in accurately assessing the extent of a lesion for better treatment planning and functional outcomes. Imaging studies including conventional radiographs and advanced imaging studies are helpful in the preoperative evaluation of intramuscular hemangiomas. The choice of imaging modality depends on factors such as the age of the patient, site and size of the lesion, and the availability and cost effectiveness of the technique.

Plain film imaging is advantageous in terms of ease of imaging and availability, affordability, and comparatively lower radiation exposure. Plain film findings may include calcifications in the form of either phleboliths, which appear as multiple, round or concentric in shape, randomly located radiopacities with a radiolucent center, or rarely ossification in the form of a Swiss cheese pattern suggestive of poorly defined ossification (18–21). The present plain film radiographic findings included calcification in the form of multiple phleboliths and absence of invasion of surrounding bone. These were similar to findings reported by other authors (17–19, 21). Calcified phleboliths may be seen on radiographs in only 25% of cases (22). Therefore, the diagnostic role of plain film radiography is limited in cases where calcifications are not present or in the absence of invasion into adjacent bony structures (20, 21).

Ultrasonography of intramuscular hemangiomas most commonly reveals a poorly defined mass with mixed echogenicity. Phleboliths are imaged as bright dots with posterior acoustic shadowing located within the hyperechoic component of the hemangioma; this finding was also seen in the present case. Color Doppler ultrasonography reveals a lesion with intratumoral hypoechogenicity with hypervascularity and abnormally low resistance arterial signals with forward flow during systole and diastole (23, 24). The use of color Doppler ultrasonography for intramuscular hemangiomas was first reported by Griffin et al. (25, 26). Ultrasound imaging has been suggested for the initial evaluation of intramuscular lesions, especially in cases where MRI cannot be done (26).

MRI of intramuscular hemangioma shows a mass of intermediate signal intensity on T1WI and high signal intensity on T2WI. According to Wu et al. (25), the internal structure of hemangiomas has a characteristic appearance that can be differentiated; the cavernous type shows lobulation, while the capillary type is serpiginous. Contrast-enhanced MRI shows serpiginous or lattice-like enhancement of the lesions. Calcifications within the lesion can be suggested by low intensity spots located in enhanced vascular channels on T2WI. In the present case, the MRI findings were similar to those reported previously. These findings could provide useful information to differentiate between cavernous and capillary hemangiomas. MRI is superior to all imaging modalities for intramuscular hemangioma, as it provides distinct contrast between normal muscle and hemangioma, along with the exact extent of the lesion (8, 10, 11).

Intramuscular hemangiomas can be differentiated from lesions such as salivary gland tumors, benign masseter hypertrophy, and myositis ossificans.

**Management of intramuscular hemangioma**

Management of intramuscular hemangioma in the head and neck region is based on tumor location, accessibility, depth of invasion, patient age, and cosmetic considerations. Medical therapy, sclerotherapy, lasers, cryosurgery, and surgical therapy, or combinations of these therapies, are among the various treatment modalities used for management of intramuscular hemangioma (Fig. 12). In general, active interventions such as surgery, laser therapy, and cryosurgery should be considered in all disfiguring hemangiomas. In uncomplicated lesions that are cosmetically
acceptable, benign neglect may be chosen together with careful follow-up for 4–5 years.

Medical therapy is indicated in the proliferative phase of lesions. High doses of steroids, either in the form of systemic or intralesional injections, are first-line treatment, which includes prednisolone at a dose of 20–30mg/day for 2 weeks to 4 months (7, 10, 13) and intralesional triamcinolone at a dose of 4mg/mL per week for 4 weeks (7, 10). Interferon alpha 2a has also been used, but there are concerns about its safety in children. Sclerotherapy is indicated in large lesions to debulk the tumor before surgery. Agents such as STS (sodium tetradecyl sulfate), ethanol, hypertonic saline, and sodium morrhuate are used in sclerotherapy. Laser therapy is advantageous in selective photothermolysis within the lesion rather than nonselective tissue destruction. Pulsed dye lasers, Nd-YAG lasers, argon lasers, and carbon dioxide lasers are used effectively in superficial hemangiomas. Cryotherapy is beneficial in the treatment of small intraoral lesions, but in cutaneous lesions it can result in hypopigmentation and scarring. The surgical approach to masseter muscle hemangiomas involves a preauricular approach using superficial skin flaps or intraoral excision (7, 10, 13). In the present case, the patient was informed about his condition, and the treatment options were discussed, but the patient elected to forgo treatment at that time.

Conclusion

Intramuscular hemangiomas are uncommon benign tumors with a low incidence of occurrence in the head and neck region. A lack of specific signs (pulsations, bruits, or thrills) suggesting a vascular etiology and a deep intramuscular location may lead to inaccurate preoperative diagnosis and inappropriate management for such cases. In cases of neoplastic-like swelling and the presence of the turkey wattle sign, an accurate preoperative diagnosis can be established only with radiographic intervention. Correlations with radiographic, ultrasonographic, MRI, or computed tomographic findings are essential for correctly diagnosing intramuscular hemangioma with ossification. This case report was presented in an attempt to increase awareness of the clinical and radiological presentation of such silent tumors. In addition, the radiographic findings described in this case report might prove useful in the accurate diagnosis of such conditions.

References


